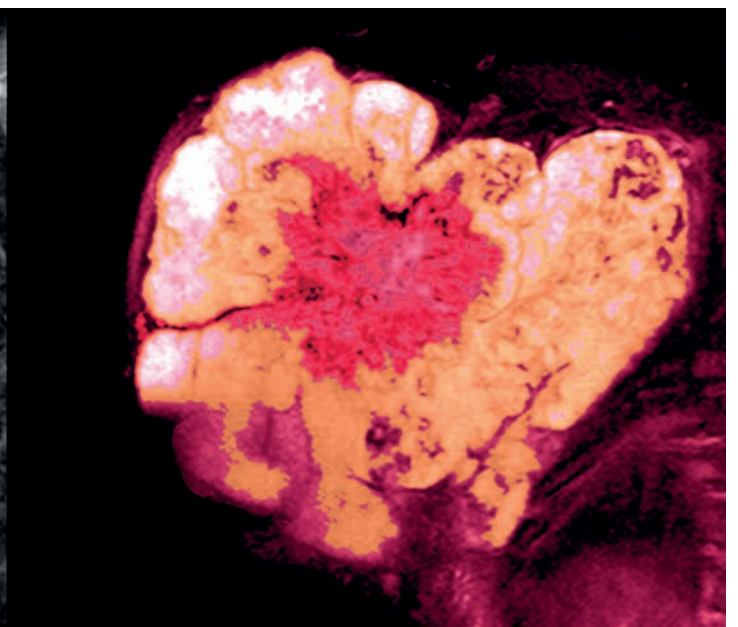
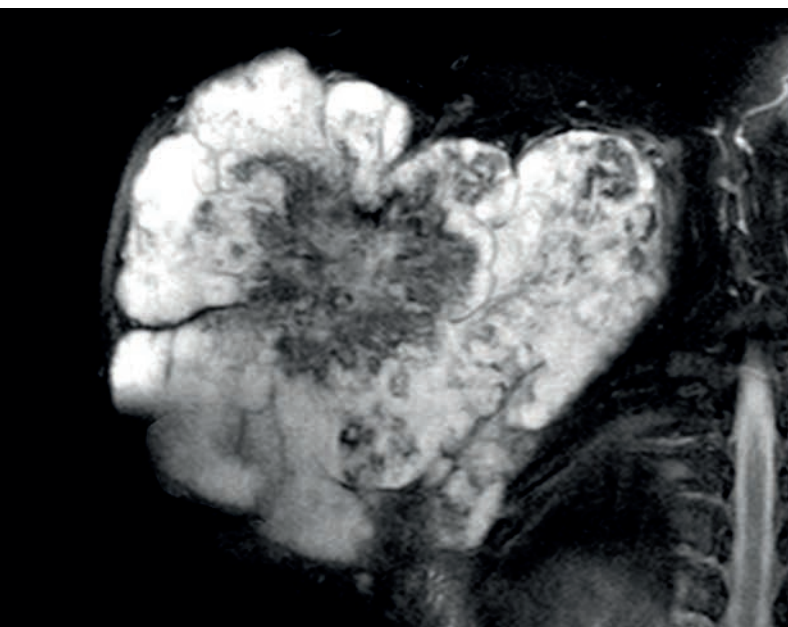
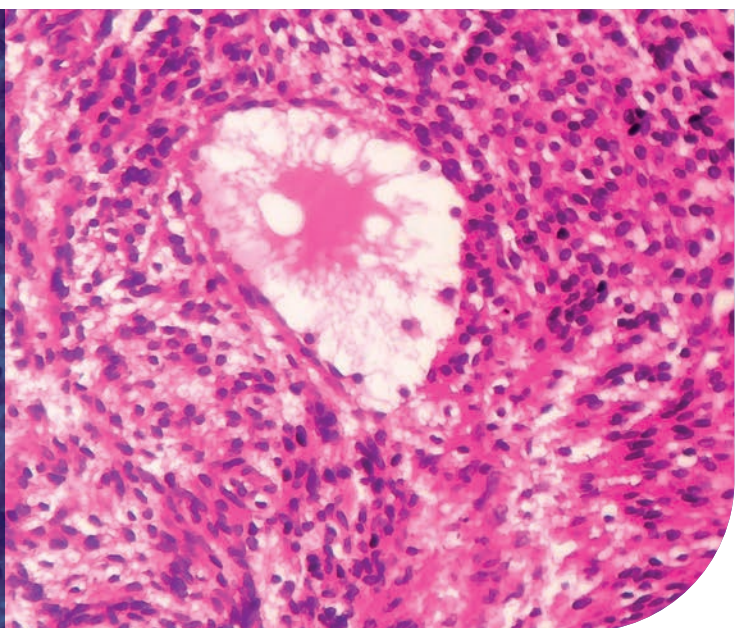


Belgian Cancer Registry



**Bone & soft
tissue tumour
epidemiology**
in Belgium 2004-2019



Cover page:

Top image: Leg x-ray showing advanced stage bone cancer at tibia with pathologic fracture.

Middle image: Sarcoma, MRI image of shoulder region. Black and white image as well as colored image with enhanced tumour lesion.

Bottom image: Microscopic picture of a sarcoma.

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LIST OF ACRONYMS

AAPC	Average Annual Percentage Change
AIDS	Acquired immunodeficiency syndrome
ALT	Atypical lipomatous tumour
APC	Annual Percentage Change
ARMS	Alveolar rhabdomyosarcoma
AYA	Adults and young adolescents
BCR	Belgian Cancer Registry
BSPHO	Belgian Society of Paediatric Haematology Oncology
CBSS	Crossroads Bank for Social Security
CI	Confidence interval
CR	Crude incidence rate
CRi	Cumulative risk
DDLPS	Dedifferentiated liposarcoma
DFSP	Dermatofibrosarcoma protuberans
EBV	Epstein-Barr Virus
ERMS	Embryonal rhabdomyosarcoma
ESR	European Standardised incidence rate
FNCLCC	French National Federation of Cancer Centers
FRANCIM	French network of cancer registries
GIST	Gastrointestinal stromal tumour
HIV	Human Immunodeficiency Virus-1
ICD-O-3	International Classification of Diseases for Oncology (3rd edition)
IMA	InterMutualistic Agency
IKNL	Netherlands Comprehensive Cancer Organization
INSZ-NISS	National social security number
M/F-ratio	Male/Female ratio
MLPS	Myxoid liposarcoma
MOC-COM	Multidisciplinary oncological consult
MPNST	Malignant peripheral nerve sheath tumour
N	Number of new diagnoses
NOS	Not otherwise specified
PEComa	Perivascular epithelioid cell tumour
PITTER	Predictive test for a therapeutic response
PNST	Peripheral nerve sheath tumour
SFT	Solitary fibrous tumour
WDLPS	Well-differentiated liposarcoma
WHO	World Health Organization
WSR	World Standardised incidence rate

1 INTRODUCTION

The main objective of this publication is to describe the incidence of malignant bone and soft tissue tumours, often referred to as sarcomas, in Belgium between 2004 and 2019, with a specific focus on the ten most recent years. This report provides the first complete overview of the sarcoma incidence in Belgium. Together with the population-based nature of the Belgian Cancer Registry, the completeness of the database provides an opportunity to get a comprehensive description of the incidence, prevalence, survival and trends over time of sarcomas in Belgium over a period of 16 years (2004-2019).

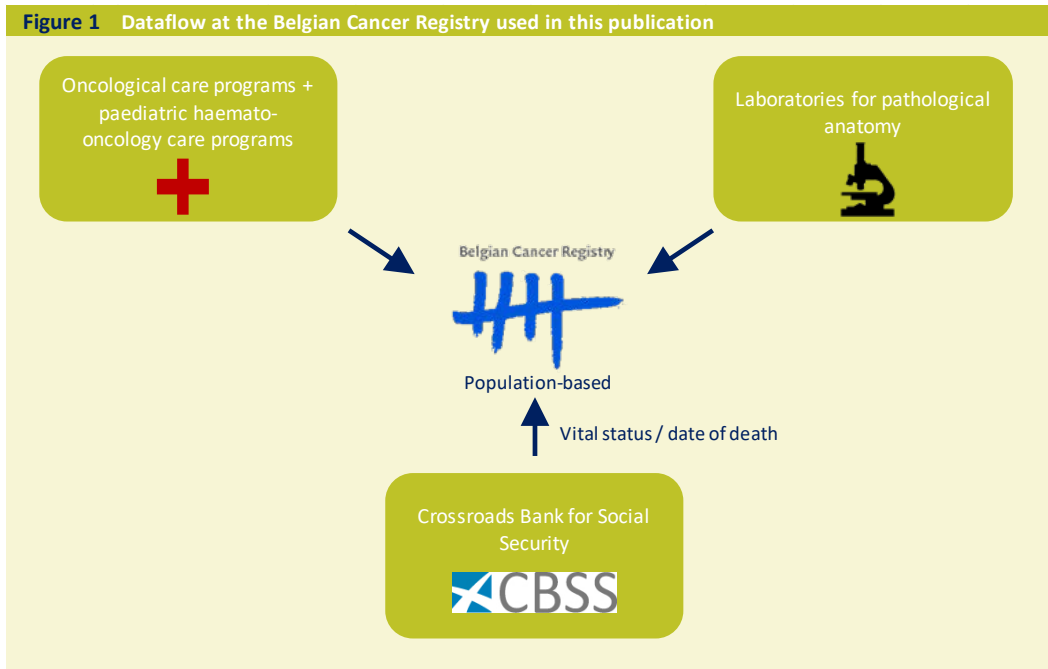
Sarcomas constitute a very rare group of tumours and represent less than one percent of malignancies worldwide⁽¹⁾. In addition, it is an extremely heterogeneous group of tumours, mostly considered as having a mesenchymal origin. The precise inclusion criteria are presented in the second chapter. The various sarcoma subtypes are characterised by very different behaviour and prognosis. Sarcomas can arise anywhere throughout the human body, originating from soft tissues of the limbs, head and neck region, trunk, including the viscera. The classification is based on the tumour cell differentiation⁽¹⁻³⁾, nevertheless, an important part of the sarcomas has an uncertain differentiation. For simplicity, all malignant bone and soft tissue tumours and related tumours will be designated as sarcomas in this report.

1.1 NOTIFICATION AND SUBMISSION TO THE CANCER REGISTRY

Since 2003, both new legislation initiatives and the foundation of the Belgian Cancer Registry (BCR) in 2005, forced a breakthrough in the Belgian cancer registration. Especially the Royal Decree on the oncological care programs in 2003 with the reimbursement of the multidisciplinary oncological consult (MOC-COM) and the creation of the specific law on the Cancer Registry in 2006 provided a firm legal basis for cancer registration in Belgium⁽⁴⁻⁵⁾. This legislation makes cancer registration compulsory, both for the oncological care programs and for the laboratories for pathological anatomy and clinical biology / haematology. From 2004 onwards, Belgian cancer incidence data are available. The general data flow (**figure 1**) thus relies on information (notifications) coming from the oncological care programs, the paediatric haematology-oncology care programs ('clinical network') and the laboratories for pathological anatomy ('pathology network').

Furthermore, the law authorises the use of the national social security number (INSZ-NISS) as unique identifier of the cancer patient as well as linkage with other medical and/or administrative databases. Through linkage with the Crossroads Bank for Social Security (CBSS), the Cancer Registry can perform active follow-up of the vital status and date of death of the cancer patients. The methodology of the cancer data registration and collection related to the hospitals and the pathology laboratories has been previously reported in several publications⁽⁶⁻¹⁵⁾.

Figure 1 Dataflow at the Belgian Cancer Registry used in this publication



1.2 PRIVACY & PROTECTION OF PERSONAL DATA

The core business of the Belgian Cancer Registry (BCR) includes the collection and processing of sensitive personal data in order to fulfil its legal obligations as stated in the Coordinated Act of 10 May 2015 on the exercise of health care professions. Consequently, BCR attaches great importance to privacy and the protection of personal data and has taken strict measures to comply with the General Data Protection Regulation EU 2016/679. For more information, please read the Privacy Statement available on our website (https://belgiancancerregistry.be/privacy_en).

2 METHODS & DATA QUALITY

2.1 CLASSIFICATION & REPORTING: MALIGNANT BONE, SOFT TISSUES & RELATED TUMOURS

Malignant bone, soft tissue and related tumours are an extremely heterogeneous group of tumours and will be designated as sarcomas in this report. To organise this diverse group, two different classifications are applied throughout this publication:

- A traditional subdivision is made by primary tumour location (**chapter 3.1**) into soft tissue and visceral sarcomas on the one hand, and bone sarcomas on the other.
- A subdivision by histological tumour type (**chapter 3.2**) differentiates the various microscopically distinct tumour types and was inspired by the two latest WHO classifications of Soft Tissue and Bone Tumours^(1,2) and the work of the French (FRANCIM⁽¹⁶⁾) and Dutch cancer registries (IKNL⁽¹⁷⁾).

The WHO classification of sarcomas has evolved over time and in 2020 the fifth and most recent edition was published⁽¹⁾. The data presented in this report (2004-2019) were registered according to the WHO edition in use at the time of diagnosis (third published in 2002, fourth in 2013)^(2,18).

Table 1 provides an overview of all histological tumour types included in this publication, with the time period during which these codes are applicable. This 'retrospective' table also includes entities and categories for poorly specified registrations (e.g. sarcoma not otherwise specified), that currently are obsolete. To address the progressive insights in the classification of sarcomas and the corresponding changes between the consecutive editions of the WHO classification, the editors decided to include a selection of entities which were formerly classified as malignant but today are considered benign or of intermediate biologic potential. As registration of benign tumours and tumours of intermediate biological potential is, for most cases, not obligatory at the Belgian Cancer Registry, trends of these tumour registrations were assessed to detect a potential impact of registrations changes on the incidence over time.

Tumours with intermediate biologic potential correspond to borderline tumours in the ICD-O-3 classification. They include both locally aggressive and rarely metastasizing tumours.

Chapters that include tumour entities with a non-malignant behaviour are:

- Liposarcoma: atypical lipomatous tumours with intermediate biologic potential behaviour were grouped with well differentiated liposarcomas not otherwise specified (NOS), as they belong to the same entity after systematic revision.
- Dermatofibrosarcoma protuberans (DFSP): this entity was previously regarded as malignant, but the behaviour was changed to 'borderline' (i.e. intermediate biologic potential) in the 5th edition of the WHO classification (exception for the fibrosarcomatous subtype). The editors opted to include DFSP as this publication concerns the period 2004-2019 and offers an excellent opportunity to discuss the epidemiology of this rather frequently occurring subgroup of mesenchymal tumours.
- Myoepithelioma: since 2020, only the myoepithelial carcinoma subtype is regarded as malignant. However, as this publication concerns the period 2004-2019 also myoepithelioma is included.

In appendix I, an up to date 'prospective' classification is provided to facilitate the precise coding of sarcomas, considering the changes introduced by the 5th edition of the WHO classification and integrations in the last version of the ICD-O-3 classification^(1,3). We warmly recommend everyone involved in cancer registration to consult this 'prospective table' for future registrations.

Table 1 Classification of tumours of soft tissues/bones (inclusion criteria used in this publication)¹

Sarcomas classified by histological type	Classification ICD-O-3	Period during which the code applies
Liposarcoma		
Atypical lipomatous tumour / well-differentiated liposarcoma (ALT/WDLPS)		
Atypical lipomatous tumour	8850/1	2002 and later
Liposarcoma, well differentiated, NOS	8851/3	1992 and later
Dedifferentiated liposarcoma (DDLPS)		
Dedifferentiated liposarcoma	8858/3	1992 and later
Myxoid liposarcoma (MLPS)		
Myxoid liposarcoma	8852/3	1992 and later
Round cell liposarcoma	8853/3	1992 and later
Liposarcoma NOS & other		
Liposarcoma, NOS	8850/3	1992 and later
Pleomorphic liposarcoma	8854/3	1992 and later
Mixed liposarcoma	8855/3	1992 and later
Fibroblastic liposarcoma	8857/3	2002 and later
Myxoid pleomorphic liposarcoma	8859/3	/
Malignant (myo)fibroblastic tumours and so-called fibrohistiocytic tumours		
Dermatofibrosarcoma protuberans (DFSP)		
Dermatofibrosarcoma protuberans, NOS	8832/3; 8832/1	8832/3: 1992-2019; 8832/1: 2013 and later
Dermatofibrosarcoma protuberans, fibrosarcomatous	8832/3	2013 and later
Pigmented dermatofibrosarcoma protuberans	8833/3; 8833/1	8833/3: 1992-2019; 8833/1: 2013 and later
Solitary fibrous tumour (SFT)		
Solitary fibrous tumour	8815/3	2002 and later
Fibrosarcoma		
Fibrosarcoma, NOS	8810/3	1992 and later
Fascial fibrosarcoma	8813/3	1992 and later
Infantile fibrosarcoma	8814/3	1992 and later
Periosteal fibrosarcoma	8812/3	1992 and later
Myxofibrosarcoma		
Myxofibrosarcoma	8811/3	1992 and later
Miscellaneous malignant (myo)fibroblastic tumours and so-called fibrohistiocytic tumours		
Low-grade myofibroblastic sarcoma	8825/3	2013 and later
Low grade fibromyxoid sarcoma / sclerosing epithelioid fibrosarcoma (myxosarcoma)	8840/3	1992 and later
Malignant giant cell tumour of soft parts	9251/3	1992 and later
Malignant tenosynovial giant cell tumour	9252/3	2002 and later
Vascular sarcoma		
Kaposi sarcoma		
Kaposi sarcoma	9140/3	1992 and later
Angiosarcoma		
Hemangiosarcoma	9120/3	1992 and later
Kupffer cell sarcoma	9124/3	1992 and later
Lymphangiosarcoma	9170/3	1992 and later
Haemangioendothelioma		
Haemangioendothelioma, malignant	9130/3	1992 and later
Epithelioid haemangioendothelioma, NOS	9133/1; 9133/3	9133/1 2002 and later; 9133/3: 2020 and later
Mvopericvtoma. including mvofibroma	8824/0	2002 and later
Leiomyosarcoma		
Leiomyosarcoma, NOS	8890/3	1992 and later
Epithelioid leiomyosarcoma	8891/3	1992 and later
Myxoid leiomyosarcoma	8896/3	1992 and later
Rhabdomyosarcoma		
Rhabdomyosarcoma, NOS	8900/3	1992 and later
Pleomorphic rhabdomyosarcoma, adult type	8901/3	1992 and later
Mixed type rhabdomyosarcoma	8902/3	1992 and later
Embryonal rhabdomyosarcoma, NOS	8910/3	1992 and later
Spindle cell rhabdomyosarcoma	8912/3	2002 and later
Alveolar rhabdomyosarcoma	8920/3	1992 and later
Ectomesenchymoma	8921/3	2002 and later
Peripheral nerve sheath tumour		
Malignant peripheral nerve sheath tumour, NOS	9540/3	1992 and later
Malignant melanotic nerve sheath tumour		
Malignant peripheral nerve sheath tumour, epithelioid	9542/3	2013 and later
Schwannoma	9560/3	1992 and later
Malignant peripheral nerve sheath tumour with rhabdomyoblastic differentiation	9561/3	1992 and later
Malignant Triton tumour		
Perineurioma, malignant	9571/3	2002 and later
Granular cell tumour, malignant	9580/3	1992 and later

Sarcomas classified by histological type	Classification ICD-O-3	Period during which the code applies
Other tumours of uncertain differentiation		
Synovial sarcoma		
Synovial sarcoma, NOS	9040/3	1992 and later
Synovial sarcoma, spindle cell	9041/3	1992 and later
Synovial sarcoma, epithelioid cell	9042/3	1992 and later
Synovial sarcoma, biphasic	9043/3	1992 and later
Myoepithelioma		
Myoepithelial carcinoma	8982/3	2002 and later
Myoepithelioma, myoepithelial carcinoma, and mixed tumour	8982/0	1992 and later
Rhabdoid tumours		
Rhabdoid tumour, NOS	8963/3	1992 and later
Atypical teratoid/rhabdoid tumour	9508/3	2002 and later
Remaining other tumours of uncertain differentiation		
Desmoplastic small round cell tumour	8806/3	2002 and later
Alveolar soft part sarcoma	9581/3	1992 and later
Clear cell sarcoma, NOS	9044/3	1992 and later
Clear cell sarcoma of kidney	8964/3	1992 and later
PEComa, malignant	8714/3	2013 and later
Mesenchymoma, malignant	8990/3	1992 and later
Phosphaturic mesenchymal tumour, malignant		
Embryonal sarcoma	8991/3	1992 and later
Biphenotypic sinonasal sarcoma	9045/3	New 2020
Intimal sarcoma	9137/3	2013 and later
Epithelioid sarcoma, NOS	8804/3	1992 and later
Mixed tumour, malignant, NOS	8940/3	1992 and later
Ossifying fibromyxoid tumour, malignant	8842/3	2013 and later
NTRK-rearranged spindle cell neoplasm (emerging)		
Gastrointestinal stromal tumour (GIST)		
Gastrointestinal stromal tumour	8936/3	2002 and later
Gastrointestinal stromal tumour, benign	8936/0	2002-2019
Gastrointestinal stromal tumour, borderline	8936/1	2002-2019
Endometrial stromal sarcoma		
Endometrial stromal sarcoma, NOS	8930/3	1992 and later
Endometrial stromal sarcoma, low grade	8931/3	2002 and later
Undifferentiated small round cell sarcomas of bone and soft tissue		
Ewing sarcoma	9364/3	New 2020
Ewing sarcoma	9260/3	1992-2019
Askin tumour	9365/3	2002 and later
Chondrosarcoma		
Chondrosarcoma, NOS	9220/3	1992 and later
Chondrosarcoma, NOS, grade 2 & 3	9220/3	New 2020
Periosteal chondrosarcoma (juxtacortical)	9221/3	1992 and later
Mesenchymal chondrosarcoma	9240/3	1992 and later
Clear cell chondrosarcoma	9242/3	2002 and later
Dedifferentiated chondrosarcoma	9243/3	2002 and later
Myxoid chondrosarcoma	9231/3	1992 and later
Osteosarcoma		
Osteosarcoma, NOS	9180/3	1992 and later
Chondroblastic osteosarcoma	9181/3	1992 and later
Fibroblastic osteosarcoma	9182/3	1992 and later
Telangiectatic osteosarcoma	9183/3	1992 and later
Osteosarcoma in Paget disease of bone	9184/3	1992 and later
Small cell osteosarcoma	9185/3	1992 and later
Central osteosarcoma, NOS	9186/3	2002 and later
Low grade central osteosarcoma	9187/3	2002 and later
Parosteal osteosarcoma	9192/3	2002 and later
Periosteal osteosarcoma	9193/3	2002 and later
High grade surface osteosarcoma	9194/3	2002 and later
Intracortical osteosarcoma	9195/3	2002 and later
Other bone tumours of uncertain differentiation		
Conventional chordoma	9370/3	1992 and later
Poorly differentiated chordoma		
Chordoma, NOS		
Chondroid chordoma	9371/3	2002 and later
Dedifferentiated chordoma	9372/3	2002 and later
Adamantinoma of long bones	9261/3	1992 and later
Unclassified and poorly characterised sarcoma		
Myosarcoma	8895/3	1992 and later
Stromal sarcoma, NOS	8935/3	2002 and later
Sarcoma, NOS	8800/3	1992 and later
Spindle cell sarcoma	8801/3	1992 and later
Pleomorphic or Giant cell sarcoma (except of bone 9250/3)	8802/3	1992-2020
Pleomorphic or Giant cell sarcoma	8802/3	New 2020
Small cell sarcoma	8803/3	1992 and later
Undifferentiated sarcoma	8805/3	2002 and later
Malignant fibrous histiocytoma	8830/3	1992 and later

Source: Belgian Cancer Registry 

ⁱRecently the classification of tumours of soft tissues and bones has been updated. The correct inclusion criteria to be applied starting from incidence year 2020 are presented in the Appendices.

2.2 QUALITY OF INCIDENCE DATA

2.2.1 COMPLETENESS OF THE CANCER REGISTRY

Notifications from multiple sources

Combining registrations from independent sources ensures a more complete cancer registration and allows for the detection of registration errors and the inclusion of partly different sets of variables. As explained in the introduction (**chapter 1**), the Belgian Cancer Registry combines data from two main sources: the oncological care programs and the paediatric haematology-oncology care programs ('clinical network') and the laboratories for pathological anatomy ('pathology network').

In 2019, the BCR has recorded 71,651 new cancer diagnoses (excl. non-melanoma skin cancer) for official Belgian residents. Of all these cancers, 94% was covered by the clinical network and 90% via the pathological network which resulted in an overlap of 84%. The percentage of notifications by the clinical network has significantly increased over time (**figure 1**). The pathological network shows, after an initial rapid increase, a stable percentage of notifications over time of circa 90% (a biopsy or resection specimen is not always available). Of all cancers that were registered in 2019 (both networks combined), microscopic confirmation was indicated for over 96% of cases. Specifically for sarcomas, this concerns over 99% of the registrations.

Sarcomas show a lower percentage of registrations by the clinical network when compared to all cancer types combined (**figure 1**). Although the improvement over time of clinical sarcoma registrations resembles the trend observed for all cancers combined, only 88% of all sarcomas are registered by the clinical network for incidence year 2019. This 6-percentage points difference between clinical registration of sarcomas and all cancers combined is remarkable. There could be several explanations for this finding.

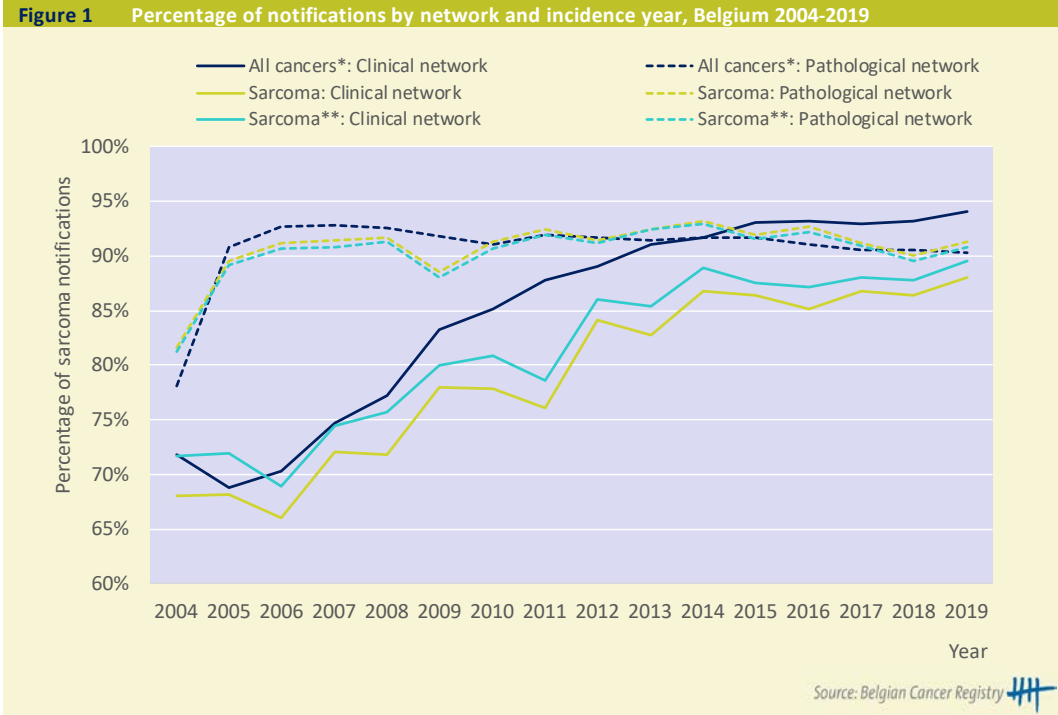
A first explanation is the diverse nature of sarcomas (**table 2**). Rhabdoid tumours for example, which almost exclusively occur in paediatric patients, show a percentage of clinical notifications of 100%. Dermatofibrosarcoma protuberans (DFSP) on the other hand has one of the lowest percentages (74%) of clinical notifications. DFSP has a 10-year relative survival of around 100% (see **chapter 3.2.2.1**) and most subtypes are no longer being regarded as malignant (see **chapter 2.1**). This tumour type might therefore be more prone to escape cancer registration in the hospitals. Furthermore, a lot of dermatologists have a private practice and do not attend a MOC/COM and have no legal basis or obligation for participating in cancer registration when working outside of an oncological care program. To illustrate this, separate curves for sarcomas *without* DFSP and Kaposi sarcoma are included in **figure 1**, showing a slightly better completeness for the clinical sarcoma registration.

A second and partly overlapping explanation, are the cases discussed at a multidisciplinary oncology team meeting (MOC/COM) (**figure 2**). Although mandatory cancer registration is not limited to tumours discussed at a MOC/COM, tumours which are not discussed are more likely to escape registration by oncological care programs. Patient referral to a secondary or tertiary centre might also implicate that a new MOC/COM might not be reimbursed and consequently be (wrongly) forgotten or not flagged for cancer registration. In 2018, 81% of all sarcoma cases was discussed at a MOC/COM. The evolution over time of the percentage of cases discussed at a MOC/COM (**figure 2**) shows a similar trend as the percentage of notifications by the clinical network (**figure 1**). Sarcoma types with a lower percentage of clinical notifications generally correspond to those that are less often discussed in a MOC/COM (**table 2**). An association between MOC/COM discussion and more complete registrations by the clinical network is also observed by age group (**figure 3 & 4**). In children, sarcomas are almost always discussed at a MOC/COM and accordingly show a very high (>95%) percentage of clinical notifications. The paediatric haematology-oncology care programs are actively involved in cancer registration and there is a close collaboration between the BCR and the Belgian Society of Paediatric Haematology Oncology (BSPHO). There is a considerable drop in the percentage of registrations by the clinical network and in the percentage of cases discussed in a MOC/COM discussion after the age of 15 year. The lowest percentages are observed for adults and young adolescents (AYA) who are probably treated in an adult care setting.

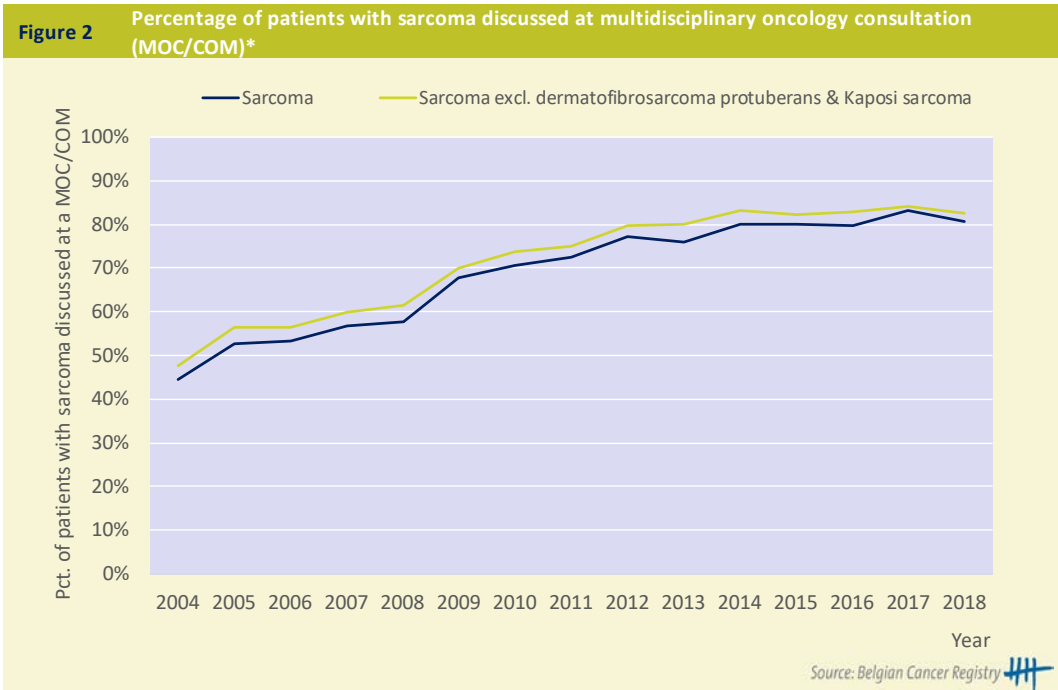
An additional element is the inclusion of non-malignant entities (**table 1**) in this publication for which cancer registration is not always mandatory. However, consistency of incidence trends upon changes in the coding of tumour behaviour was assessed (as mentioned in **chapter 2.1**) and this only has a marginal impact of around 0.5 percentage points.

Another possible cause are incidental findings or so-called *whoops* procedures. Especially for soft tissue sarcoma, some cases are only identified as being a sarcoma during histological analysis after resection. Due to the very rare nature of sarcomas and difficulties in diagnosis, a surgeon might not be aware he or she is resecting a sarcoma which might explain the case was not discussed on a MOC/COM and hence preventing a cancer registration for this case. If information from the pathology report after surgery is not picked up by the hospital's cancer registration team, these incidental findings contribute to the lower number of notifications to the BCR by the clinical network.

The combination of data from the clinical and pathological network provides a valuable tool to achieve a complete database. However, the lower percentage of clinical notifications demands attention and strongly suggests room for improvement in the sarcoma cancer registration.



* All cancers excl. non-melanoma skin cancer **All sarcomas excl. dermatofibrosarcoma protuberans and Kaposi sarcoma



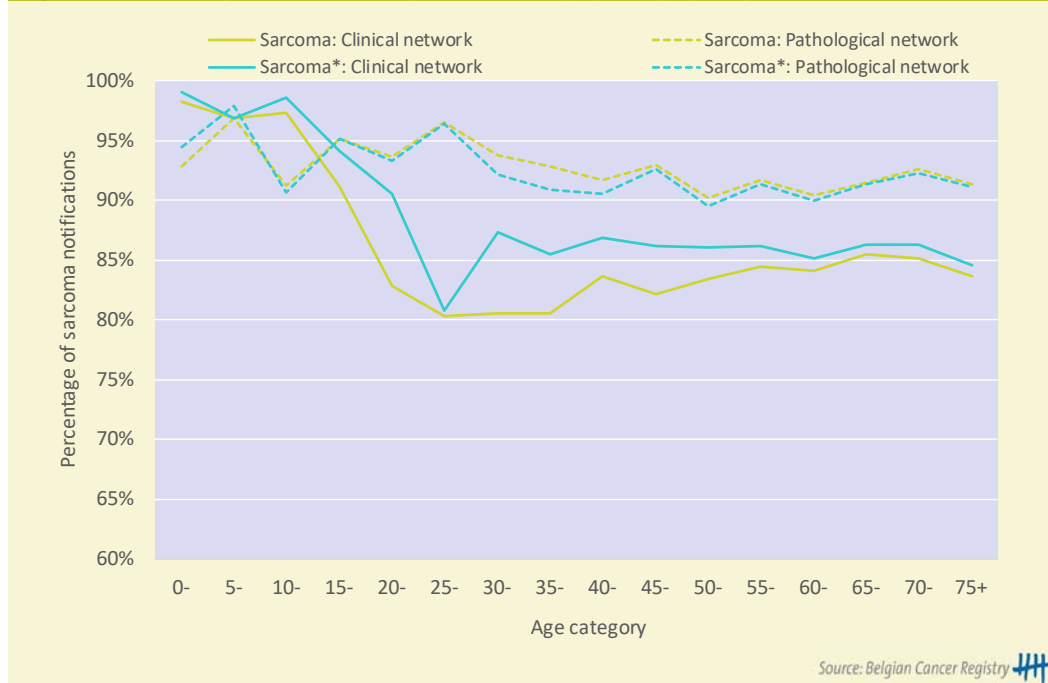
* Multidisciplinary oncology team meeting. Based on MOC/COM nomenclature codes using data from the Intermutualistic Agency (IMA) in a period of 1 month before until 6 months after diagnosis

Table 2 Percentage of registrations made by the clinical and pathological networks, and percentage of patients discussed at a MOC/COM* 2010-2019

	Clinical network	Pathological network	MOC/COM
3. All sarcomas	84%	92%	77%
3.1 Sarcomas classified by primary tumour location			
3.1.1 Soft tissue and visceral sarcoma	84%	92%	77%
3.1.2 Bone sarcoma	90%	94%	77%
3.2 Sarcomas classified by histological type			
3.2.1 Liposarcoma	86%	92%	78%
3.2.2 Malignant (myo)fibroblastic tumours and so-called fibrohistiocytic tumours	80%	91%	71%
3.2.2.1 Dermatofibrosarcoma protuberans	74%	95%	63%
3.2.2.2 Solitary fibrous tumour	87%	65%	84%
3.2.2.3 Fibrosarcoma	83%	86%	75%
3.2.2.4 Myxofibrosarcoma	86%	96%	75%
3.2.3 Vascular sarcoma	70%	95%	62%
3.2.3.1 Kaposi sarcoma	56%	97%	40%
3.2.3.2 Angiosarcoma	84%	95%	81%
3.2.4 Leiomyosarcoma	88%	96%	79%
3.2.5 Rhabdomyosarcoma	93%	97%	87%
3.2.6 Peripheral nerve sheath tumours	93%	82%	84%
3.2.7 Other tumours of uncertain differentiation	88%	92%	82%
3.2.7.1 Synovial sarcoma	87%	96%	78%
3.2.7.2 Myoepithelioma	77%	90%	77%
3.2.7.3 Rhabdoid tumours	100%	92%	97%
3.2.8 Gastrointestinal stromal tumour	83%	90%	80%
3.2.9 Endometrial stromal sarcoma	89%	93%	83%
3.2.10 Ewing sarcoma	95%	96%	90%
3.2.11 Chondrosarcoma	85%	94%	71%
3.2.12 Osteosarcoma	94%	94%	84%
3.2.13 Other bone tumours of uncertain differentiation	88%	92%	73%
3.2.13.1 Chordoma	90%	91%	80%
3.2.14 Unclassified and poorly characterised sarcoma	87%	87%	79%

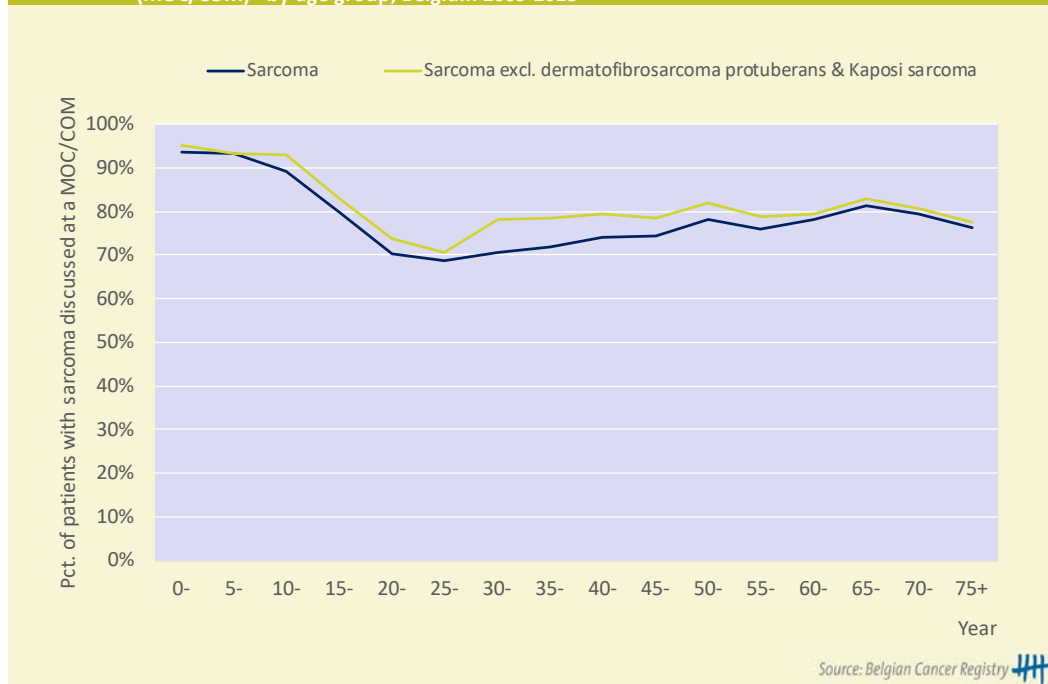
* Multidisciplinary oncology team meeting (data included for 2010-2018)

Figure 3 Percentage of notifications by network and by age category, Belgium 2010-2019



*All sarcomas excl. dermatofibrosarcoma protuberans and Kaposi sarcoma

Figure 4 Percentage of patients with sarcoma discussed at a multidisciplinary oncology consultation (MOC/COM)* by age group, Belgium 2009-2018



* Multidisciplinary oncology team meeting. Based on MOC/COM nomenclature codes using data from the Inter-mutualistic Agency (IMA) in a period of 1 month before until 6 months after diagnosis

2.2.2 VALIDITY

Quality checks


The cancer registry validates the data quality on a regular basis^(13,19). In the context of this publication, the BCR performed additional quality checks. More than 2,200 cases from all incidence years (2004-2019) were selected for an additional specific data cleaning effort. Focus points included a more precise classification of not well specified histological sarcoma types whenever possible (e.g. sarcoma, not otherwise specified), coding of liposarcoma, verification of atypical primary tumour locations, correct coding of tumour behaviour (malignant vs. non-malignant), coding of central nervous system embryonal tumours, and several other smaller topics.

Stability of incidence data over time

As a result of delays in notification or by recovering additional information not available at the time of registration, the incidence for a given year will change over time. Due to the continuous and thorough data cleaning, additional data is incorporated at a later date resulting in small changes over time in the number of new diagnoses for the same incidence year. Very often, the number of cases in the first year after publication will increase due to the inclusion of 'late arrivals', while later on, the number of cases decreases a little due to the thorough and consistent data cleaning that results in for example the exclusion of cases that after additional investigations were confirmed as non-malignant. The incidence for all sarcomas (**table 3**) remains fairly stable and rarely exceeds 1% change between 2 consecutive publication years.

Table 3 Sarcomas: Stability of incidence data (N) over time, Belgium 2004-2019

Publication year	Incidence year																
	2004	2005	2006	2007	2008	2009	2010	2011	2012	2013	2014	2015	2016	2017	2018	2019	
2004	885																
2005	883	863															
2006	916	887	809														
2008	915	884	827	959	953												
2009	917	882	832	944	947	819											
2010	919	884	832	946	956	829	958										
2011	920	887	833	951	958	838	960	1002									
2012	915	874	818	935	933	809	930	994	1070								
2013	934	911	876	1038	1030	896	933	984	1060	1046							
2014	925	905	867	1039	1032	903	941	988	1055	1047	1189						
2015	920	905	866	1034	1029	903	945	984	1050	1048	1167	1171					
2016	919	908	875	1041	1041	917	958	991	1073	1064	1186	1196	1169				
2017	918	906	874	1040	1041	911	957	986	1063	1062	1186	1198	1168	1157			
2018	926	908	881	1044	1043	913	963	994	1069	1064	1192	1208	1175	1164	1193		
2019	912	896	877	1031	1030	905	958	990	1061	1059	1186	1202	1165	1162	1184	1171	

Source: Belgian Cancer Registry 

2.3 CALCULATION OF INCIDENCE, TRENDS, PREVALENCE AND SURVIVAL

2.3.1 INCIDENCE

Incidence is the number of new cases occurring in a given time period in a specific population. It can be used to estimate the probability or risk of illness and can be expressed in different ways. The incidence data presented in the current publication encompass the time period 2004-2019.

- The **crude incidence rate (CR)** is calculated by dividing the number of new cases observed during a given time period by the corresponding population time at risk in that time period. The crude rate is expressed as the number of new cases per 100,000 person years.
- The **age-specific incidence rate** is the crude incidence rate in a particular 5-year age group and expressed per 100,000 person years.
- The **age-standardised incidence rate** is a weighted average of the individual age-specific rates using an external standard population. It is the incidence that would be observed if the population had the age structure of the standard population (European (1976) or World Standard Population). Since age has a powerful influence on the risk of cancer, this standardisation is necessary when comparing several populations that differ with respect to their age structure. In this publication, the World Standard Population is used for standardisation in the individual chapters and consequently World Standardised incidence Rates (WSR) are reported. These are expressed as the number of new cases per 100,000 person years. In the chapter on international comparisons, both European Standardised incidence Rates (ESR) and WSR are reported (see **chapter 2.4**).
- **Male/Female (M/F) ratios** are calculated by dividing the corresponding age-standardised incidence rates (WSR).

2.3.2 PREVALENCE

Prevalence is the number of persons who are still alive at a given index date, and who received a cancer diagnosis during a specified time period preceding the index date. For example, 5-year prevalence is the number of persons who received at least one new diagnosis of cancer during a specific five-year period and who are still alive at the end of the five-year period. The prevalence data in this publication were estimated with an index date of 31st December 2019, representing people living in Belgium who were diagnosed with at least one sarcoma in the period from 1st January 2015 to 31st December 2019 and who were still alive at the end of 2019 (index date) for 5-year prevalence or from 1st January 2010 to 31st December 2019 for 10-year prevalence. Persons with more than one malignancy were included as prevalent cases in each subtype, but were counted only once in analyses regrouping multiple sarcoma subtypes.

The methodology for results on prevalence was described in detail in our publication ‘Cancer Prevalence in Belgium 2010’⁽¹¹⁾.

2.3.3 INCIDENCE TRENDS

Since data have been collected from 2004 onwards, age-standardised incidence rates (WSR) could also be compared over time. In total, 16 consecutive years of incidence data are available for Belgium. The corresponding incidence trends are shown with the corresponding 95% confidence intervals (95% CI).

Trends in age-standardised incidence (WSR) were quantified by the Annual Percentage Change (APC), which expresses a mean multiplicative change per year. Trends and APC calculations are given by sex, age group and subtype. The APC is estimated from a least squares regression on the logarithm of the age-standardised rate (WSR) versus incidence year. Due to the log transformation, no APC can be obtained if the WSR is zero for at least one year. In cases where the relation of the WSR with incidence year cannot be adequately fit with a loglinear model (i.e. a constant APC for the full data range cannot be assumed), a piecewise log-linear model was estimated in which the different linear segments are connected at certain joinpoints. This model results in an estimated APC per time segment of which an Average Annual Percentage Change (AAPC) is calculated as the average of the APC estimates per segment weighted by the corresponding segment length⁽²⁰⁾. The model building process on the logarithm of the WSR was fully automated in SAS (version 9.3) and consists of the following steps:

1. The simple linear regression model, assuming a normal error structure, was compared with a nonparametric smoother fit (PROC REG and PROC LOESS respectively) using an F-test on the residual sets for both models. When the linear regression model was not significantly different from the smoother at the 5% level, the linear model was accepted as final model and a single APC value resulted to quantify the trend over the full time range.
2. When the linear model at the log scale was rejected, a piecewise model with one joinpoint was fitted. The optimal position of the joinpoint was determined using a non-linear optimisation procedure (PROC NLIN). Joinpoints were not allowed to be the first or second time point or the before last and last time point, as those endpoints can be influential points and induce spurious segments. The estimated joinpoint position was rounded to the nearest integer value and fixed in a re-estimation of the piecewise model with PROC GENMOD. As in the previous step, an F-test was used to accept or reject the piecewise model against the smoother. When the regression model was accepted, the final model consisted of a piecewise model with two connected linear segments each quantified by their own APC and a weighted overall AAPC.
3. When the piecewise model with one joinpoint was not accepted, the process continues to evaluate two joinpoints in the same way as described in step 2. As an additional restriction, the difference in position between the two joinpoints should be at least three years. If the two joinpoints were closer, the piecewise model with only one joinpoint from the previous step was retained. A 95% confidence interval (CI) and p-value for the individual segments and the overall AAPC were calculated from the final regression model. The loss in degrees of freedom due to the optimisation of the joinpoint position(s) was not taken into account for the construction of the CI and final p-values. When the 95% CI for the AAPC contains the value zero, no significant trend with incidence year is observed. Combined changes in trends of incidence, mortality and survival can have various causes and are often difficult to interpret and are not considered as an objective of this publication. However, a manuscript by Karim-Kos et al. on trends of cancer in Europe provides an excellent framework to help gaining insights and provide possible explanations for the observed trends⁽²¹⁾.

2.3.4 INCIDENCE PROJECTIONS

The incidence projections for the period 2020-2030 were obtained from linear and log-linear Poisson regression models by extrapolating the observed incidence trends for the period 2004-2019. As the observed number of cancer diagnoses represent a counting process, Poisson models were used to model the relation between the crude incidence rate and the incidence year. The population size at the start of the calendar year was taken as the (log-) offset in the Poisson rate models and the number of observed cancer diagnoses as dependent variable. The modelling process consisted of 2 main steps. First a log-linear Poisson model was estimated. If a significant slope at the 5% level was obtained, the estimated log-linear Poisson model was selected as final model in case of a decreasing time trend (this to avoid projections that end up with a negative number of cancer cases) while a new linear Poisson model was estimated in case of an increasing time trend (to avoid exponential extrapolation). When the slope coefficient of the initial log-linear Poisson model was found to be non-significant, the mean yearly crude rate was estimated over the available time period.

Evolutions in the population size and age distribution were taken into account using the projections of potential population growth as published by Statistics Belgium. Gender specific incidence projections were performed per 5-year age category (0-4, 5-9, ..., 80-84, 85+) to obtain projected sex and age specific crude rates. These projected rates were then applied to the projected population to obtain age-sex specific projected incidence counts. Finally, these age-sex cancer incidence counts were summed and overall projected numbers of cancer diagnoses and crude incidence rates were obtained. Age-standardised rates (WSR) were directly calculated based on the age-sex specific projected cancer incidence rates. All projections were performed using SAS software version 9.3 (SAS Institute, Cary, NC, USA), p-values below 0.05 were considered statistically significant.

A more detailed description of the methodology can be found in our publication 'Cancer Incidence Projections in Belgium'⁽¹⁴⁾.

2.3.5 RELATIVE SURVIVAL

The relative survival gives an estimate of the net survival, which is the survival when causes of death not related to the cancer have been eliminated. The relative survival is obtained by comparing the observed survival with the expected survival for a comparable group of the general population matched for age, sex, region and calendar period. The expected survival was obtained with the Ederer II method for **chapters 3, 3.1, 3.2**⁽²²⁾. Both Ederer II and Pohar-Perme⁽²³⁾ methods were used for the chapter with international comparisons (**chapter 4**) (methods detailed in **chapter 2.4**).

In this publication, 5-year and 10-year relative survival ratios are reported stratified by age group, sex and type of sarcoma. For the survival analyses with results for all ages together, cases with age younger than 15 years were included, this in contrast to most other reports from the BCR. In addition, the chapters also contain results for the age-specific 5-year relative survival showing more detailed data by age group (including cases below 15 years of age). The methodology was described in detail in our publication 'Cancer Survival in Belgium'⁽⁹⁾.

The empirical life tables (by sex, age, region and calendar-year), used in the calculation for expected survival, vary considerably by year of age for young (<30 years) and old ages (>90 years)⁽⁷⁾. To reduce the sampling variability and to ensure that death probabilities evolve consistently from one age and calendar year to another, the life tables were smoothed on age and calendar year using the LOESS-method⁽²⁴⁻²⁷⁾.

In this publication, relative survival results are not shown when the number of patients at risk is less than 50 cases and all relative survival results are presented with the corresponding 95% confidence intervals (95% CI). After a check of the consistency of survival data, for rhabdoid tumours, a lower N at risk was exceptionally allowed due to the rarity of the disease (see **chapter 3.2.7.3**).

2.3.6 CONDITIONAL RELATIVE SURVIVAL

The conditional relative survival reported in this publication is the relative survival proportion given that the person has already survived the first X years since diagnosis (results are shown for X = 1, 2 and 3 years). It is calculated as the standard relative survival, although only patients who survived the first X years since diagnosis are considered. So in case of X = 1, the reported 5-year conditional relative survival therefore corresponds with the relative survival 6 years after diagnosis for patients that at least survived the first year since diagnosis.

2.3.7 RELATIVE SURVIVAL TRENDS

Relative survival is compared between the cohorts 2004-2009, 2009-2014 and 2014-2019. Overlapping periods are reported to obtain sufficiently large cohorts, especially in case of rare tumour types. Note that the follow-up period for the cohorts is not the same, as the last date of follow-up is the 1st of April 2021. In case sarcoma subtypes with low incidence, only two periods were compared.

2.4 INTERNATIONAL COMPARISON

In this report we also aim to compare the Belgian epidemiological situation (namely, age-standardized incidence and relative survival) with the most recent available international data. We included three main studies which enables us to make a 3-level comparison of Belgium with:

- (i) Europe and the United States of America (data from the RARECARENet study ⁽²⁸⁾), both sexes and over the period 2000-2007;
- (ii) with different parts of Europe (data of the RARECARENet study ⁽²⁹⁾), both sexes and over the same period 2000-2007 and
- (iii) with 2 neighbouring countries (the Netherlands (supplemental data provided by the Netherlands Comprehensive Cancer Organisation (IKNL)) and France ^(16,30-32) and supplemental data provided by the FRANCIM network), by sex and over a more recent period 2010-2015.

We would like to thank the RARECARENet Study for the availability of their data on <http://rarecarenet.istitutotumori.mi.it/index.php>, the FRANCIM network for the collection of supplemental data and advice, and the registration team of the Netherlands Comprehensive Cancer Organisation (IKNL) for providing supplemental data of the Netherlands Cancer Registry.

The Belgian data are harmonized with those from other countries to cope with the wide heterogeneity of the available data on sarcomas regarding classification, inclusion criteria, incidence and survival indicators. Consequently, the classification in the **chapter 4** is different (**table 4**) from the one used throughout the rest of the publication (**table 1**).

Cancer data on a national level for Belgium are only available since 2004. Prior to 2004, only for the Flemish region incidence and survival data are available with complete coverage. Therefore, for the period 2000-2007, we used data from Flanders only (comparison with RARECARENet study) as indicated in the respective figures.

The European regions used in the RARECARENet study are defined as follows:

- Northern Europe: Finland, Iceland, Norway
- United Kingdom and Ireland: England, Northern Ireland, Republic of Ireland, Scotland, Wales
- Central Europe: Austria, Belgium (Flanders), France, Germany, Switzerland, the Netherlands
- Eastern Europe: Bulgaria, Czech Republic, Estonia, Latvia, Lithuania, Poland, Slovakia
- Southern Europe: Croatia, Italy, Malta, Portugal, Slovenia, Spain

The first part of this international comparison focuses on incidence based on age-standardised incidence rates (using 1976 European Standard population or World Standard population). Two time periods (2000-2007 and 2010-2013) and two different groupings (**table 4**) are presented in **chapter 4**.

In the second part concerning survival data, relative survival (Ederer II or Pohar-Perme method) are presented depending on the methodology described in the respective publications ⁽²⁸⁻³²⁾. Data presented in **figure 7 and table 3 of chapter 4** only include patients aged 15 years or older at time of diagnosis.

Selections of ICD-O-3 topographical and morphological codes used to establish sarcoma subgroups are detailed in **table 4**.

Table 4 Classifications used for international comparison

Sarcomas classified by localisation and/or histological type	Topography classification (ICD-O-3)	Morphology classification (ICD-O-3)
Classification based on RARECARENet²⁸ used in Figure 1-4;7-8 and Table 1;3 of chapter 4		
Soft tissue sarcoma	C00.0-C39.9; C42.0-C80.9	8710-8711; 8800-8935; 8959; 8963-8964; 8990-8991; 9020; 9040-9044; 9120-9133; 9150; 9170; 9180; 9231; 9240; 9251; 9260; 9364-9365; 9540; 9560-9571; 9580-9581
	C00.0-C06.9; C09.0-C39.9; C42.0-C43.9; C45.0-C59.9; 8940 C61.0-C63.1; C63.3-C80.9	
	C00.0-C39.9; C42.0-C55.9; C57.0-C61.9; C63.0-C70.9; 9473 C73.0-C80.9	
Soft tissue sarcoma of head and neck	C00.0-C14.8; C30.0-C32.9; C49.0; C73.9; C75.2; C75.4; C76.0	8710-8711; 8800-8902; 8912; 8921; 8933-8935; 8963; 8990-8991; 9040-9044; 9120-9133; 9150; 9170; 9180; 9231; 9240; 9364-9365; 9540; 9560-9571; 9580-9581
Soft tissue sarcoma of limbs	C49.1-C49.2; C76.4-C76.5	8710-8711; 8800-8902; 8912; 8921; 8933-8935; 8940; 8963; 8990-8991; 9040-9044; 9120-9133; 9150; 9170; 9180; 9231; 9240; 9365; 9540; 9560-9571; 9580-9581
Soft tissue sarcoma of superficial trunk	C49.3-C49.4; C49.6; C76.1-C76.2; C76.7	8710-8711; 8800-8902; 8912; 8921; 8933-8935; 8940; 8963; 8990-8991; 9040-9044; 9120-9133; 9150; 9170; 9180; 9231; 9240; 9365; 9540; 9560-9571; 9580-9581
Soft tissue sarcoma of mediastinum	C38.1-C38.3; C38.8	8710-8711; 8800-8902; 8912; 8921; 8933-8935; 8940; 8963; 8990-8991; 9040-9044; 9120-9133; 9150; 9170; 9180; 9231; 9240; 9365; 9540; 9560-9571; 9580-9581
Soft tissue sarcoma of heart	C38.0	8710-8711; 8800-8902; 8912; 8921; 8933-8935; 8940; 8963; 8990-8991; 9040-9044; 9120-9133; 9150; 9170; 9180; 9231; 9240; 9365; 9540; 9560-9571; 9580-9581
Soft tissue sarcoma of breast	C50.0-C50.9	8710-8711; 8800-8902; 8912; 8921; 8933-8935; 8940; 8963; 8990-8991; 9020; 9040-9044; 9120-9133; 9150; 9170; 9180; 9231; 9240; 9365; 9540; 9560-9571; 9580-9581
Soft tissue sarcoma of uterus	C53.0-C55.9	8710-8711; 8800-8902; 8912; 8921-8931; 8933; 8934-8935; 8940; 8963; 8990-8991; 9040-9044; 9120-9133; 9150; 9170; 9180; 9231; 9240; 9365; 9540; 9560-9571; 9580-9581
Soft tissue sarcoma of other genitourinary tract (vulva; vagina; ovary; penis; prostate; testis; kidney; renal pelvis; ureter; bladder; urethra)	C51.0-C52.9; C56.9-C57.9; C60.0-C62.9; C63.8-C63.9; C64.9-C68.9	8710-8711; 8800-8902; 8912; 8921; 8933; 8934-8935; 8959; 8963-8964; 8990-8991; 9040-9044; 9120-9133; 9150; 9170; 9180; 9231; 9240; 9365; 9540; 9560-9571; 9580-9581
	C51.0-C52.9; C56.9-C57.9; C61.9-C62.9; C63.8-C63.9; C64.9-C68.9	8940
Soft tissue sarcoma of viscera	C15.0-C26.9; C33.9-C37.9; C38.4; C39.0-C39.9; C422	8710-8711; 8800-8902; 8912; 8921; 8933-8935; 8940; 8963; 8990-8991; 9040-9044; 9120-9133; 9150; 9170; 9180; 9231; 9240; 9365; 9540; 9560-9571; 9580-9581
Soft tissue sarcoma of paratestis	C63.0-C63.7	8710-8711; 8800-8902; 8912; 8921; 8933-8935; 8940; 8963; 8990-8991; 9040-9044; 9120-9133; 9150; 9170; 9180; 9231; 9240; 9365; 9540; 9560-9571; 9580-9581
Soft tissue sarcoma of retroperitoneum and peritoneum	C48.0-C48.8	8710-8711; 8800-8902; 8912; 8921; 8933-8935; 8940; 8963; 8990-8991; 9040-9044; 9120-9133; 9150; 9170; 9180; 9231; 9240; 9365; 9540; 9560-9571; 9580-9581
Soft tissue sarcoma of pelvis	C49.5; C76.3	8710-8711; 8800-8902; 8912; 8921; 8933-8935; 8940; 8963; 8990-8991; 9040-9044; 9120-9133; 9150; 9170; 9180; 9231; 9240; 9365; 9540; 9560-9571; 9580-9581
Soft tissue sarcoma of skin	C44.0-C44.9	8710-8711; 8800-8902; 8912; 8921; 8933-8935; 8963; 8990-8991; 9040-9044; 9120-9133; 9150; 9170; 9180; 9231; 9240; 9365; 9540; 9560-9571; 9580-9581
Soft tissue sarcoma of paraorbit	C69.0-C69.9	8710-8711; 8800-8902; 8912; 8921; 8933-8935; 8940; 8963; 8990-8991; 9040-9044; 9120-9133; 9150; 9170; 9180; 9231; 9240; 9365; 9540; 9560-9571; 9580-9581
Soft tissue sarcoma of brain and other parts of the nervous system	C47.0-C47.9; C70.0-C72.9; C75.1; C75.3	8710-8711; 8800-8902; 8912; 8921; 8933-8935; 8940; 8963; 8990-8991; 9040-9044; 9120-9133; 9150; 9170; 9180; 9231; 9240; 9365; 9540; 9560-9571; 9580-9581
Embryonal rhabdomyosarcoma of soft tissue	C00.0-C39.9; C42.0-C80.9	8910
Alveolar rhabdomyosarcoma of soft tissue	C00.0-C39.9; C42.0-C55.9; C57.0-C61.9; C63.0-C80.9	8920
Ewing sarcoma of soft tissue	C00.0-C39.9; C42.0-C55.9; C57.0-C61.9; C63.0-C70.9; 9260; 9364; 9473 C73.0-C80.9	
Bone sarcoma	C40.0-C41.9	8800-8801; 8803-8806; 8810-8812; 8815; 8830; 8840; 8850-8855; 8890-8891; 8894-8896; 8900-8902; 8910; 8912; 8920; 9040-9044; 9120-9133; 9150; 9170; 9180-9250; 9260-9261; 9310; 9364; 9370-9372; 9540-9581
Osteogenic sarcoma	C40.0-C41.9	9180; 9183; 9185; 9187; 9192-9194
Chondrogenic sarcoma	C40.0-C41.9	9220; 9240; 9242-9243
Notochordal sarcoma; chordoma	C40.0-C41.9	9370-9372
Vascular sarcoma	C40.0-C41.9	9120-9133; 9150; 9170
Ewing sarcoma	C40.0-C41.9	9260; 9364
Other high grade sarcomas (fibrosarcoma; malignant fibrous histiocytoma)	C40.0-C41.9	8810; 8830
Gastrointestinal stromal tumours	All cancer sites	8936
Kaposi sarcoma	All cancer sites	9140

Sarcomas classified by localisation and/or histological type	Topography classification (ICD-O-3)	Morphology classification (ICD-O-3)
Classification based on Amadeo <i>et al.</i>; 2020¹⁶ used in Figure 5-6 and Table 2 of chapter 4		
Gastrointestinal stromal tumours	All cancer sites	8936*
Liposarcoma	All cancer sites	8850; 8852; 8853; 8854; 8855; 8857; 8858
Chondrosarcoma	All cancer sites	9220; 9221; 9231; 9240; 9242; 9243
Dermatofibrosarcoma	All cancer sites	8832; 8833
Kaposi sarcoma	All cancer sites	9140
Angiosarcoma	All cancer sites	9120; 9124; 9170
Osteosarcoma	All cancer sites	9180; 9181; 9182; 9183; 9184; 9185; 9186; 9187; 9192; 9193; 9194; 9195
Ewing sarcoma	All cancer sites	9260; 9364
Myxofibrosarcoma	All cancer sites	8811
Rhabdomyosarcoma	All cancer sites	8900; 8901; 8902; 8910; 8912; 8920; 8921
Nerve sheath tumours	All cancer sites	9540; 9542; 9560; 9561; 9571; 9580
Endometrial stromal sarcoma	All cancer sites	8930; 8931; 8935
Synovial tumours	All cancer sites	9040; 9041; 9042; 9043
Chordoma	All cancer sites	9370; 9371; 9372
Solitary fibrous tumour	All cancer sites	8815
Fibrosarcoma	All cancer sites	8810; 8812; 8813; 8814
Classification based on Francim national reports²⁹⁻³¹ used in Figure 9 and Table 4 of chapter 4		
All sarcomas	All cancer sites	8710; 8711; 8714; 8800; 8801; 8802; 8803; 8804; 8805; 8806; 8810; 8811; 8812; 8813; 8814; 8815; 8825; 8830; 8832; 8833; 8840; 8842; 8850; 8851; 8852; 8853; 8854; 8855; 8857; 8858; 8890; 8891; 8894; 8895; 8896; 8900; 8901; 8902; 8910; 8912; 8920; 8921; 8930; 8931; 8932; 8933; 8935; 8936; 8940; 8963; 8964; 8973; 8982; 8990; 8991; 9040; 9041; 9042; 9043; 9044; 9045; 9120; 9130; 9133; 9137; 9140; 9150; 9170; 9180; 9181; 9182; 9183; 9184; 9185; 9186; 9187; 9192; 9193; 9194; 9195; 9220; 9221; 9230; 9231; 9240; 9242; 9243; 9250; 9251; 9252; 9260; 9261; 9364; 9365; 9370; 9371; 9372; 9473; 9508; 9540; 9542; 9560; 9561; 9571; 9580; 9581
Soft tissue sarcoma	C00-C06; C09-C14; C30-C32; C38.1-C38.3; C47-C49; C69.8	8710; 8711; 8714; 8800; 8801; 8802; 8803; 8804; 8805; 8806; 8810; 8811; 8812; 8813; 8814; 8815; 8825; 8830; 8832; 8833; 8840; 8842; 8850; 8851; 8852; 8853; 8854; 8855; 8857; 8858; 8890; 8891; 8894; 8895; 8896; 8900; 8901; 8902; 8910; 8912; 8920; 8921; 8930; 8931; 8932; 8933; 8935; 8936; 8940; 8963; 8964; 8973; 8982; 8990; 8991; 9040; 9041; 9042; 9043; 9044; 9045; 9120; 9130; 9133; 9137; 9140; 9150; 9170; 9180; 9181; 9182; 9183; 9184; 9185; 9186; 9187; 9192; 9193; 9194; 9195; 9220; 9221; 9230; 9231; 9240; 9242; 9243; 9250; 9251; 9252; 9260; 9261; 9364; 9365; 9370; 9371; 9372; 9473; 9508; 9540; 9542; 9560; 9561; 9571; 9580; 9581
Gastrointestinal stromal tumour	All cancer sites	8936*
Bone sarcoma	C40.0-C41.9	8710; 8711; 8714; 8800; 8801; 8802; 8803; 8804; 8805; 8806; 8810; 8811; 8812; 8813; 8814; 8815; 8825; 8830; 8832; 8833; 8840; 8842; 8850; 8851; 8852; 8853; 8854; 8855; 8857; 8858; 8890; 8891; 8894; 8895; 8896; 8900; 8901; 8902; 8910; 8912; 8920; 8921; 8930; 8931; 8932; 8933; 8935; 8936; 8940; 8963; 8964; 8973; 8982; 8990; 8991; 9040; 9041; 9042; 9043; 9044; 9045; 9120; 9130; 9133; 9137; 9140; 9150; 9170; 9180; 9181; 9182; 9183; 9184; 9185; 9186; 9187; 9192; 9193; 9194; 9195; 9220; 9221; 9230; 9231; 9240; 9242; 9243; 9250; 9251; 9252; 9260; 9261; 9364; 9365; 9370; 9371; 9372; 9473; 9508; 9540; 9542; 9560; 9561; 9571; 9580; 9581
Classification based on Botta <i>et al.</i>, 2020²⁷ used in Figure 10 and Table 5 of chapter 3.3		
Soft tissue sarcoma of limbs	C49.1-C49.2; C76.4-C76.5	8710; 8711; 8714; 8800; 8801; 8802; 8803; 8804; 8805; 8806; 8810; 8811; 8812; 8813; 8814; 8815; 8825; 8830; 8832; 8833; 8840; 8842; 8850; 8851; 8852; 8853; 8854; 8855; 8857; 8858; 8890; 8891; 8894; 8895; 8896; 8900; 8901; 8902; 8910; 8912; 8920; 8921; 8930; 8931; 8932; 8933; 8935; 8936; 8940; 8963; 8990; 8991; 9040; 9041; 9042; 9043; 9044; 9120; 9124; 9130; 9133; 9137; 9150; 9170; 9180; 9181; 9182; 9183; 9185; 9186; 9187; 9192; 9193; 9194; 9195; 9220; 9231; 9240; 9252; 9365; 9540; 9542; 9560; 9561; 9571; 9580; 9581
Soft tissue sarcoma of superficial trunk	C49.3-C49.4; C49.6; C76.1-C76.2; C76.7	8710; 8711; 8714; 8800; 8801; 8802; 8803; 8804; 8805; 8806; 8810; 8811; 8812; 8813; 8814; 8815; 8830; 8832; 8833; 8840; 8842; 8850; 8851; 8852; 8853; 8854; 8855; 8857; 8858; 8890; 8891; 8894; 8895; 8896; 8900; 8901; 8902; 8912; 8921; 8933; 8934; 8935; 8940; 8963; 8990; 8991; 9040; 9041; 9042; 9043; 9044; 9120; 9124; 9130; 9133; 9137; 9150; 9170; 9180; 9181; 9182; 9183; 9185; 9186; 9187; 9192; 9193; 9194; 9195; 9220; 9231; 9240; 9252; 9365; 9540; 9542; 9560; 9561; 9571; 9580; 9581; 9220
Soft tissue sarcoma of viscera	C15.0-C26.9; C34.1-C37.9; C38.4; C39.0-C39.9; C42.2	8710; 8711; 8714; 8800; 8801; 8802; 8803; 8804; 8805; 8806; 8810; 8811; 8812; 8813; 8814; 8815; 8830; 8832; 8833; 8840; 8842; 8850; 8851; 8852; 8853; 8854; 8855; 8857; 8858; 8890; 8891; 8894; 8895; 8896; 8900; 8901; 8902; 8912; 8921; 8933; 8934; 8935; 8940; 8963; 8990; 8991; 9040; 9041; 9042; 9043; 9044; 9120; 9124; 9130; 9133; 9137; 9150; 9170; 9180; 9181; 9182; 9183; 9185; 9186; 9187; 9192; 9193; 9194; 9195; 9220; 9231; 9240; 9252; 9365; 9540; 9542; 9560; 9561; 9571; 9580; 9581

Sarcomas classified by localisation and/or histological type	Topography classification (ICD-O-3)	Morphology classification (ICD-O-3)
Soft tissue sarcoma of retroperitoneum and peritoneum	C48.0-C48.8	8710; 8711; 8714; 8800; 8801; 8802; 8803; 8804; 8805; 8806; 8810; 8811; 8812; 8813; 8814; 8815; 8830; 8832; 8833; 8840; 8842; 8850; 8851; 8852; 8853; 8854; 8855; 8857; 8858; 8890; 8891; 8894; 8895; 8896; 8900; 8901; 8902; 8912; 8921; 8933; 8934; 8935; 8940; 8963; 8990; 8991; 9040; 9041; 9042; 9043; 9044; 9120; 9124; 9130; 9133; 9137; 9150; 9170; 9180; 9181; 9182; 9183; 9185; 9186; 9187; 9192; 9193; 9194; 9195; 9220; 9231; 9240; 9252; 9365; 9540; 9542; 9560; 9561; 9571; 9580; 9581
Soft tissue sarcoma of pelvis	C49.5; C76.3	8710; 8711; 8714; 8800; 8801; 8802; 8803; 8804; 8805; 8806; 8810; 8811; 8812; 8813; 8814; 8815; 8830; 8832; 8833; 8840; 8842; 8850; 8851; 8852; 8853; 8854; 8855; 8857; 8858; 8890; 8891; 8894; 8895; 8896; 8900; 8901; 8902; 8912; 8921; 8933; 8934; 8935; 8940; 8963; 8990; 8991; 9040; 9041; 9042; 9043; 9044; 9120; 9124; 9130; 9133; 9137; 9150; 9170; 9180; 9181; 9182; 9183; 9185; 9186; 9187; 9192; 9193; 9194; 9195; 9220; 9231; 9240; 9252; 9365; 9540; 9542; 9560; 9561; 9571; 9580; 9581
Ewing sarcoma of soft tissue	All cancer sites	All cancers sites except C40.0-C41.9, C32.3, C33.9, C34.0, C30.0, C30.1: 9260; 9364; All cancers sites except C40.0-C41.9, C32.3, C33.9, C34.0, C30.0, C30.1, C56, C71, C72: 9473
Osteogenic sarcoma	C40.0-C41.9; C32.3; C33.9; C34.0; C30.0; C30.1	9180; 9181; 9182; 9183; 9185; 9186; 9187; 9192; 9193; 9194; 9195
Chondrogenic sarcoma	C40.0-C41.9; C32.3; C33.9; C34.0; C30.0; C30.1	9220; 9221; 9230; 9231; 9240; 9242; 9243
Notochordal sarcoma, chordoma	All cancer sites	9370; 9371; 9372
Kaposi sarcoma	All cancer sites	9140

Only malignant neoplasms were included, excepted for gastrointestinal stromal tumour indicated by an asterisk for which both malignant and borderline neoplasms were included

Source: Belgian Cancer Registry 

¹⁶ Amadéo B; Penel N; Coindre JM; Ray-Coquard I; Ligier K et al. Incidence and time trends of sarcoma (2000–2013): results from the French network of cancer registries (FRANCLIM). *BMC Cancer*. 2020; 6,20(1):190

²⁷ Botto L, Gatta G, Trama A, Bernasconi A, Sharon E, Capocaccia R, Mariotto A, the RARECAREnet working group. Incidence and survival of rare cancers in the US and Europe. *Cancer Medicine* 2020;9:5632-5642

²⁸ « RARECARENet – On line Analysis ». Information Network on Rare Cancers. RARECARENet. <http://rarecarenet.istitutotumori.mi.it/analysis.php>. 29 Sept. 2021

²⁹ Désandes E; Amadéo B; Delafosse P; Lecoffre C; Lafay L; Mounier M et al. *Survie des personnes atteintes de cancer en France métropolitaine 1989-2018 – Sarcomes*. Boulogne-Billancourt : Institut national du cancer mars 2021

³⁰ Amadéo B; Désandes E; Delafosse P; Lecoffre C; Lafay L; Mounier M et al. *Survie des personnes atteintes de cancer en France métropolitaine 1989-2018 – Sarcomes des tissus mous*. Boulogne-Billancourt : Institut national du cancer mars 2021

³¹ Seigneurin A; Plouvier S; Désandes E; Amadéo B; Delafosse P; Lecoffre C et al. *Survie des personnes atteintes de cancer en France métropolitaine 1989-2018 – Sarcomes des os*. Boulogne-Billancourt : Institut national du cancer mars 2021

3 ALL SARCOMAS

MAIN STRUCTURE:

- Sarcomas classified by their primary tumour location (chapter 3.1)
- Sarcomas classified by histological type (chapter 3.2)

KEYNOTES

Incidence (table 1-2; figure 1-5)

- Sarcomas are rare tumours and make up 1.7% of all cancer diagnoses. However, the percentage of sarcomas in children (0-19 years) is higher and reaches 13% of all cancer diagnoses in children.
- Sarcoma incidence increases with age, with the highest incidence in the oldest age groups.
- Between 2004 and 2019 the incidence of sarcomas in Belgium is stable in younger age groups but shows an increasing trend in the older age groups (60+ years), especially in males. This increase is mainly observed in gastrointestinal stromal tumours (GIST, chapter 3.2.8) and liposarcoma (in males, chapter 3.2.1). Data for GIST also include so-called “micro-GIST” (tumours smaller than 1 cm in diameter). There is an increased awareness among pathologists and increased use of biomarkers (KIT, DOG1, mutational analysis) to identify GIST. Doctors become more confident that these tumours can be treated in the older age groups, whereas in the past most 80+ people were not deemed fit for treatment.

Survival (table 3; figure 6-8)

- The 5-year relative survival is very similar between males (73%) and females (74%).
- Survival of all sarcoma patients combined decreases with age (with differences according to histological type of sarcoma - see chapter 3.2).
- There is a trend indicating a potential improvement in the 5-year relative survival over time from 69% in 2004-2009 to 74% to 2014-2019.

Table 1 All sarcomas: Overview of incidence, prevalence and survival by sex in Belgium, 2010-2019

Males				
Incidence	N	CR	WSR	
Incidence, 2010-2019	5,749	10.5	6.8	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	2,280	40.3	24.9	
10-year prevalence, 31.12.2019	3,697	65.3	40.5	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	5,675	72.9	[71.3;74.5]	
10-year relative survival, 2010-2019	5,675	67.7	[65.0;70.4]	
Females				
Incidence	N	CR	WSR	
Incidence, 2010-2019	5,389	9.5	6.0	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	2,105	36.1	22.3	
10-year prevalence, 31.12.2019	3,634	62.3	38.0	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	5,335	74.1	[72.5;75.6]	
10-year relative survival, 2010-2019	5,335	69.0	[66.5;71.4]	
Median age at diagnosis, 2010-2019 (y)	63 [Q1: 49; Q3: 75]			
M/F-ratio	1.1			

Source: Belgian Cancer Registry 

N: number of new diagnoses

CR: crude (all ages) rate (N/100,000 person years)

WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

M/F-ratio: Male/Female ratio based on the age-standardised rates (2010-2019)

Incidence

Figure 1 All cancers: Incidence by type of malignancy and age category, Belgium, 2010-2019

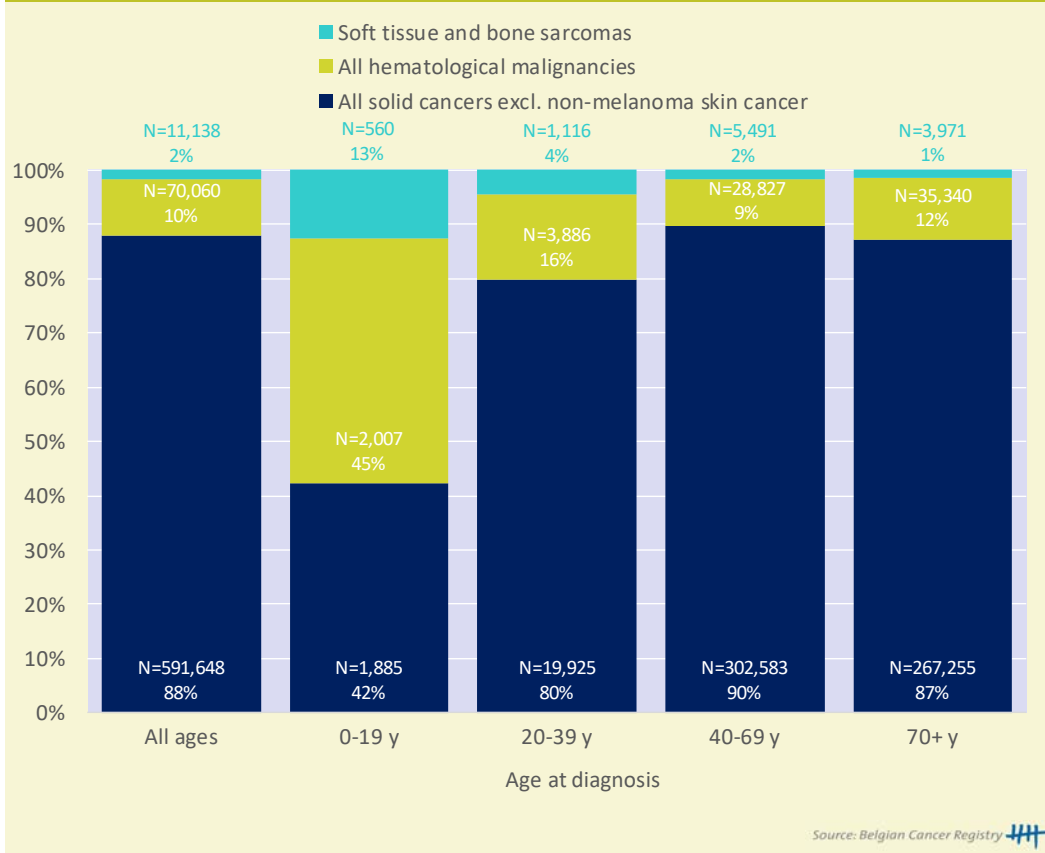
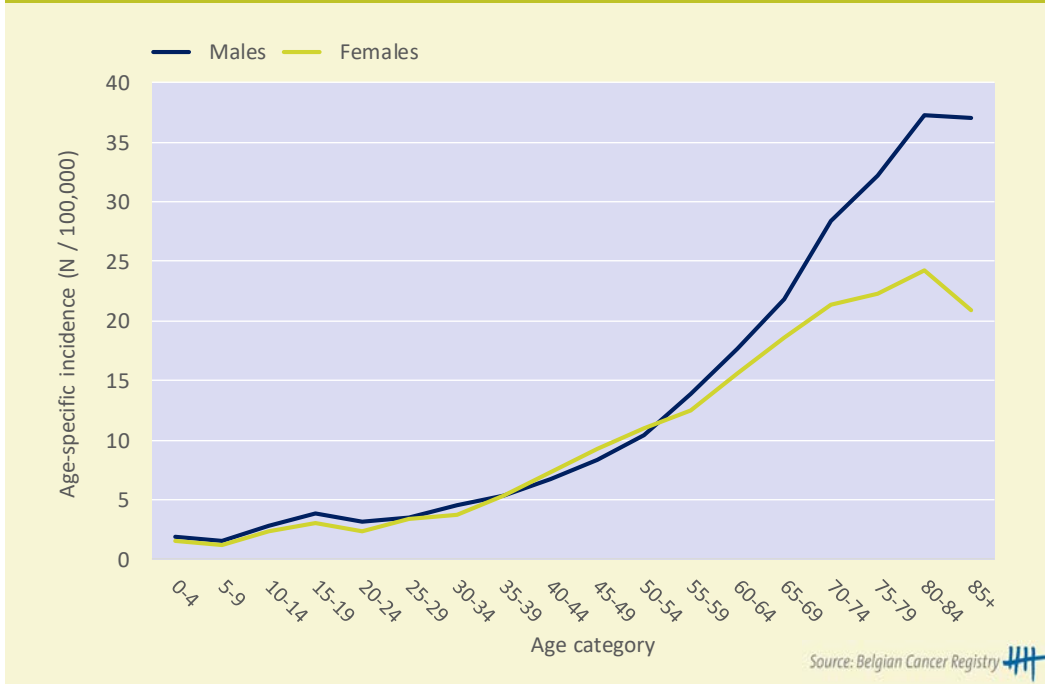
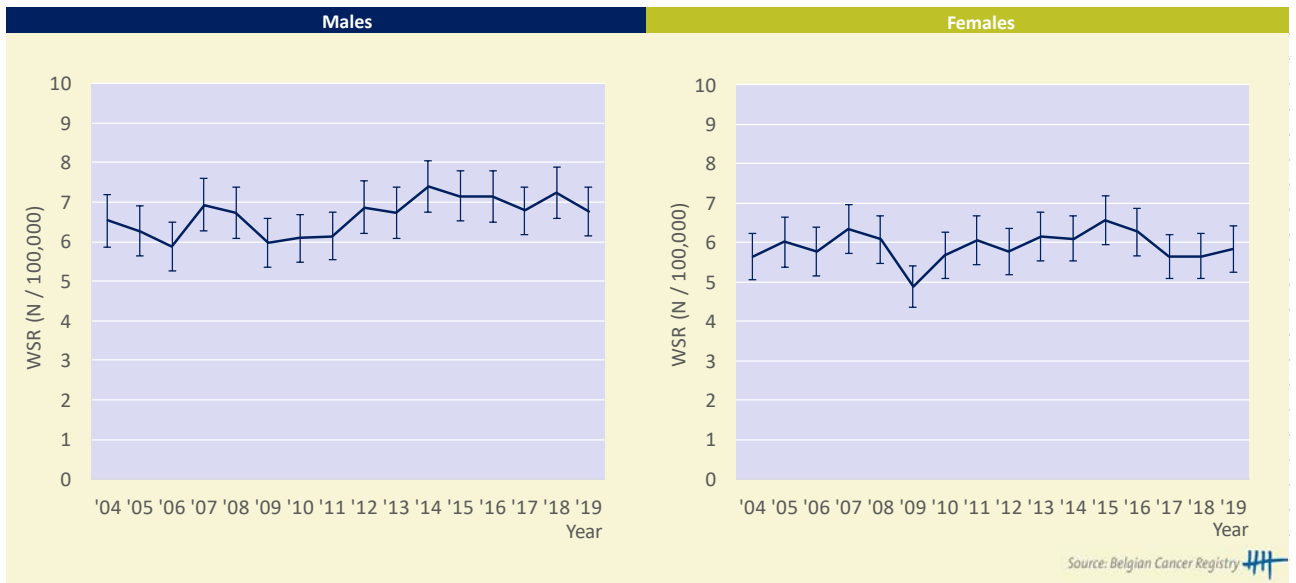


Figure 2 All sarcomas: Age-specific incidence rates (N/100,000) by sex, Belgium 2004-2019



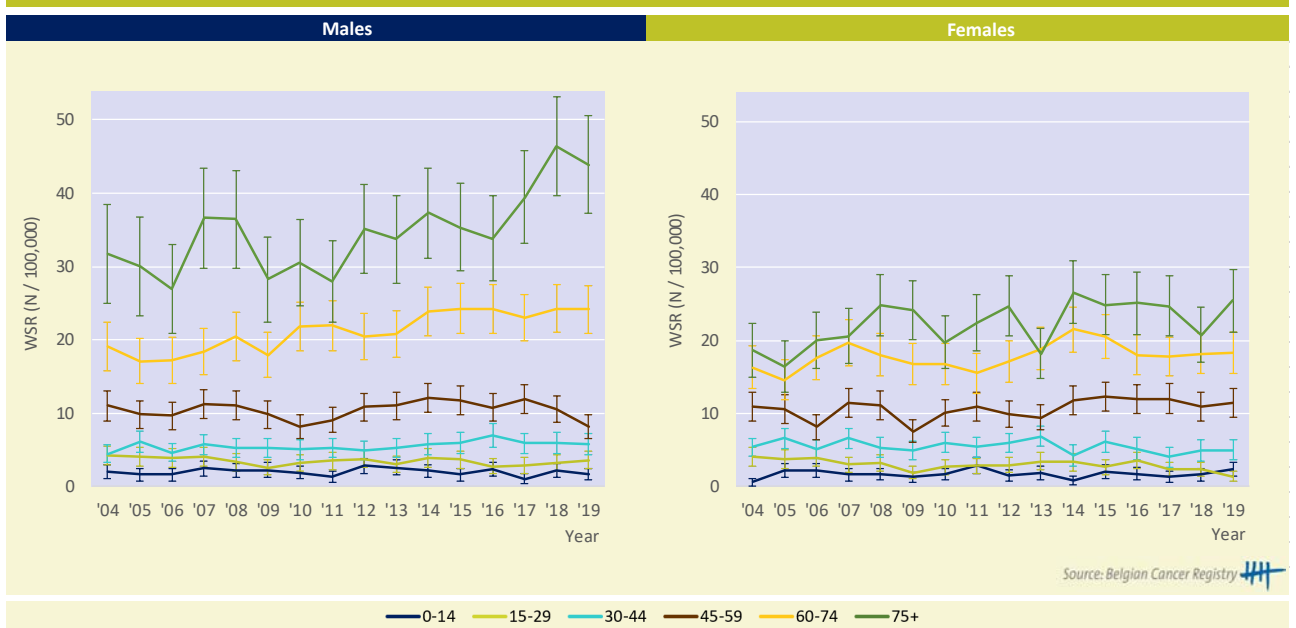
Incidence trends

Figure 3 All sarcomas: Age-standardised incidence rates* (WSR) by sex, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals.

Figure 4 All sarcomas: Age-standardised incidence rates* (WSR) by sex and age category, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals.

Table 2 All sarcomas: Incidence trends by sex and age category in Belgium, 2004-2019

Age category	Males			Females		
	AAPC (%)	95%CI	Period	AAPC (%)	95%CI	Period
All ages	0.9	[0.2; 1.6]	2004-2019	0.2	[-0.6; 1.0]	2004-2019
0 - 14 y	-0.9	[-3.9; 2.2]	2004-2019	1.7	[-2.8; 6.4]	2004-2019
15 - 29 y	-1.4	[-3.0; 0.2]	2004-2019	-6.1	[-8.3; -3.9]	2004-2019
				-11.9	[-18.0; -5.2]	2004-2009
				9.1	[2.3; 16.4]	2009-2014
				-13.9	[-19.9; -7.4]	2014-2019
30 - 44 y	1.3	[0.2; 2.5]	2004-2019	-1.3	[-3.0; 0.3]	2004-2019
45 - 59 y	-1.1	[-2.5; 0.2]	2004-2019	1.2	[-0.3; 2.8]	2004-2019
	-3.3	[-6.6; 0.2]	2004-2010			
	8.2	[3.1; 13.5]	2010-2014			
	-5.5	[-9.6; -1.2]	2014-2019			
60 - 74 y	2.4	[1.7; 3.1]	2004-2019	0.4	[-0.9; 1.6]	2004-2019
				0.0	[-3.1; 3.3]	2004-2010
				4.5	[-0.0; 9.1]	2010-2014
				-2.4	[-6.2; 1.6]	2014-2019
75+ y	2.3	[1.0; 3.7]	2004-2019	1.8	[0.4; 3.3]	2004-2019

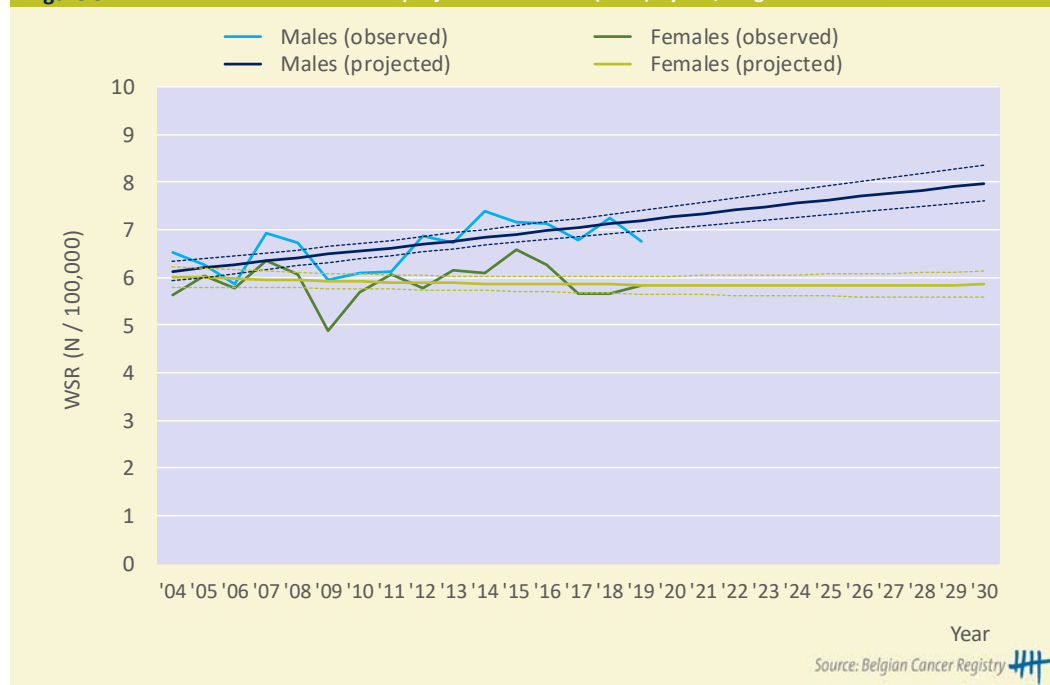
Source: Belgian Cancer Registry 

AAPC: average annual percentage change

Period: when a joinpoint occurred, APC's are calculated for the period before and after the joinpoint. This column represents the corresponding time interval. AAPC's are always calculated over the entire study-period.

Incidence projections

Figure 5 All sarcomas: Observed and projected incidence (WSR) by sex, Belgium 2004-2030

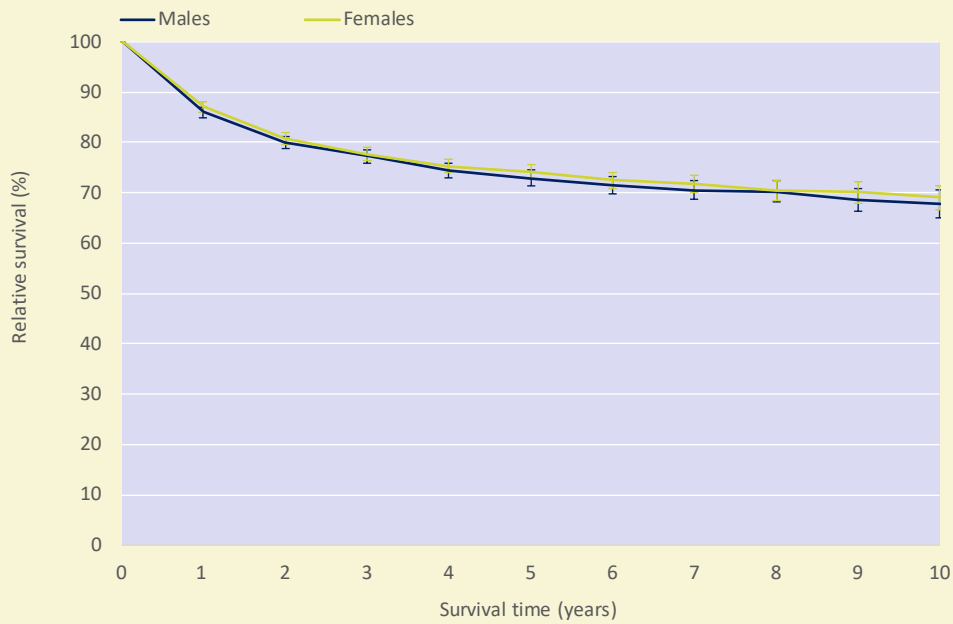


Source: Belgian Cancer Registry 

WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

Survival

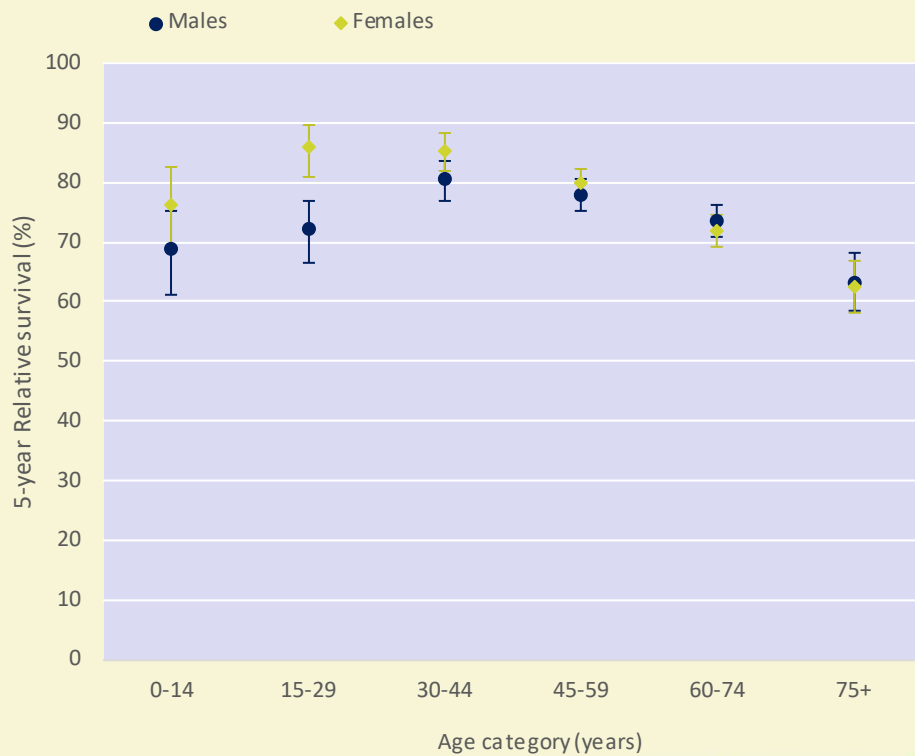
Figure 6 All sarcomas: Relative survival* by sex, Belgium 2010-2019



Source: Belgian Cancer Registry

* The relative survival values are represented with 95% Confidence Intervals

Figure 7 All sarcomas: 5-year relative survival* by age category and sex, Belgium 2010-2019



Source: Belgian Cancer Registry

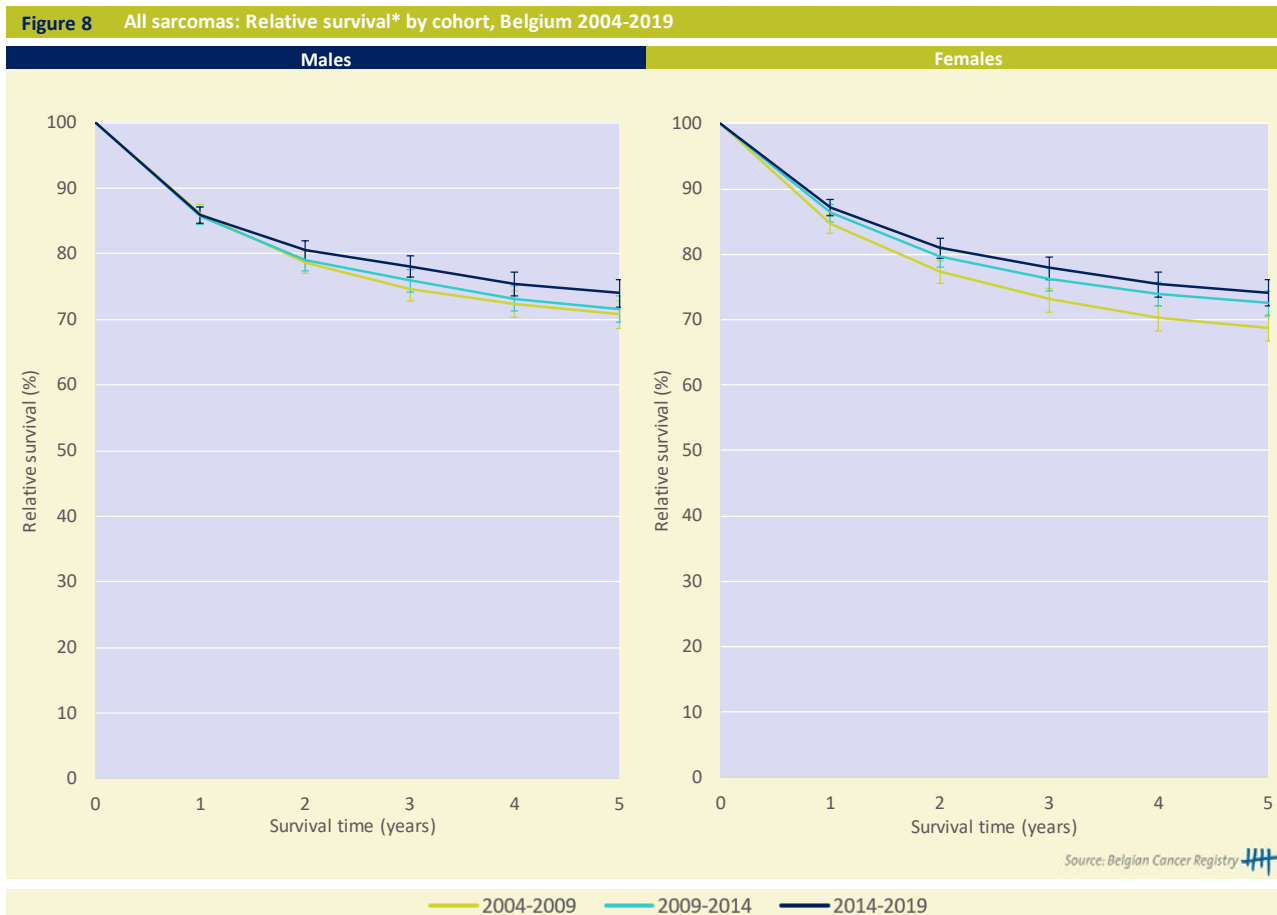
* The relative survival values are represented with 95% Confidence Intervals

Table 3 All sarcomas: Conditional 5-year relative survival* by sex in Belgium, 2010-2019		
Males		
X years since diagnosis	N at risk	%
1 year	4,728	83.2
2 year	3,914	88.2
3 year	3,211	91.0
Females		
X years since diagnosis	N at risk	%
1 year	4,531	83.3
2 year	3,798	88.8
3 year	3,173	90.8

* Unadjusted 5-year relative survival probability conditional on surviving the first X years since diagnosis, %

* Interpretation in lay-man's terms: Given that a patient has already survived X years, what is the relative survival probability 5 years later.

Survival trends



* The relative survival values are represented with 95% Confidence Intervals.

3.1 SARCOMAS CLASSIFIED BY PRIMARY TUMOUR LOCATION

MAIN SUBTYPES:

- *Soft tissue and visceral sarcoma (& related tumours)*
- *Bone sarcoma (& related tumours)*

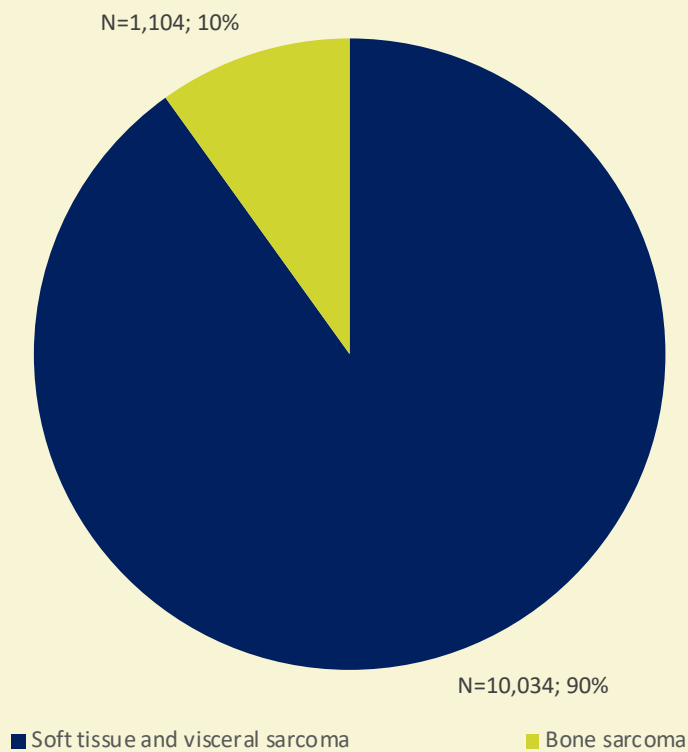
KEYNOTES

Incidence (figure 1-4)

- 90% of sarcomas arise in soft tissues and organs (viscera), whereas 10% originate in bones, joints and articular cartilage.
- The incidence of soft tissue and visceral sarcoma increases strongly with age. For bone sarcoma two smaller peaks are observed, under 20 and above 70 years of age. This results in a higher share of sarcomas originating in bones in younger patients.

Incidence

Figure 1 All sarcomas: Incidence distribution by primary tumour location, Belgium 2010-2019




Source: Belgian Cancer Registry 

Figure 2 All sarcomas: age-specific incidence rates (N/100,000) by primary tumour location, Belgium 2004-2019

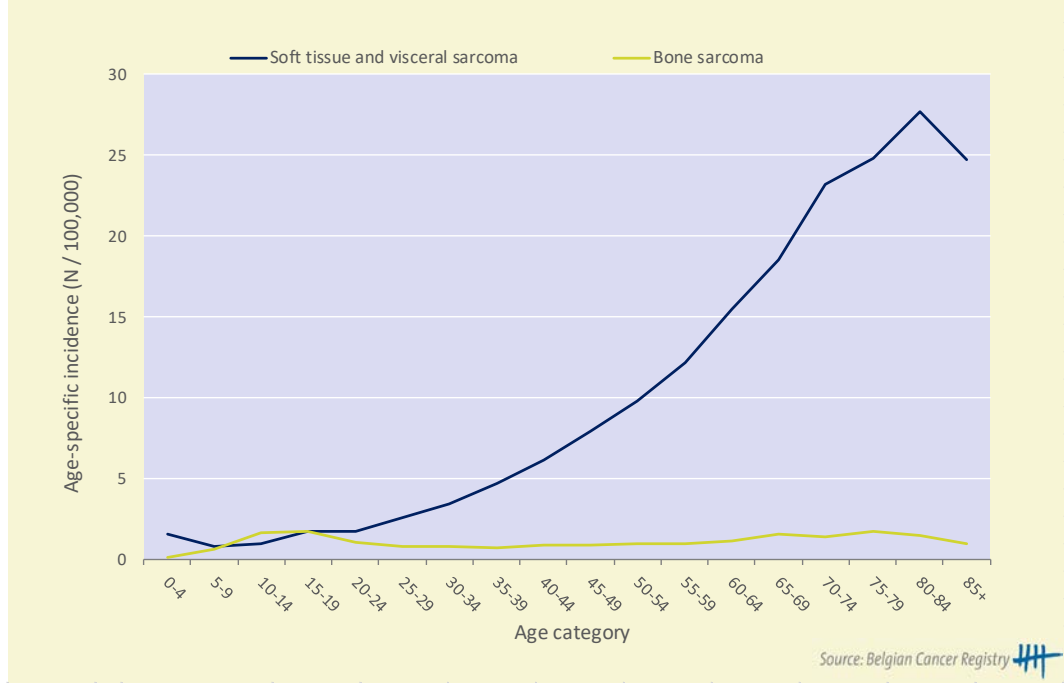
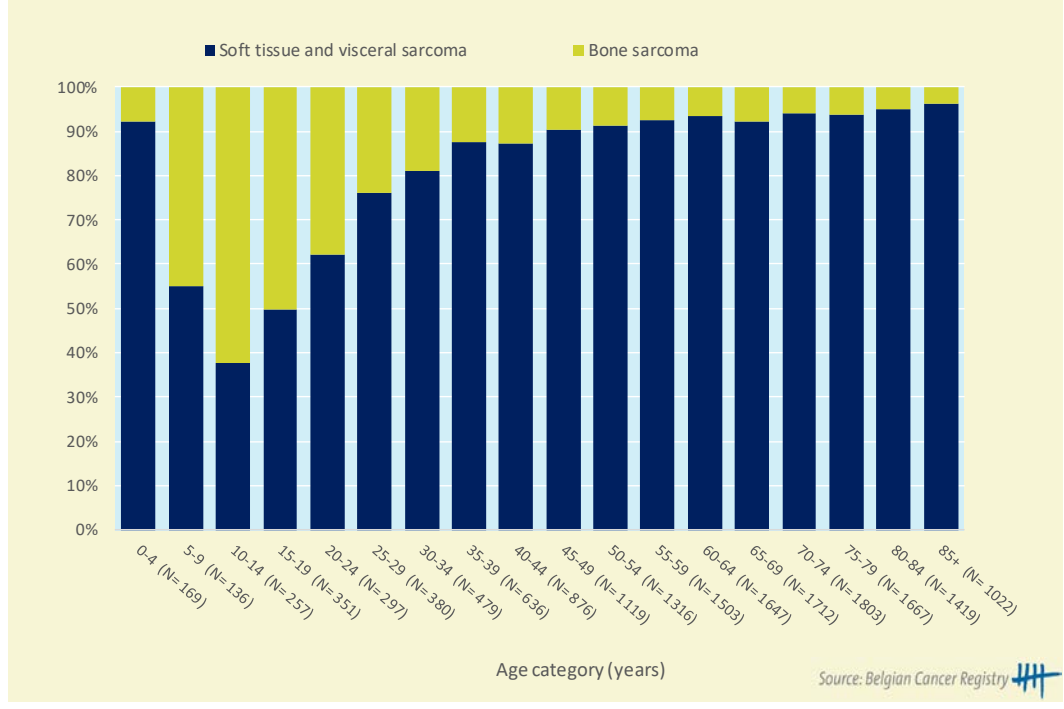


Figure 3 All sarcomas: incidence distribution by age category & primary tumour location, Belgium 2004-2019



3.1.1 SOFT TISSUE AND VISCERAL SARCOMA

KEYNOTES

Incidence (table 1-2; figure 1-6)

- The incidence of soft tissue and visceral sarcoma increases strongly with age and peaks around the age of 80 years.
- Nearly one out of three (30%) sarcomas of the soft tissue and viscera originates in the digestive system, with the stomach being the most often affected digestive organ. The second most frequent group of primary tumour locations is the connective, subcutaneous and other soft tissues (29%) followed by the skin (15%) in third position. The female genital organs, mostly corpus uteri, are ranked fourth (6%).
- The primary tumour location varies with age:
 - Under the age of 20 years, sarcomas most frequently occur in connective, subcutaneous and other soft tissues.
 - The skin is most frequently involved in young adults (20-39 years).
 - In older adults, the digestive system and soft tissues are the predominant tumour locations.
- The incidence increases over time in older age groups (60+ years), especially in males.

Survival (table 3; figure 7-10)

- Males and females with soft tissue and visceral sarcomas have a similar prognosis, with a 5-year relative survival of 74%.
- The 5-year relative survival varies slightly according to age category. Moreover, it seems to improve slightly over time from 70% in 2004-2009 to 75% in 2014-2018.

Soft tissue and visceral sarcoma: Overview of incidence, prevalence and survival by sex in Belgium, 2010-2019				
Males				
Incidence	N	CR	WSR	
Incidence, 2010-2019	5,143	9.4	5.8	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	2,060	36.4	21.4	
10-year prevalence, 31.12.2019	3,311	58.5	34.3	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	5,080	73.8	[72.1;75.4]	
10-year relative survival, 2010-2019	5,080	68.4	[65.3;71.3]	
Females				
Incidence	N	CR	WSR	
Incidence, 2010-2019	4,891	8.6	5.1	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	1,928	33.1	19.5	
10-year prevalence, 31.12.2019	3,265	56.0	32.3	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	4,844	73.5	[71.9;75.1]	
10-year relative survival, 2010-2019	4,844	68.1	[65.4;70.8]	
Median age at diagnosis, 2010-2019 (y)	64 [Q1: 51; Q3: 75]			
M/F-ratio	1.1			

Source: Belgian Cancer Registry 

N: number of new diagnoses

CR: crude (all ages) rate (N/100,000 person years)

WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

M/F-ratio: Male/Female ratio based on the age-standardised rates (2010-2019)

Incidence

Figure 1 Soft tissue and visceral sarcoma: Age-specific incidence rates (N/100,000) by sex, Belgium 2004-2019

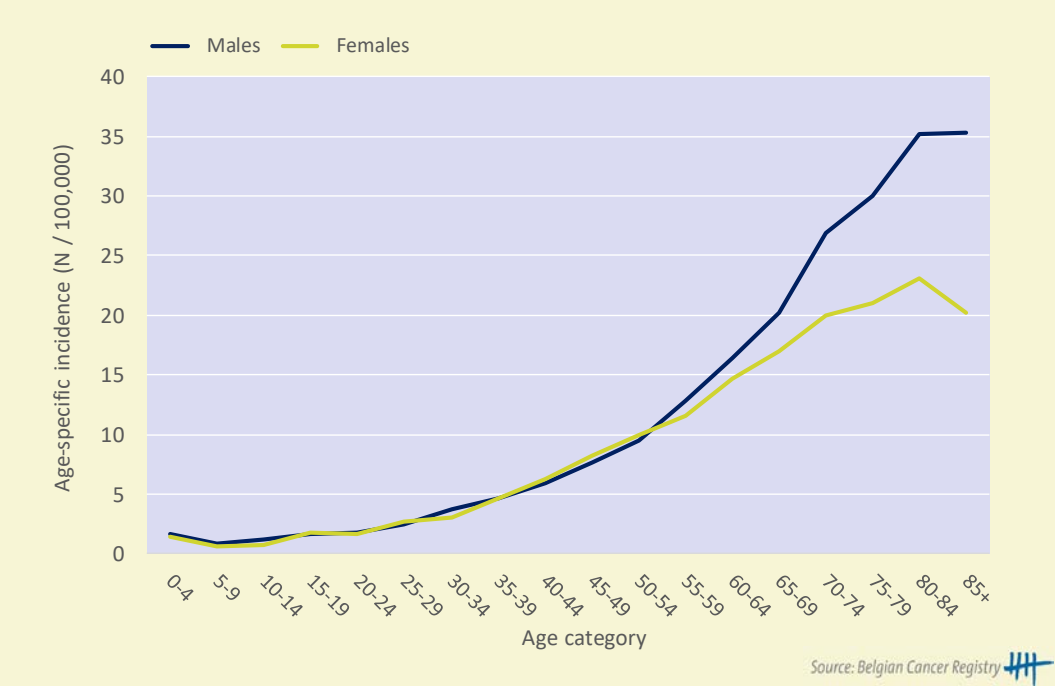
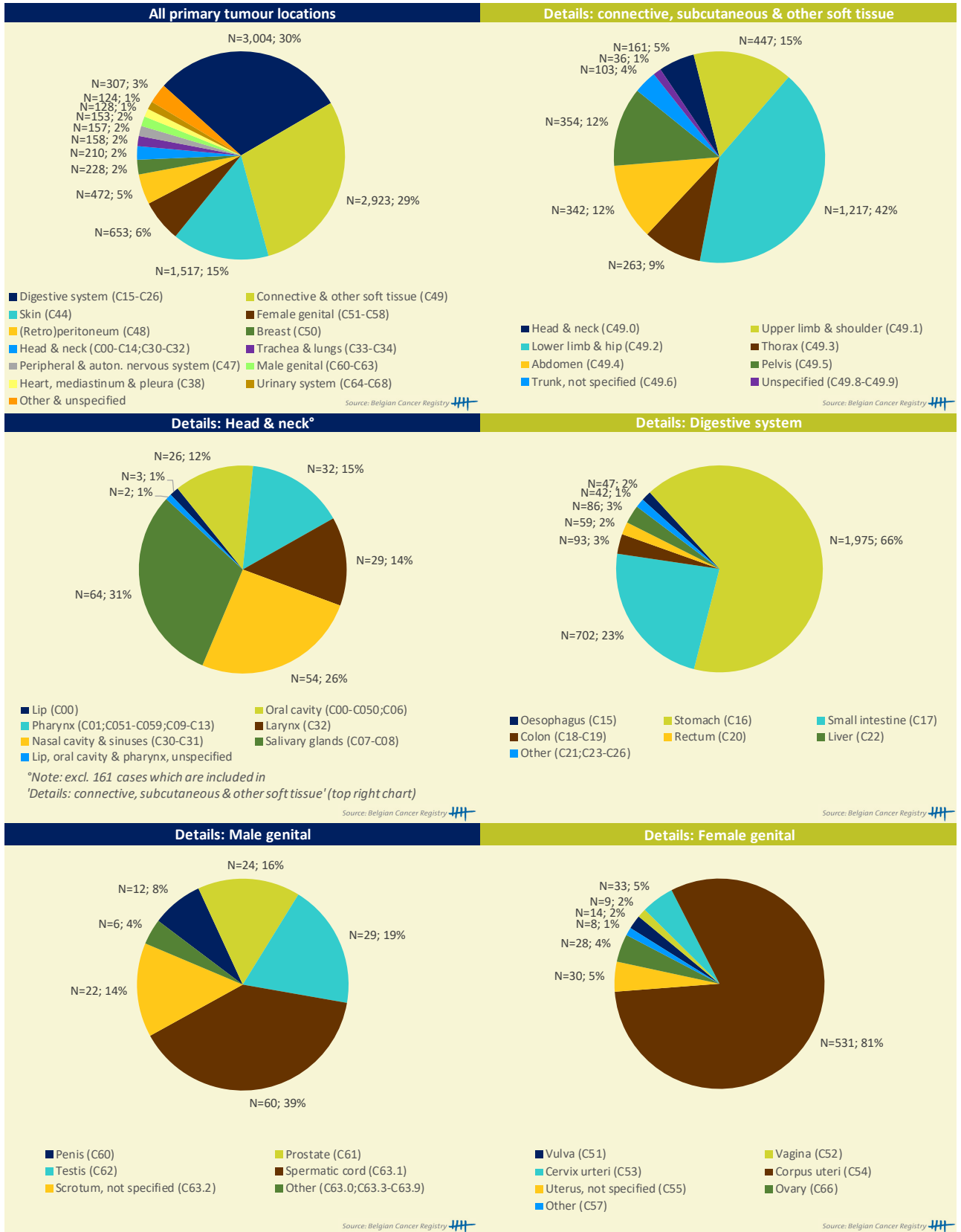
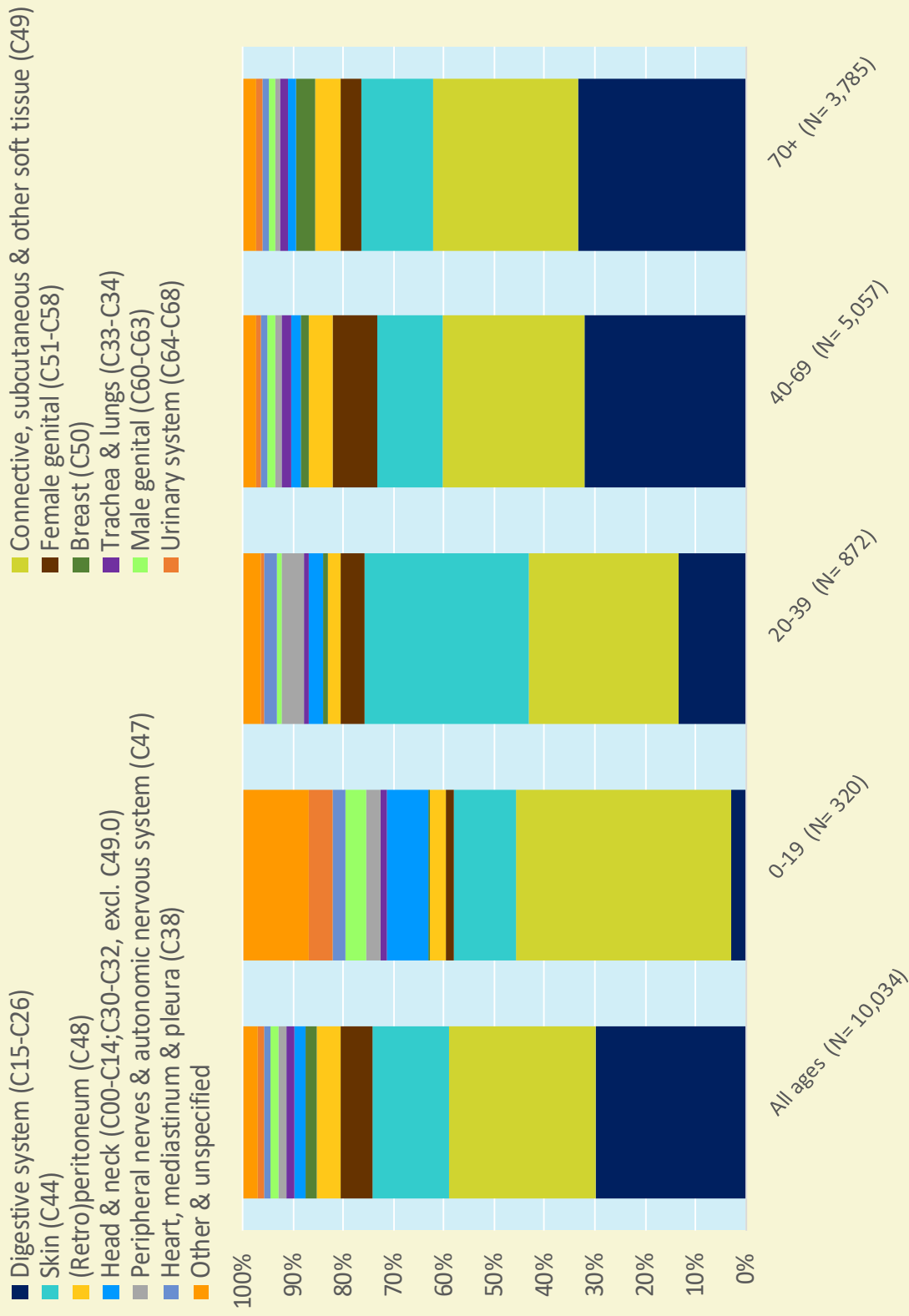


Figure 2 Soft tissue and visceral sarcoma: Incidence distribution by primary tumour location*, Belgium 2010-2019



* Tumour location defined by ICD-O-3 topography codes⁽³⁾.

Figure 3 Soft tissue and visceral sarcoma: Incidence distribution by primary tumour location and age category, Belgium 2010-2019



Source: Belgian Cancer Registry

* Tumour location defined by ICD-O-3 topography codes⁽³⁾.

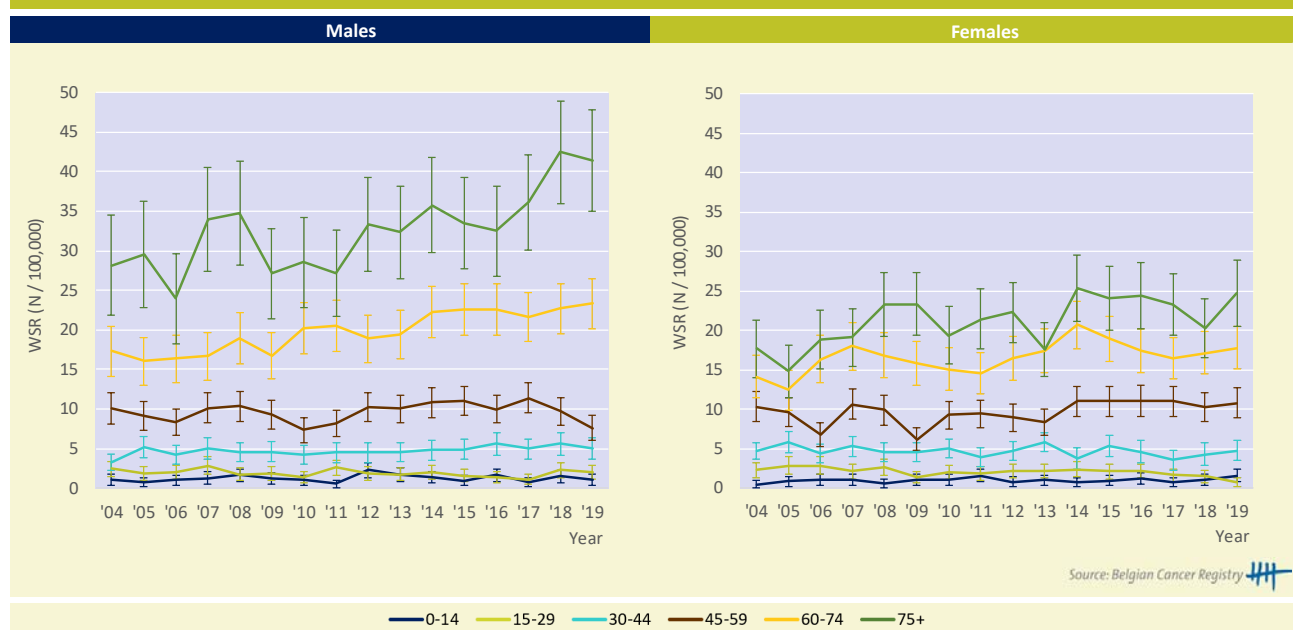
Incidence trends

Figure 4 Soft tissue and visceral sarcoma: Age-standardised incidence rates* (WSR) by sex, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals.

Figure 5 Soft tissue and visceral sarcoma: Age-standardised incidence rates* (WSR) by sex and age category, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals.

Table 2 Soft tissue and visceral sarcoma: Incidence trends by sex and age category in Belgium, 2004-2019

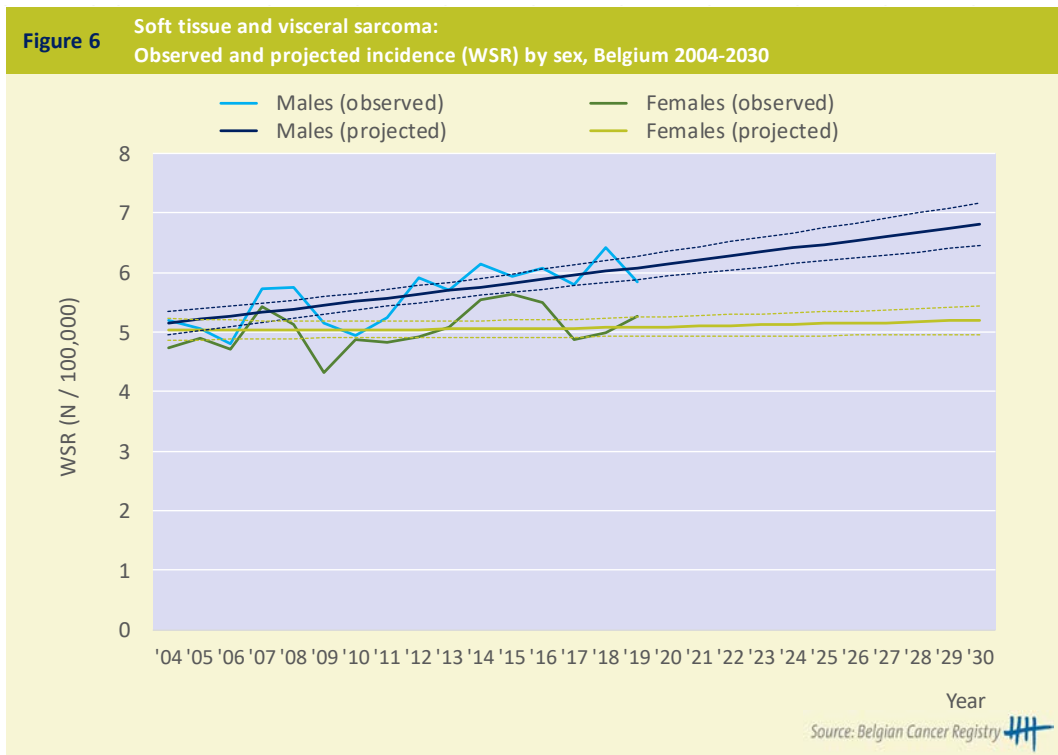
Age category	Males			Females		
	AAPC (%)	95%CI	Period	AAPC (%)	95%CI	Period
All ages	1.3	[0.6; 2.1]	2004-2019	0.6	[-0.1; 1.4]	2004-2019
0 - 14 y	1.3	[-3.0; 5.8]	2004-2019	2.8	[-0.8; 6.6]	2004-2019
15 - 29 y	-1.9	[-4.5; 0.7]	2004-2019	-6.7	[-9.3; -4.1]	2004-2019
				-5.2	[-10.6; 0.5]	2004-2011
				8.7	[-1.4; 19.9]	2011-2015
				-22.2	[-30.7; -12.6]	2015-2019
30 - 44 y	1.6	[0.4; 2.8]	2004-2019	-0.9	[-2.5; 0.7]	2004-2019
45 - 59 y	-0.9	[-2.4; 0.6]	2004-2019	1.5	[-0.3; 3.5]	2004-2019
				-3.0	[-6.9; 0.9]	2004-2010
				8.2	[2.4; 14.3]	2010-2014
				-5.2	[-9.8; -0.3]	2014-2019
60 - 74 y	2.6	[1.9; 3.2]	2004-2019	0.9	[-0.5; 2.3]	2004-2019
				1.3	[-1.7; 4.4]	2004-2011
				4.3	[-0.8; 9.8]	2011-2015
				-3.2	[-8.8; 2.9]	2015-2019
75+ y	2.4	[1.2; 3.7]	2004-2019	2.1	[0.7; 3.5]	2004-2019

Source: Belgian Cancer Registry 

AAPC: average annual percentage change

Period: When a joinpoint occurred, APC's are calculated for the period before and after the joinpoint. This column represents the corresponding time interval. AAPC's are always calculated over the entire study-period.

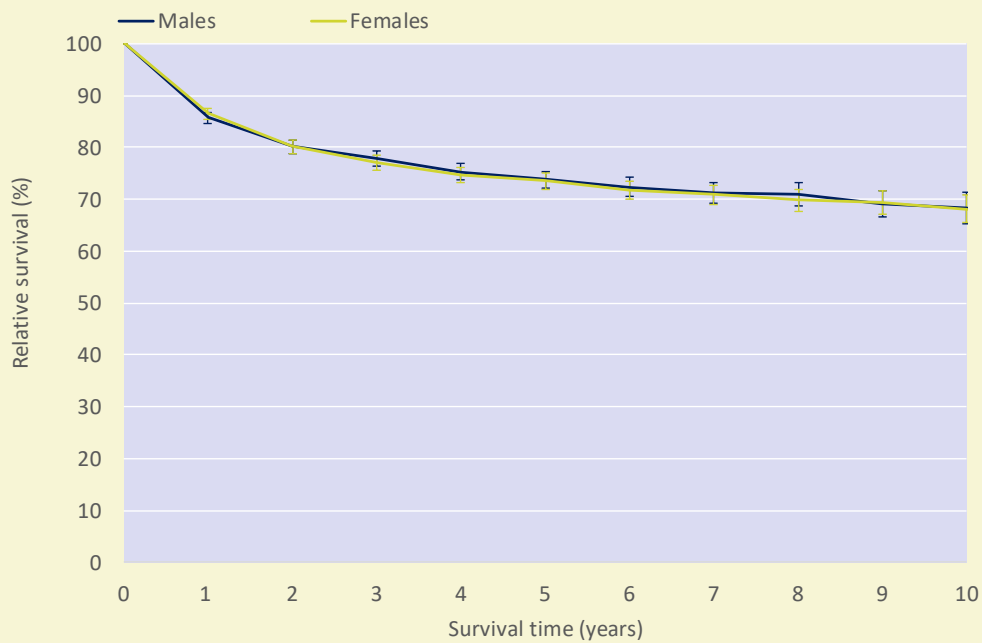
Incidence projections



WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

Survival

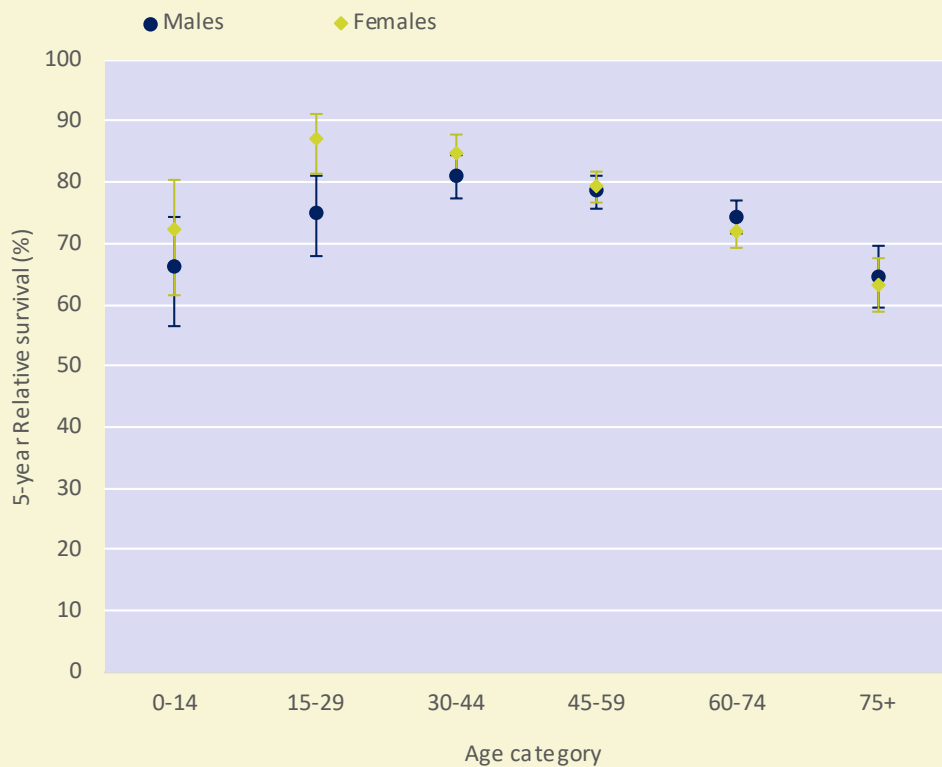
Figure 7 Soft tissue & visceral sarcoma: Relative survival* by sex, Belgium 2010-2019



Source: Belgian Cancer Registry

* The relative survival values are represented with 95% Confidence Intervals

Figure 8 Soft tissue and visceral sarcoma: 5-year relative survival* by age and sex, Belgium 2010-2019



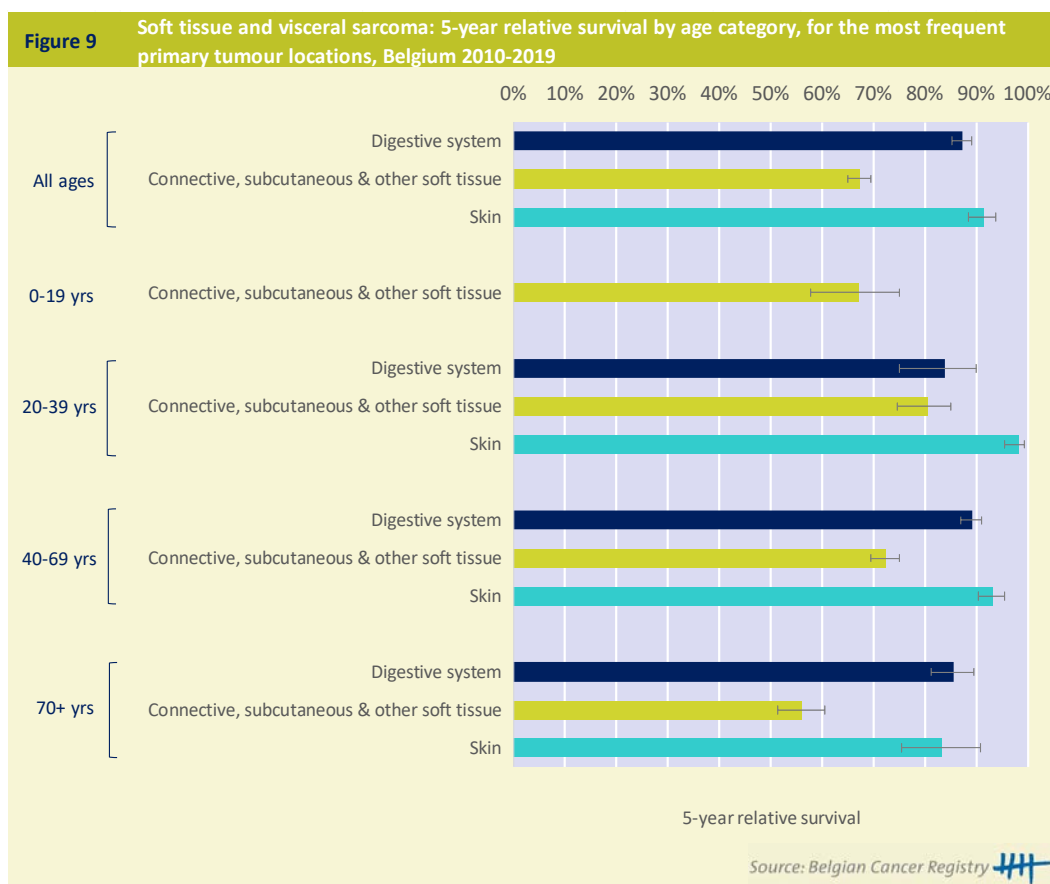
Source: Belgian Cancer Registry

* The relative survival values are represented with 95% Confidence Intervals

Table 3 Soft tissue and visceral sarcoma: Conditional 5-year relative survival* by sex in Belgium, 2010-2019		
Males		
X years since diagnosis	N at risk	%
1 year	4,219	84.5
2 year	3,500	88.9
3 year	2,871	91.2
Females		
X years since diagnosis	N at risk	%
1 year	4,091	82.9
2 year	3,409	88.5
3 year	2,828	90.6

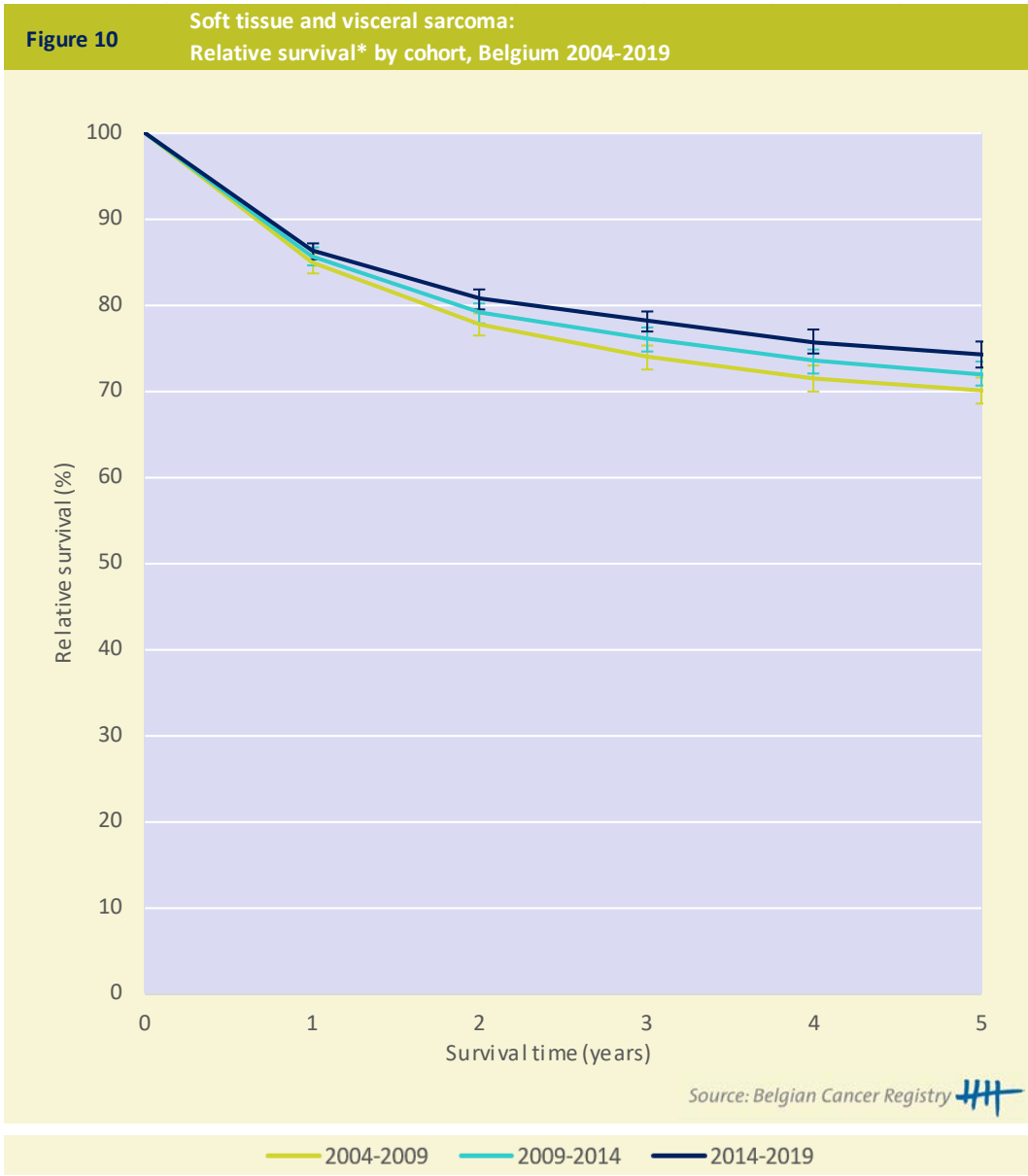
* Unadjusted 5-yr relative survival probability conditional on surviving the first X years since diagnosis, %

* Interpretation in lay-man's terms: Given that a patient has already survived X years, what is the relative survival probability 5 years later.



For 0-19 years, only one primary tumour location is shown because of too small groups for a representative survival analysis. Selection criteria (ICD-O-3): digestive system C15-C26; connective, subcutaneous & other soft tissue C49; skin C44.

Survival trends



* The relative survival values are represented with 95% Confidence Intervals

3.1.2. BONE SARCOMA

KEYNOTES

Incidence (table 1-2; figure 1-5)

- Sarcomas located in bones, joints and articular cartilage show two incidence peaks:
 - In children and adolescents (younger than 20 years).
 - In older patients (65+ years).
- The bones of the limbs represent the most frequent bone tumour location (54%), followed by pelvic bones (17%). In children, adolescents and young adults the percentage of bone tumours in peripheral locations (bones of limbs) is higher (65%) than in older patients (45%).

Survival (table 3; figure 6-9)

- The 5-year relative survival of bone sarcoma patients:
 - Is significantly better in females (79%) than in males (66%). Males present more frequently with Ewing sarcoma and osteosarcoma which have a less favourable prognosis than e.g. chondrosarcoma. Also, within these subtypes, women show a better prognosis.
 - Decreases strongly with age (<50% in the age group 75+ years). Possible explanations include the possibility to give higher chemotherapy doses in younger patients and a higher survival of patients with peripherally located tumours.

Table 1 Bone sarcoma: Overview of incidence, prevalence and survival by sex in Belgium, 2010-2019

Males				
Incidence	N	CR	WSR	
Incidence, 2010-2019	606	1.1	1.0	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	221	3.9	3.5	
10-year prevalence, 31.12.2019	388	6.9	6.2	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	598	66.3	[61.8;70.6]	
10-year relative survival, 2010-2019	598	62.6	[56.3;68.5]	
Females				
Incidence	N	CR	WSR	
Incidence, 2010-2019	498	0.9	0.8	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	178	3.1	2.8	
10-year prevalence, 31.12.2019	371	6.4	5.7	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	494	79.1	[74.7;83.0]	
10-year relative survival, 2010-2019	494	76.1	[70.3;81.2]	
Median age at diagnosis, 2010-2019 (y)	45 [Q1: 22; Q3: 65]			
M/F-ratio	1.2			

Source: Belgian Cancer Registry 

N: number of new diagnoses

CR: crude (all ages) rate (N/100,000 person years)

WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

M/F-ratio: Male/Female ratio based on the age-standardised rates (2010-2019)

Incidence

Figure 1 Bone sarcoma: Age-specific incidence rates (N/100,000) by sex, Belgium 2004-2019

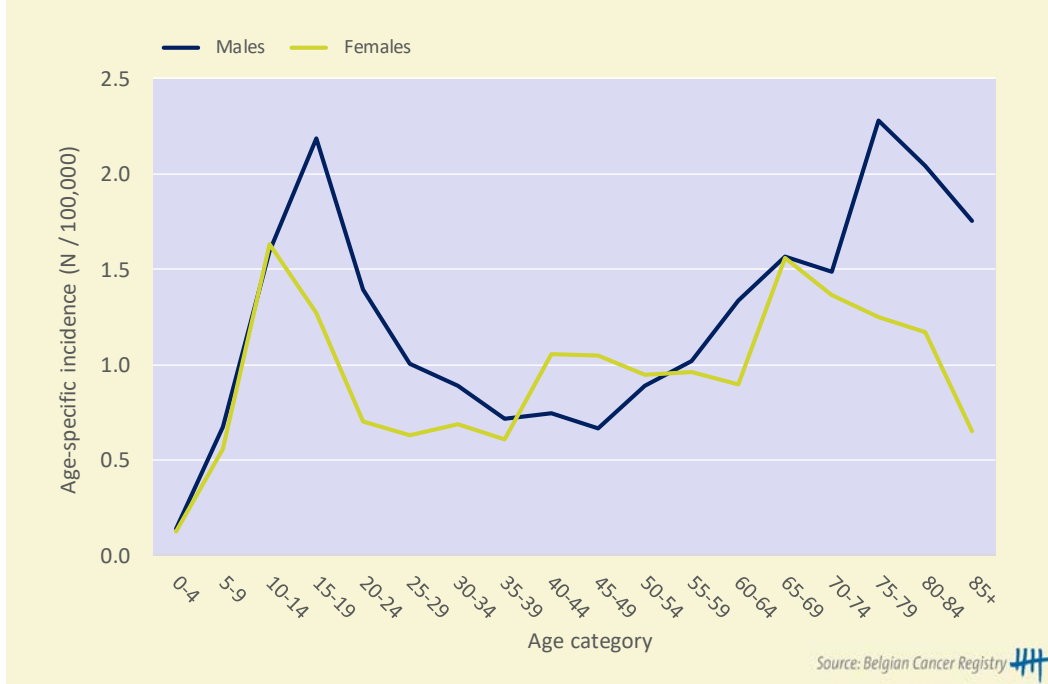
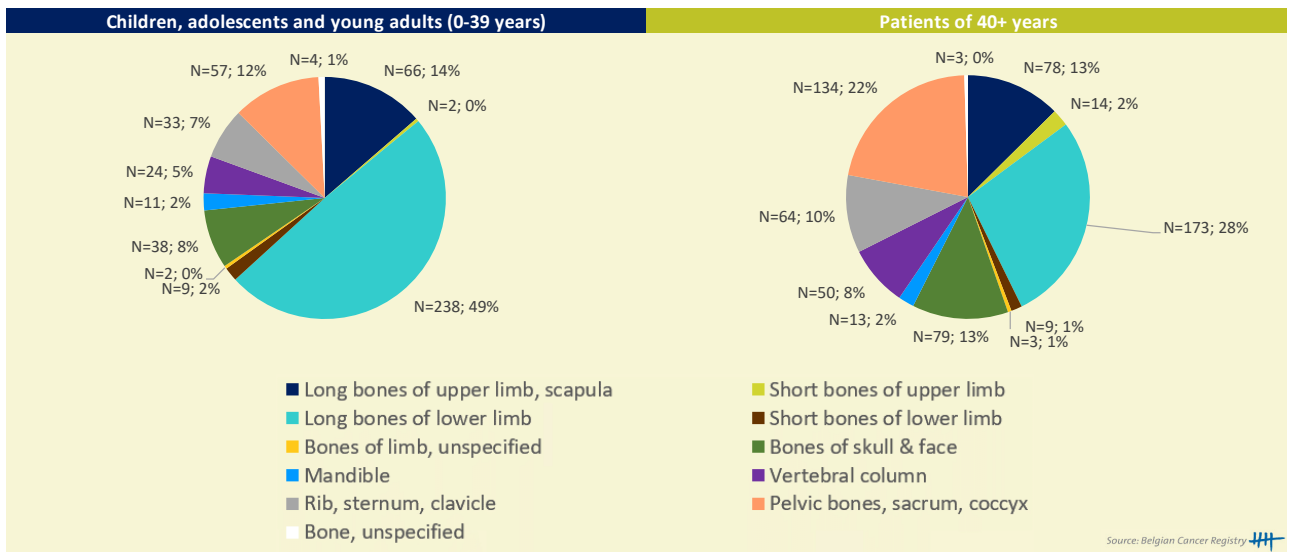


Figure 2 Bone sarcoma: Incidence distribution by primary tumour location, Belgium 2010-2019



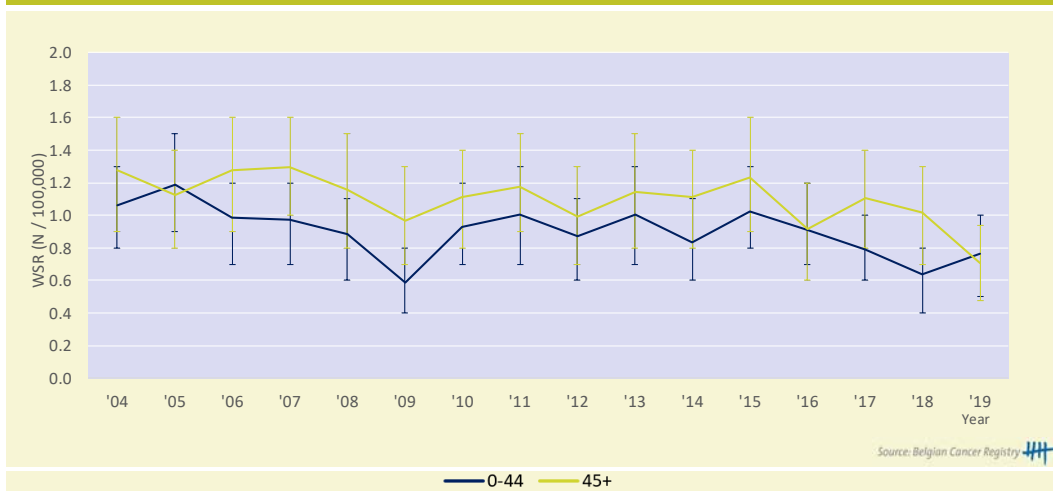
Incidence trends

Figure 3 Bone sarcoma: Age-standardised incidence rates* (WSR) by sex, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals.

Figure 4 Bone sarcoma: Age-standardised incidence rates* (WSR) by age category, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals.

Table 2 Bone sarcoma: Incidence trends by sex and age category in Belgium, 2004-2019

Age category	Males			Females		
	AAPC (%)	95%CI	Period	AAPC (%)	95%CI	Period
All ages	-2.6	[-4.2; -1.0]	2004-2019	-2.6	[-5.1; -0.1]	2004-2019
	-5.8	[-9.7; -1.7]	2004-2010			
	7.0	[1.0; 13.4]	2010-2014			
	-6.0	[-10.8; -0.8]	2014-2019			
0 - 44 y	-3.3	[-5.3; -1.2]	2004-2019	-2.2	[-5.7; 1.4]	2004-2019
	-5.5	[-9.6; -1.2]	2004-2011			
	8.5	[0.7; 17.0]	2011-2015			
	-10.3	[-17.9; -1.9]	2015-2019			
45+ y	-0.5	[-2.7; 1.8]	2004-2019	-3.5	[-5.4; -1.5]	2004-2019

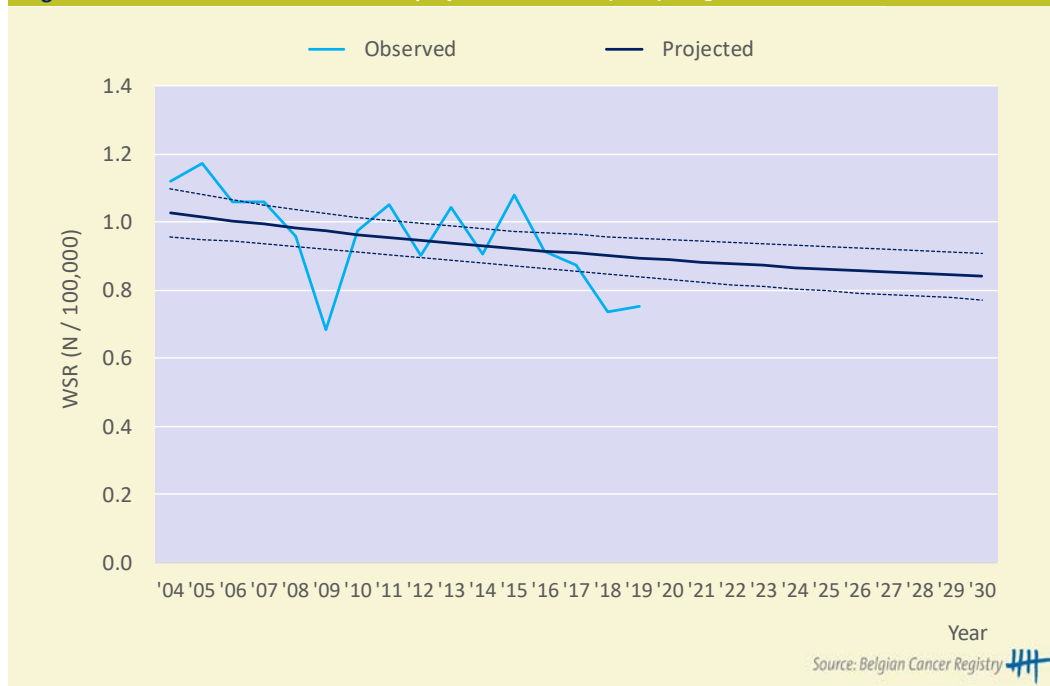
Source: Belgian Cancer Registry

AAPC: average annual percentage change

Period: When a joinpoint occurred, APC's are calculated for the period before and after the joinpoint. This column represents the corresponding time interval. AAPC's are always calculated over the entire study-period.

Incidence projections

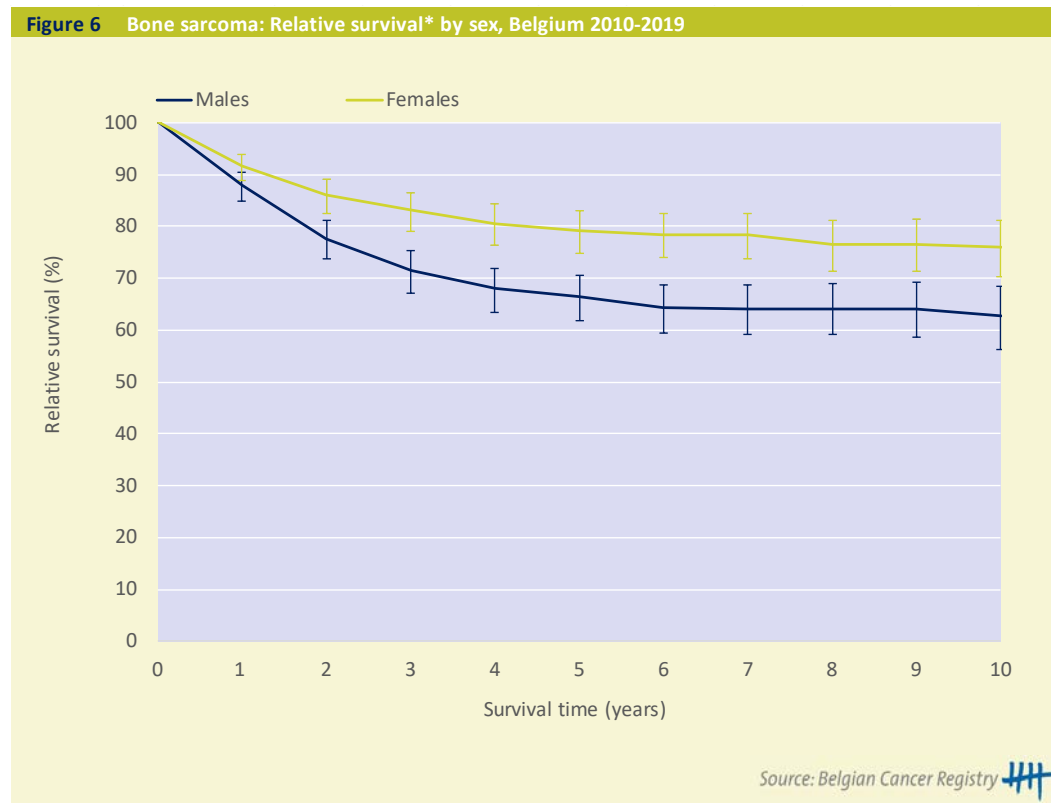
Figure 5 Bone sarcoma: Observed and projected incidence (WSR), Belgium 2004-2030



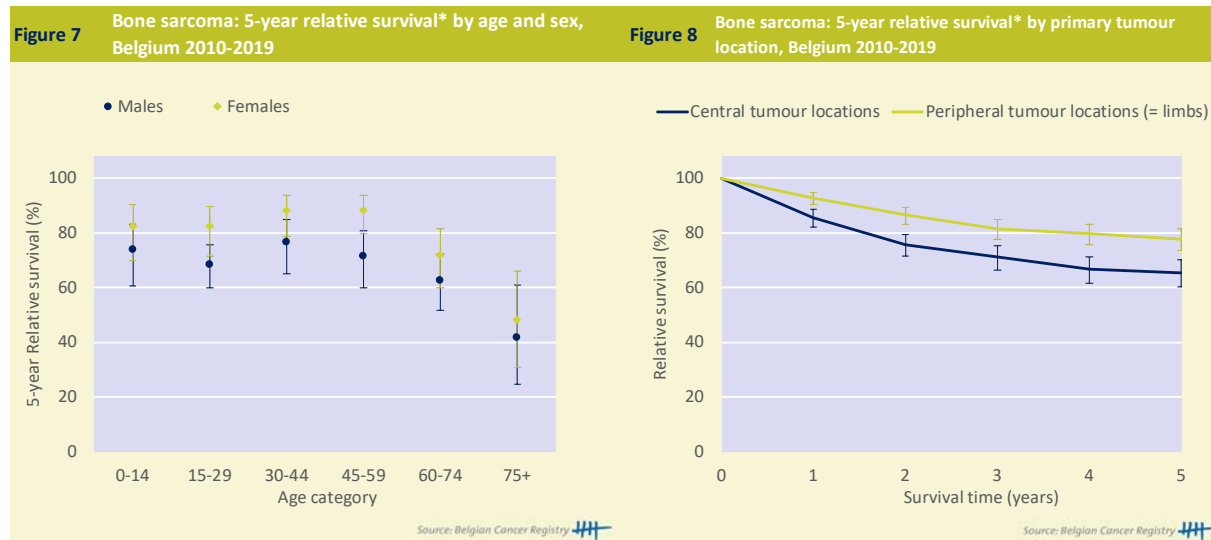
Source: Belgian Cancer Registry

WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

Survival



* The relative survival values are represented with 95% Confidence Intervals



* The relative survival values are represented with 95% Confidence Intervals

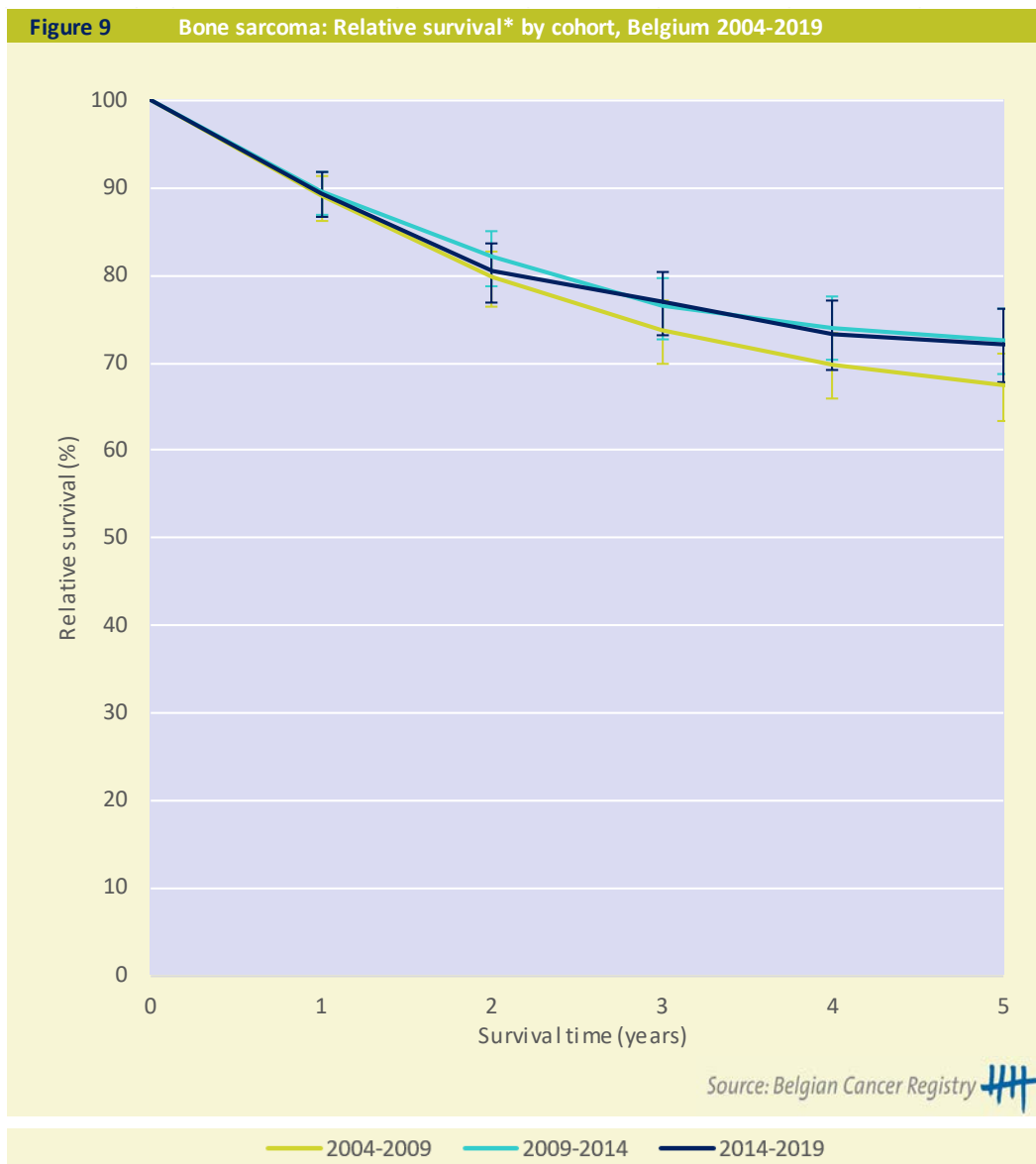
Table 3 Bone sarcoma: Conditional 5-year relative survival* by sex in Belgium, 2010-2019

Males		
X years since diagnosis	N at risk	%
1 year	511	73.1
2 year	416	82.5
3 year	341	89.9
Females		
X years since diagnosis	N at risk	%
1 year	443	85.6
2 year	391	91.0
3 year	346	92.2

* Unadjusted 5-yr relative survival probability conditional on surviving the first X years since diagnosis, %

* Interpretation in lay-man's terms: Given that a patient has already survived X years, what is the relative survival probability 5 years later.

Survival trends



* The relative survival values are represented with 95% Confidence Intervals

3.2 SARCOMAS CLASSIFIED BY HISTOLOGICAL TYPE

MAIN SUBTYPES:

- *Liposarcoma*
- *(Myo)fibroblastic tumours & so-called fibrohistiocytic tumours*
- *Vascular sarcoma*
- *Leiomyosarcoma*
- *Rhabdomyosarcoma*
- *Peripheral nerve sheath tumours (PNST)*
- *Other tumours of uncertain differentiation*
- *Gastrointestinal stromal tumour (GIST)*
- *Endometrial stromal sarcoma*
- *Ewing sarcoma*
- *Chondrosarcoma*
- *Osteosarcoma*
- *Other bone tumours of uncertain differentiation*
- *Unclassified and poorly characterised sarcoma*

KEYNOTES

Incidence (figure 1-7)

- Overall, gastro-intestinal stromal tumours (GIST) are the most frequently occurring sarcomas (23%) followed by liposarcoma in males (14%) and leiomyosarcoma in females (16%).
- Most sarcomas can be diagnosed at any age, except endometrial stromal sarcoma, malignant solitary fibrous tumour, Kaposi sarcoma and GIST which are not detected in children younger than 14 years. Conversely, rhabdoid tumours are never diagnosed in patients older than 40 years.
- The incidence of the different histological types of sarcomas varies with age:
 - In young children (younger than 10 years), rhabdomyosarcoma is the most common sarcoma (37%).
 - In older children and adolescent (10-19 years), osteosarcoma (31%) and Ewing sarcoma (23%) are the most common.
 - In young adults (20-44 years), malignant (myo)fibroblastic and fibrohistiocytic tumours represent the most common category (mostly dermatofibrosarcoma protuberans).
 - In older adults, GIST predominates.

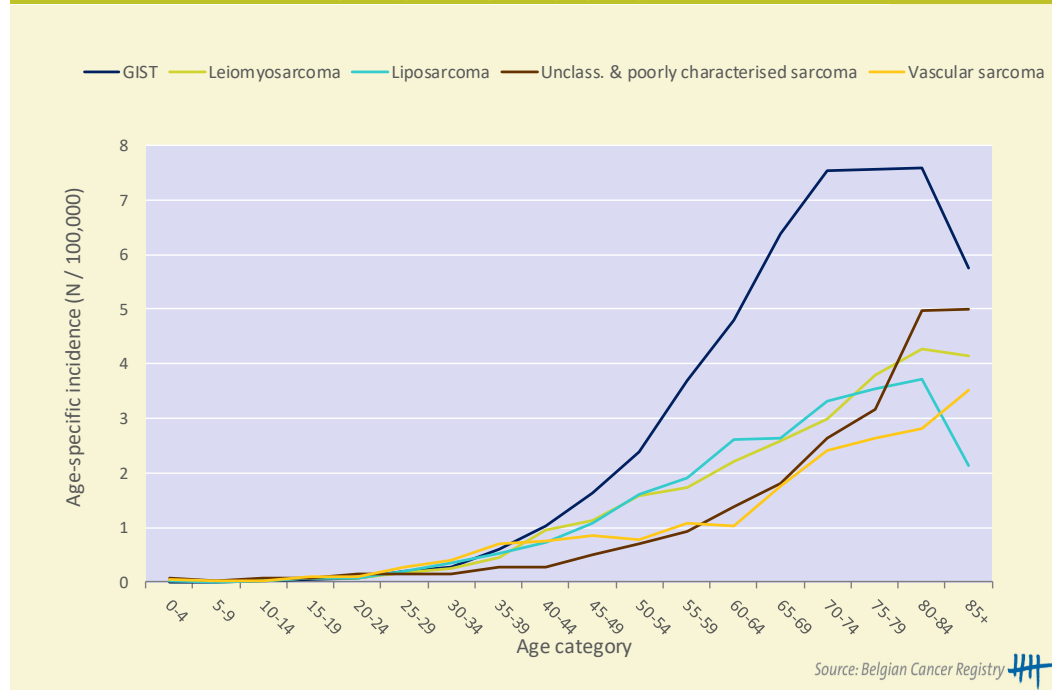
Survival (figure 8)

- The prognosis of sarcoma patients differs to a great extent between the histological subtypes, with the best 5-year relative survival for dermatofibrosarcoma protuberans (100%) patients followed by GIST (91%) patients and the worst for patients with angiosarcoma (30%) and rhabdoid tumours (32%).

Incidence

Figure 1

GIST, leiomyosarcoma, liposarcoma, unclassified & poorly characterised sarcoma & vascular sarcoma: Incidence by subtype and age category, Belgium 2004-2019



GIST: Gastrointestinal stromal tumour. This graph groups the most frequent subtypes.

Figure 2

Malignant (myo)fibroblastic & fibrohistiocytic tumours, chondrosarcoma, osteosarcoma & other tumours of uncertain differentiation: Incidence by subtype and age category, Belgium 2004-2019

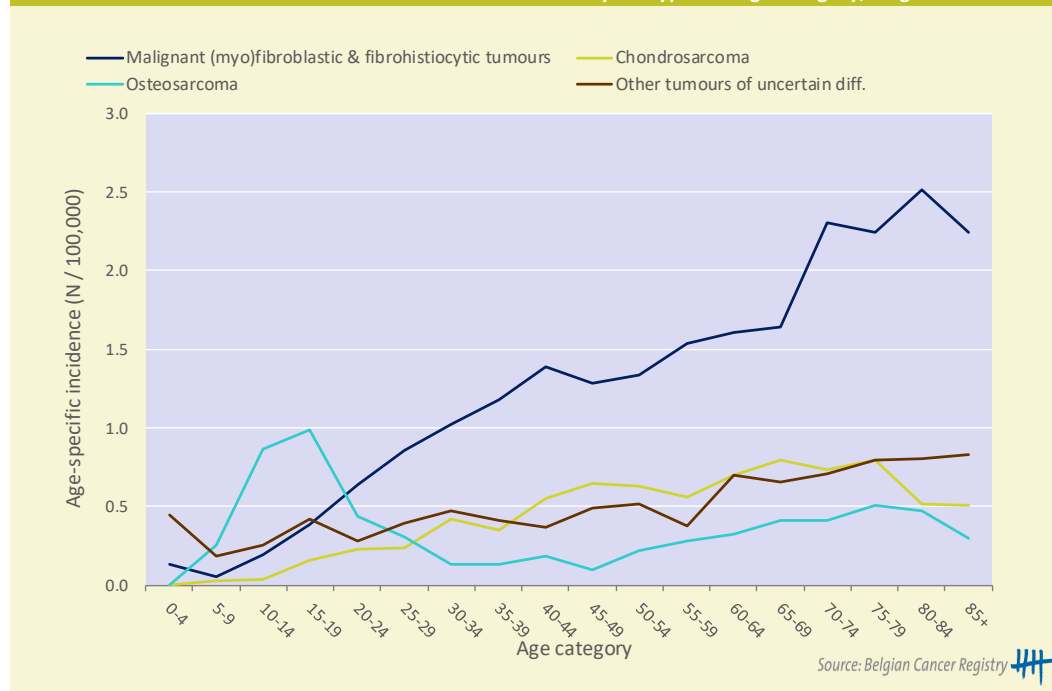
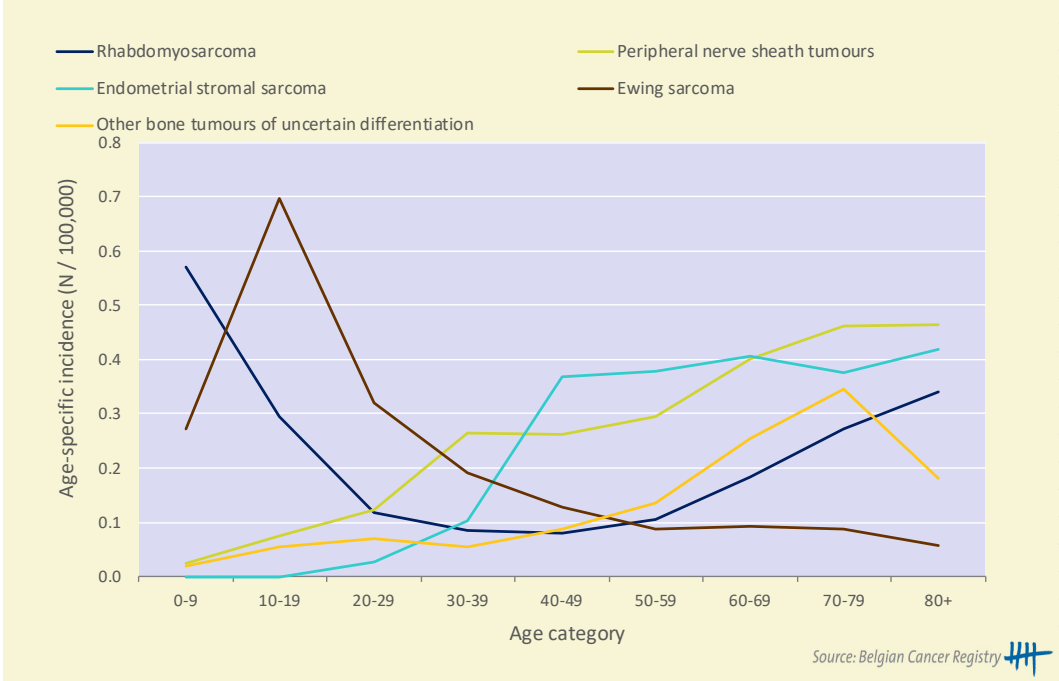
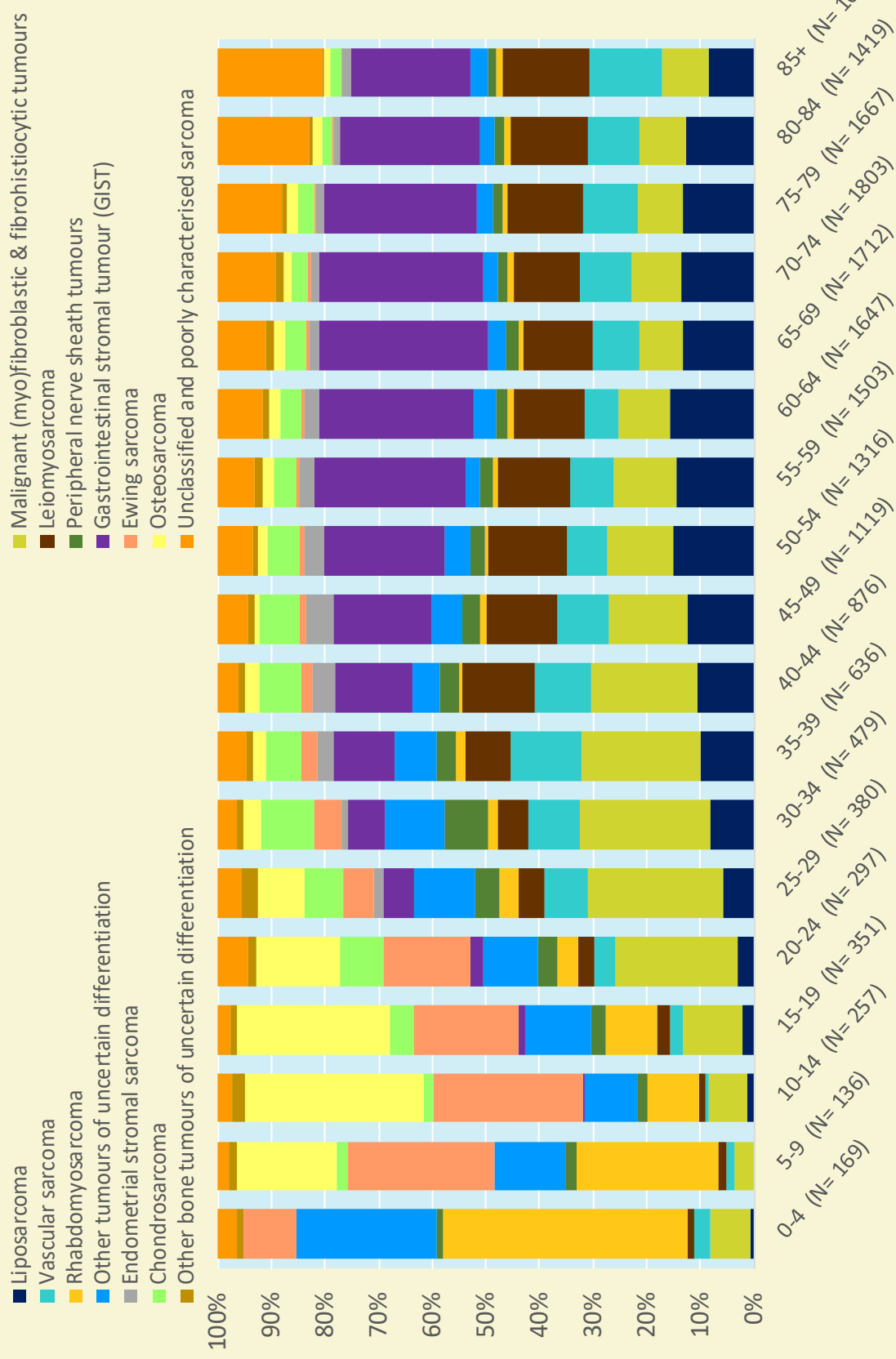


Figure 3 Rhabdomyosarcoma, peripheral nerve sheath tumours, endometrial stromal sarcoma, Ewing sarcoma & other bone tumours of uncertain differentiation: Incidence by subtype and age category, Belgium 2004-2019



This graph groups the more rare subtypes.

Figure 4 All sarcomas: incidence distribution by histological subtype and age category, Belgium, 2004-2019



Age category

Source: Belgian Cancer Registry

Figure 5 All sarcomas: Incidence by histological subtype and sex in Belgium, 2010-2019

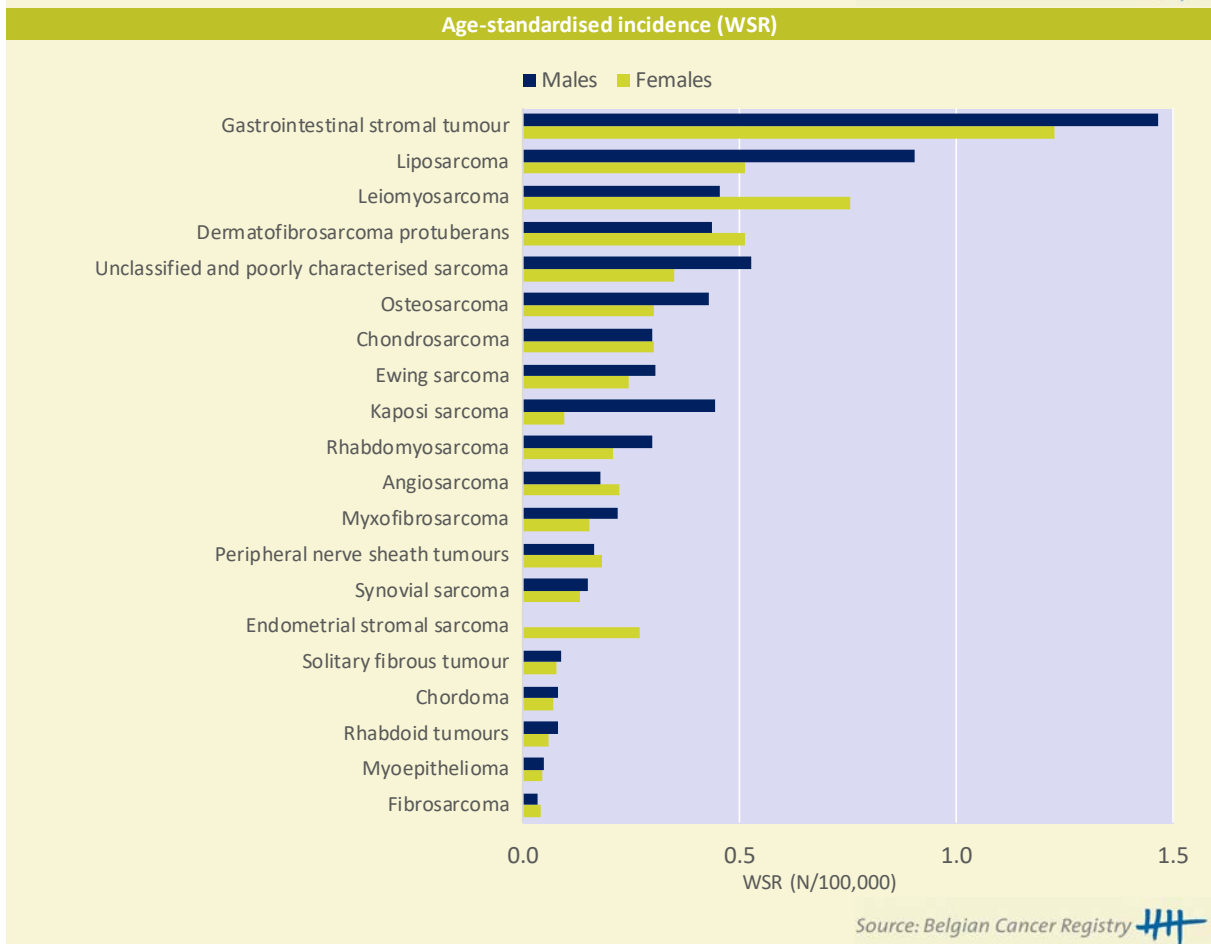
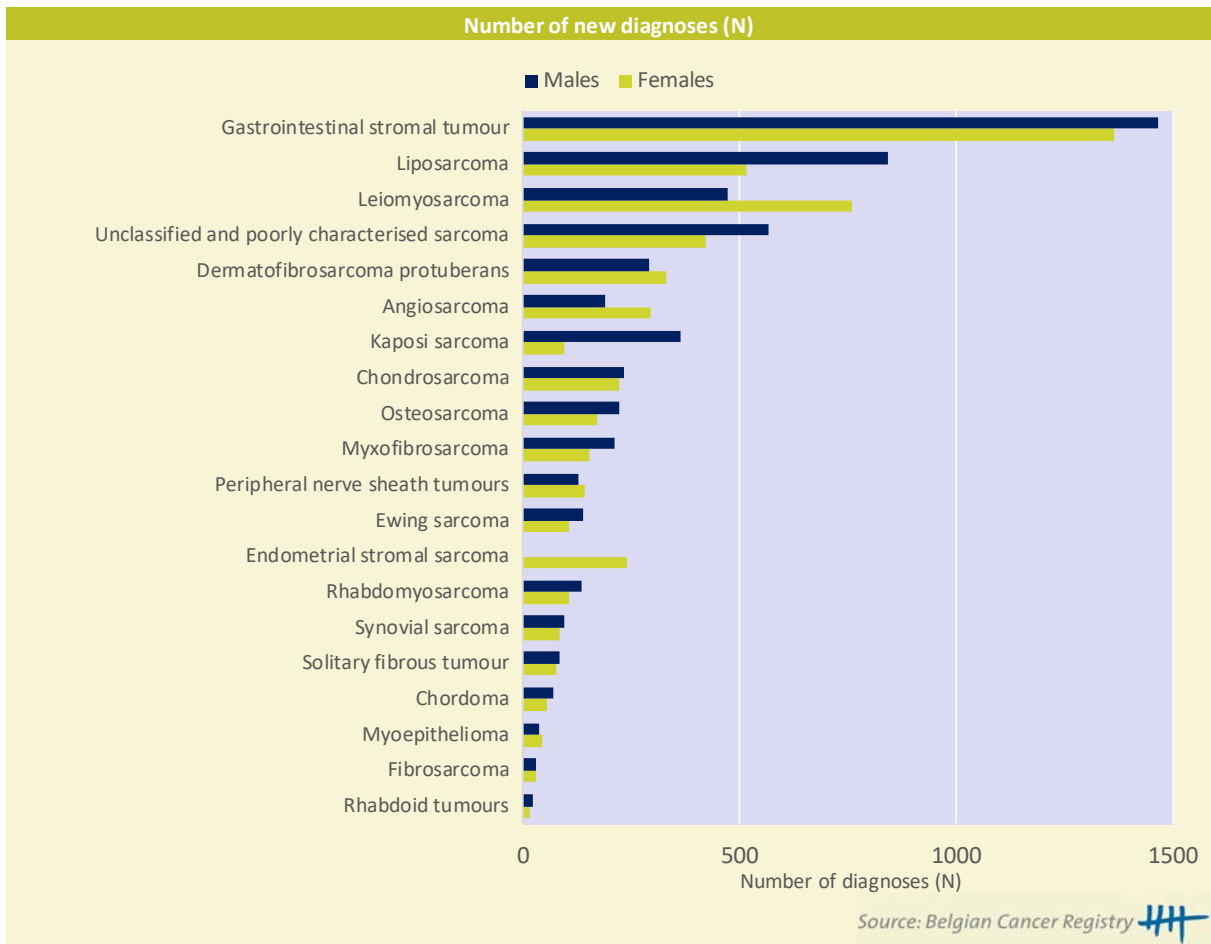
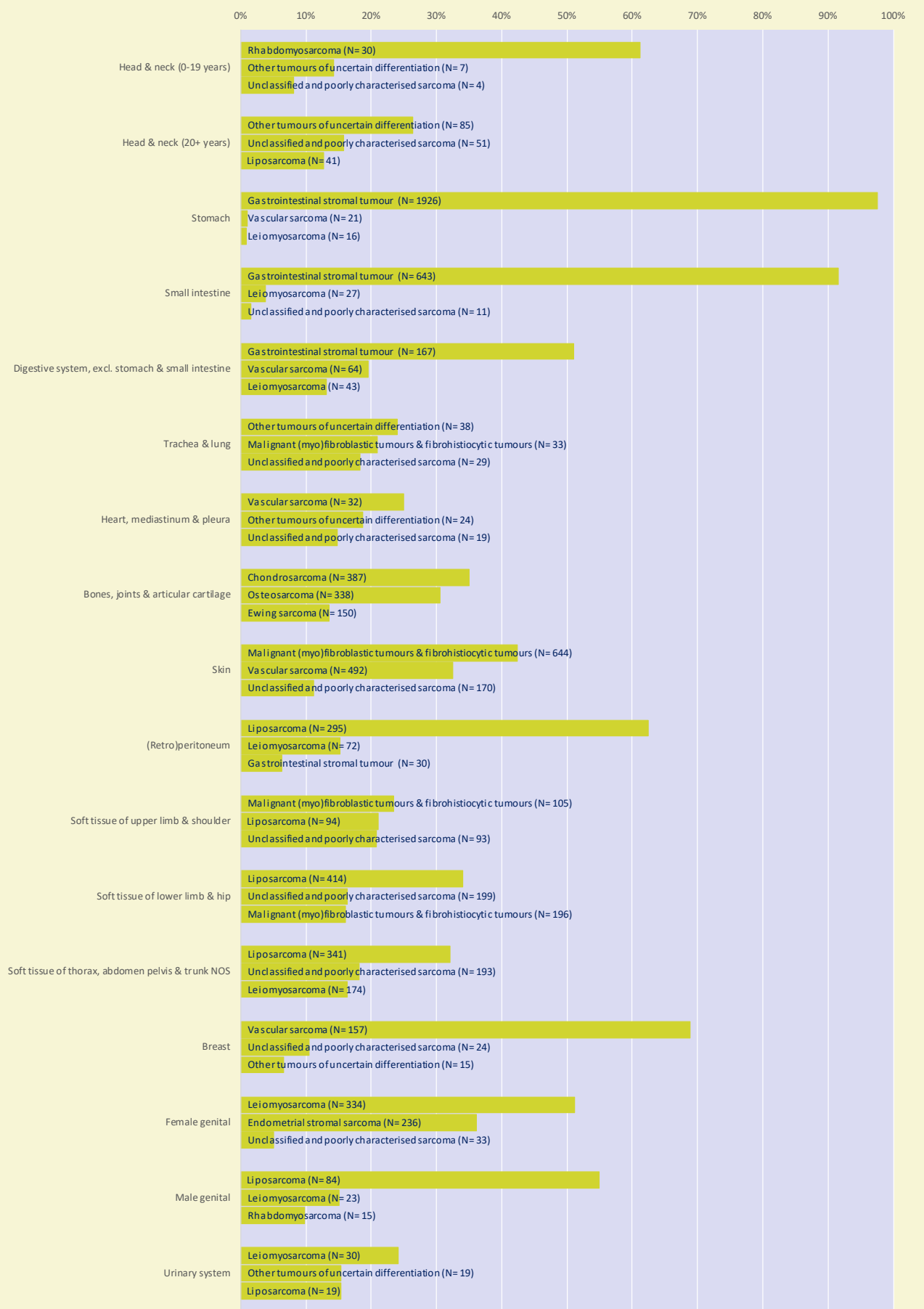


Figure 6 All sarcomas: Median age (years) at diagnosis* by histological subtype, Belgium, 2010-2019

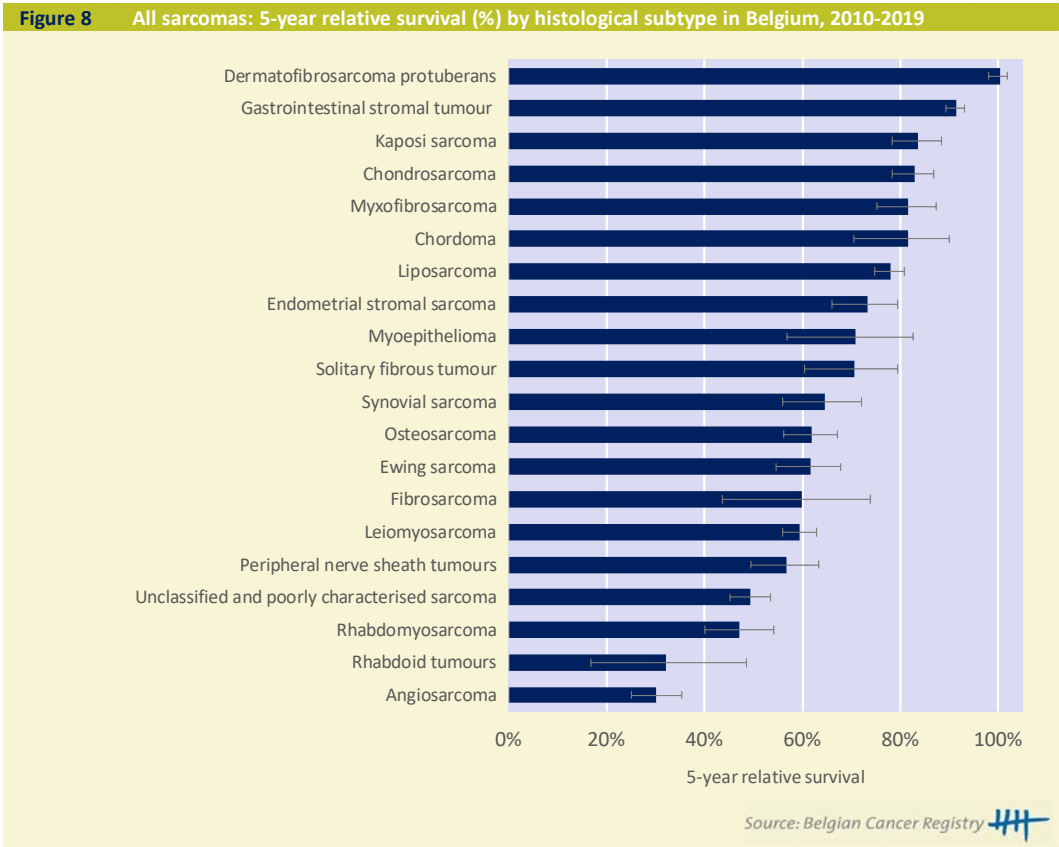


* Each bar in the figure represents the median age with the interquartile range (box containing the first quartile (Q1) and the third quartile (Q3)) and range (whiskers).

Figure 7 All sarcomas: most frequent histological subtypes (%) by primary tumour location, Belgium 2010-2019



Survival



The relative survival values are represented with 95% Confidence Intervals

3.2.1 LIPOSARCOMA

MAIN SUBTYPES:

- *Atypical lipomatous tumour/well differentiated liposarcoma (ALT/WDLPS)*
- *Dedifferentiated liposarcoma (DDLPS)*
- *Myxoid liposarcoma (MLPS)*
- *Pleomorphic liposarcoma*

KEYNOTES

Incidence (table 1-2; figure 1-9)

- Liposarcoma is more frequent in males than in females (M/F ratio= 1.8). It is the second most frequently occurring type of sarcoma in males (after GIST). In females liposarcoma is ranked third (after GIST and leiomyosarcoma) (see chapter 3.2).
- The incidence rate of liposarcoma increases with age, with peak incidence occurring around 75 years.
- The dominant liposarcoma subtype differs between age categories:
 - In younger patients (<45 years), MLPS is the dominant subtype (50%).
 - In older patients (45+), ALT/WDLPS (38%) and DDLPS are more frequent (39%).
- Liposarcoma arises in soft tissues, with most cases occurring in the hip & legs, the trunk and peritoneum. It is the most frequent sarcoma type arising in the peritoneum, male genital organs and in soft tissue of lower limbs, trunk, abdomen, and pelvis (see chapter 3.2).
- Between 2004 and 2019, the incidence rate is increasing in males, especially in the oldest age group (60+) and mostly for ALT/WDLPS and DDLPS. Potential underlying factors that could explain this increasing trend are improved registration, especially for atypical lipomatous tumours with intermediate biologic potential, improved diagnosis and classification changes.

Survival (table 3; figure 10-15)

- The 5-year relative survival is similar between males and females in all age categories.
- It varies according to:
 - The tumour grade from 95% for grade 1 to 50% for grade 3/4.
 - The primary tumour location with a better prognosis for peripheral tumours (88% vs. 71%).
 - The age of diagnosis from 90% in younger patients (0-44 years) to less than 70% in patients of 75+.
 - The subtype from 100% for ALT/WDLPS (even in older patients) to ~60% for DDLPS.
- The trends of the relative survival suggest a small improvement over time, especially at 3 years after diagnosis. However, this should be interpreted with caution as changes in the classification makes it difficult to interpret. E.g. the identification of liposarcoma subgroups and differentiation between benign lipomas and grade 1 liposarcoma has improved due to the introduction of immunohistology and genetics.

Table 1 Liposarcoma: Overview of incidence, prevalence and survival by sex in Belgium, 2010-2019				
Males				
Incidence	N	CR	WSR	
Incidence, 2010-2019	842	1.5	0.9	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	368	6.5	3.6	
10-year prevalence, 31.12.2019	582	10.3	5.7	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	838	77.7	[73.4;81.5]	
10-year relative survival, 2010-2019	838	70.6	[63.0;77.9]	
Females				
Incidence	N	CR	WSR	
Incidence, 2010-2019	513	0.9	0.5	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	221	3.8	2.1	
10-year prevalence, 31.12.2019	352	6.0	3.3	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	510	78.2	[73.3;82.7]	
10-year relative survival, 2010-2019	510	65.8	[56.8;74.2]	
Median age at diagnosis, 2010-2019 (y)	63 [Q1: 53; Q3: 74]			
M/F-ratio	1.8			

Source: Belgian Cancer Registry 

N: number of new diagnoses

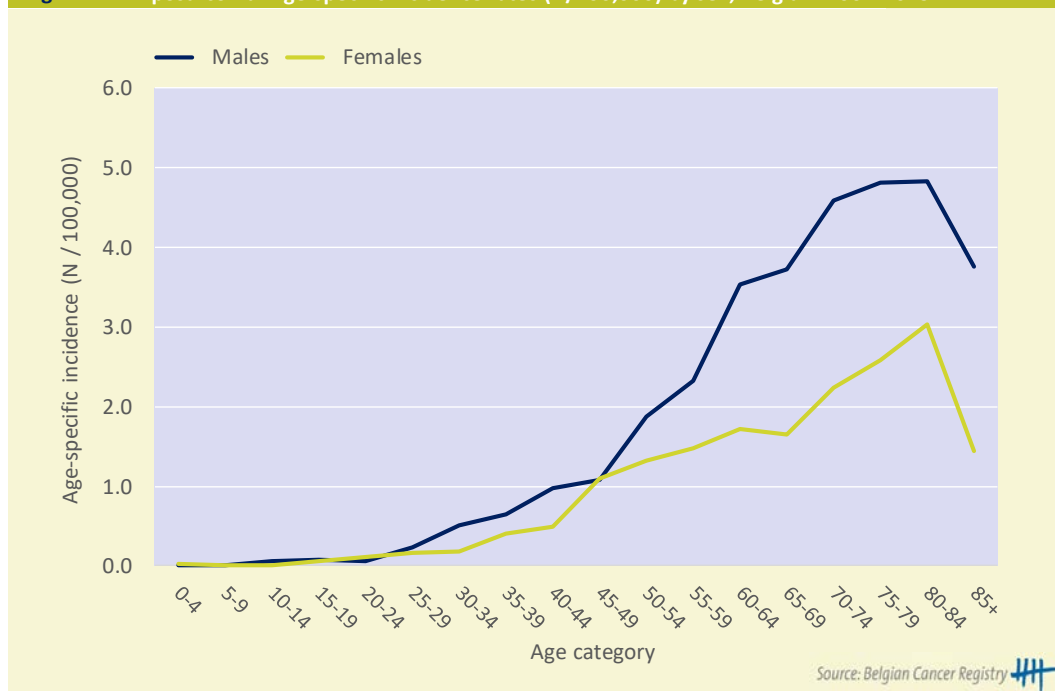
CR: crude (all ages) rate (N/100,000 person years)

WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

M/F-ratio: Male/Female ratio based on the age-standardised rates (2010-2019)

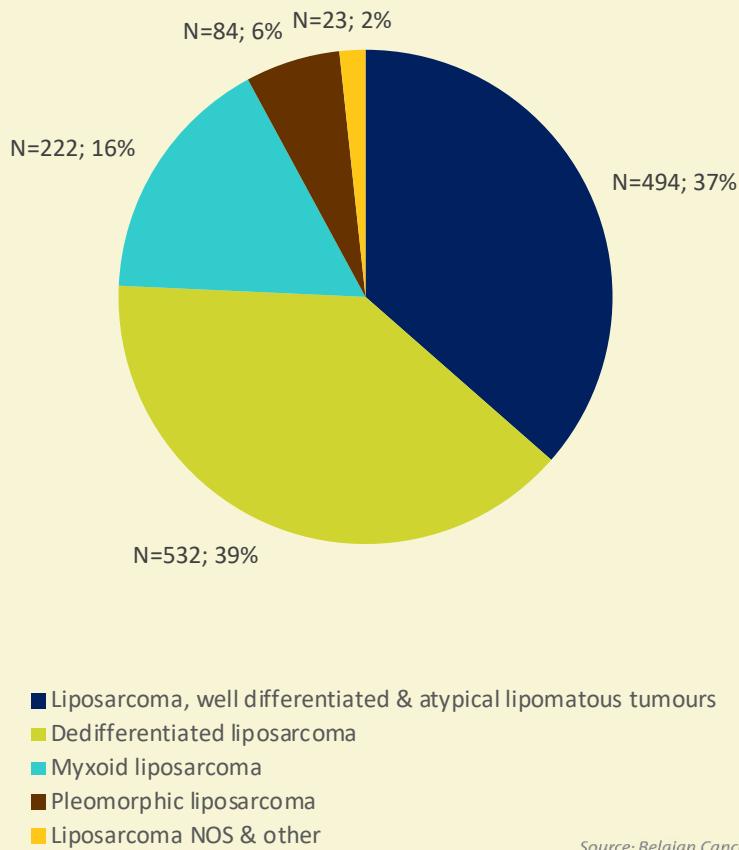
Incidence

Figure 1 Liposarcoma: Age-specific incidence rates (N/100,000) by sex, Belgium 2004-2019



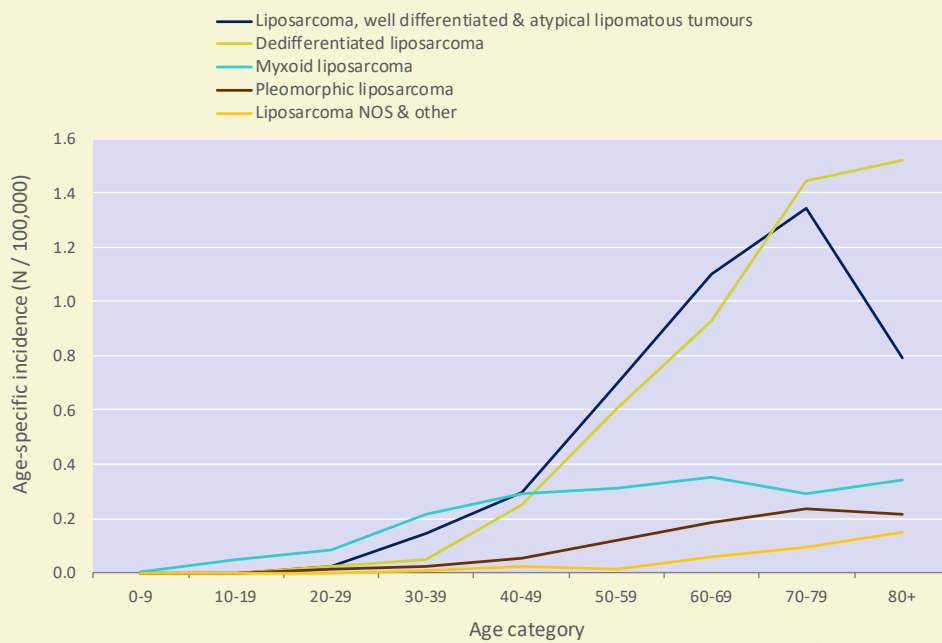
Source: Belgian Cancer Registry 

Figure 2 Liposarcoma: Incidence distribution by subtype, Belgium 2010-2019



Source: Belgian Cancer Registry

Figure 3 Liposarcoma: Age-specific incidence by subtype, Belgium 2004-2019



Source: Belgian Cancer Registry

Figure 4 Liposarcoma: Subtype incidence distribution (%) by age category, Belgium 2004-2019

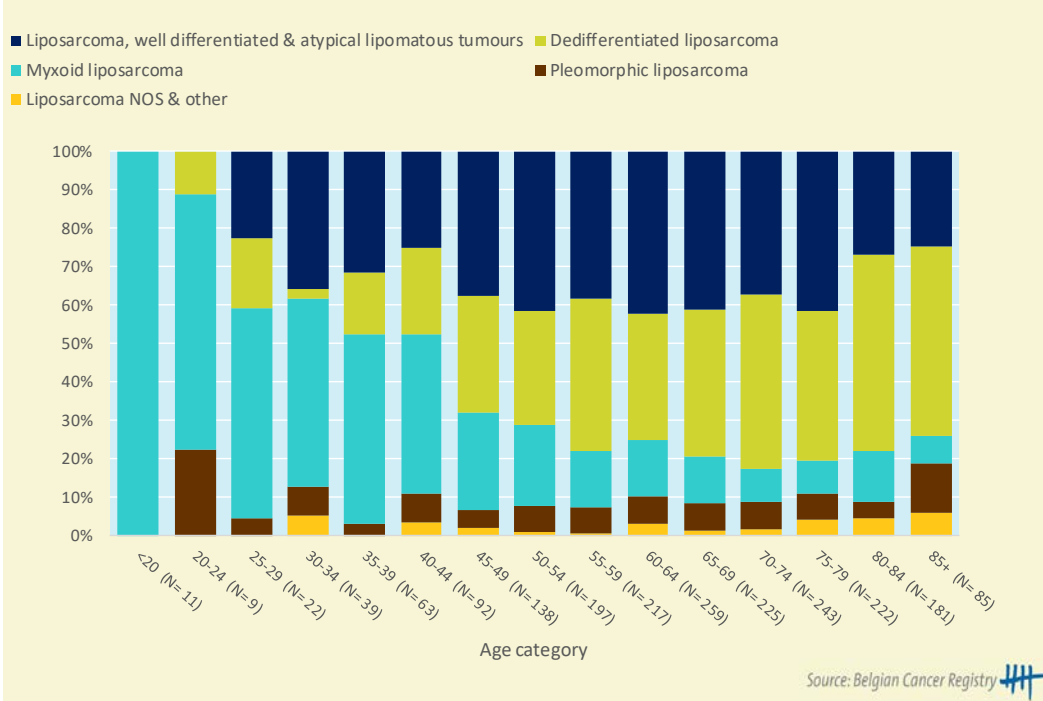
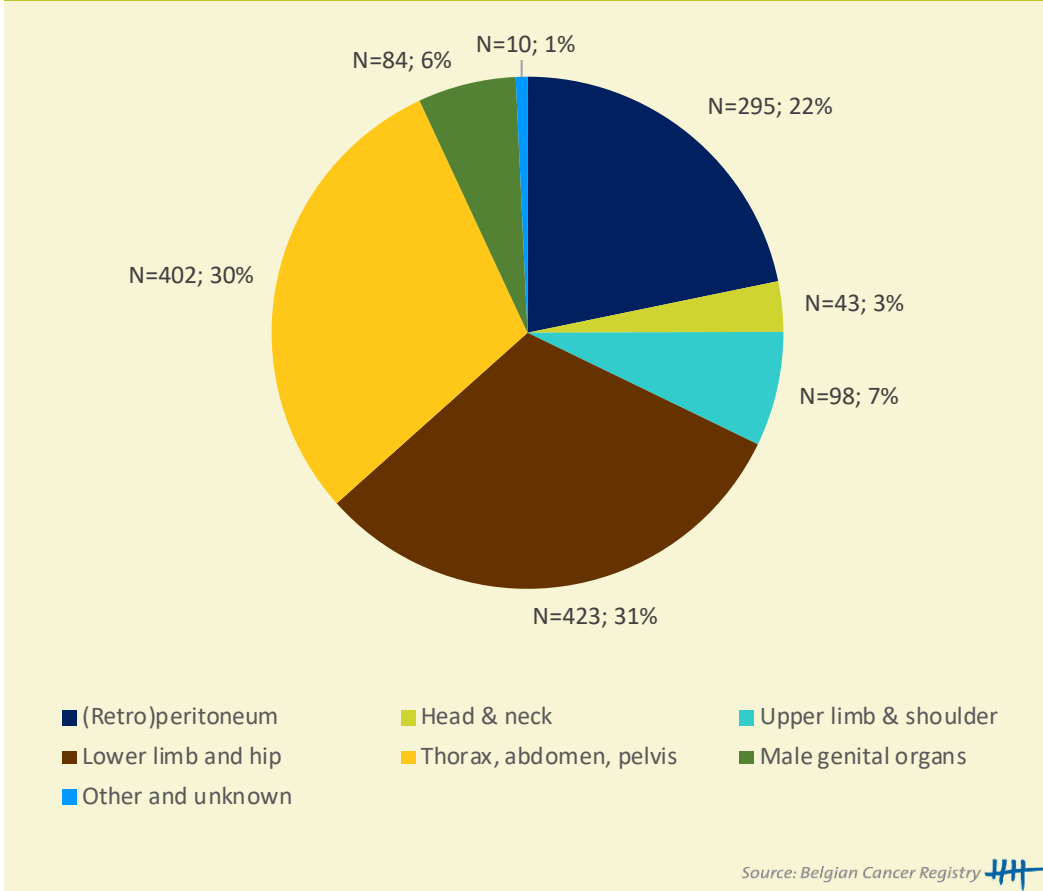


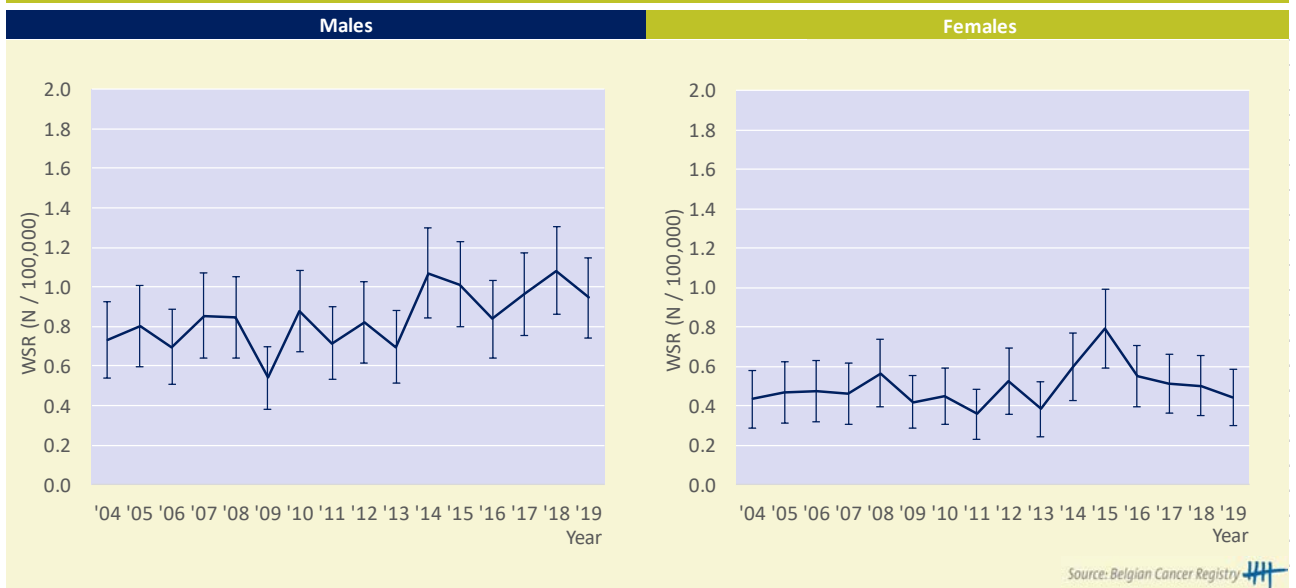
Figure 5 Liposarcoma: Incidence distribution by primary tumour location, Belgium 2010-2019



* Note: (retro)peritoneal cases occur almost exclusively in the peritoneum and retroperitoneal cases are very rare.

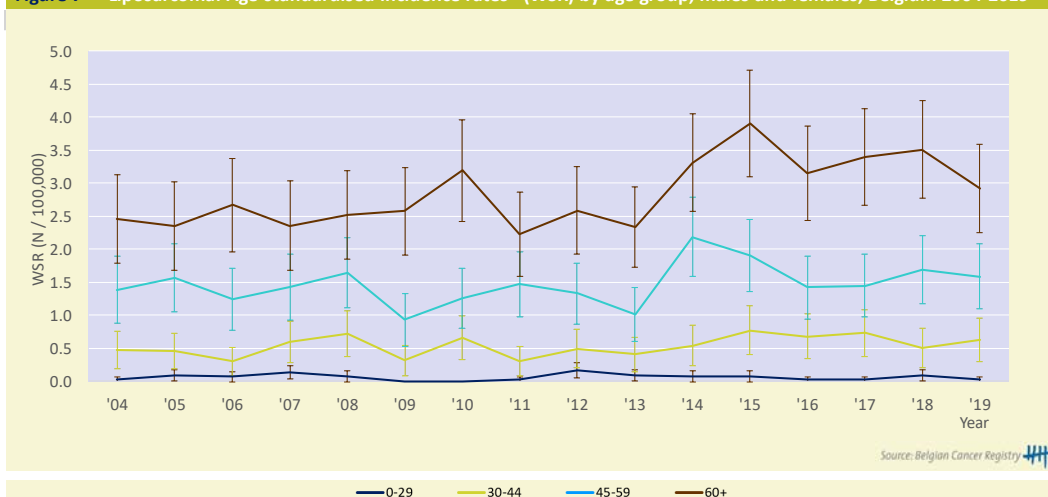
Incidence trends

Figure 6 Liposarcoma: Age-standardised incidence rates* (WSR) by sex, Belgium 2004-2019



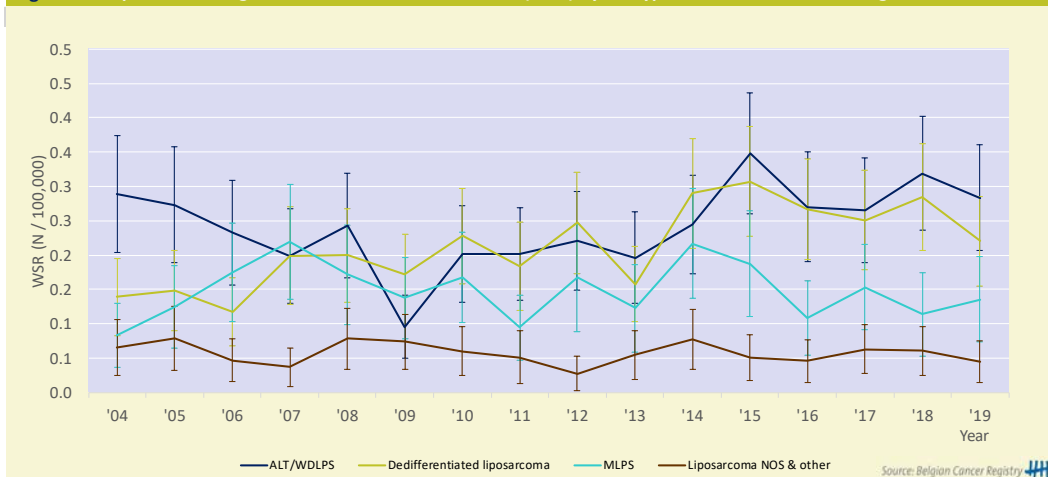
* The age-standardised incidence rates are represented with 95% Confidence Intervals.

Figure 7 Liposarcoma: Age-standardised incidence rates* (WSR) by age group, males and females, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals.

Figure 8 Liposarcoma: Age-standardised incidence rates* (WSR) by subtype, males and females, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals.

Table 2 Liposarcoma: Incidence trend by sex, age category and histological subtype in Belgium, 2004-2019

Incidence by age category	Males			Females		
	AAPC (%)	95%CI	Period	AAPC (%)	95%CI	Period
All ages	2.3	[0.5; 4.1]	2004-2019	-0.7	[-3.1; 1.8]	2004-2019
				-1.7	[-6.4; 3.2]	2004-2011
				8.5	[1.5; 16.0]	2011-2016
				-12.2	[-23.3; 0.5]	2016-2019
0 - 29 y	-	-	-	-	-	-
30 - 44 y	3.0	[-2.3; 8.6]	2004-2019	2.5	[-1.8; 7.0]	2004-2019
45 - 59 y	0.2	[-2.3; 2.8]	2004-2019	2.9	[-0.7; 6.6]	2004-2019
60+ y	3.6	[1.8; 5.4]	2004-2019	0.0	[-2.6; 2.7]	2004-2019
Incidence by subtype	AAPC (%)	95%CI	Period	AAPC (%)	95%CI	Period
ALT/WDLPS	0.7	[-1.6; 3.1]	2004-2019	0.6	[-3.0; 4.3]	2004-2019
	-12.3	[-18.9; -5.1]	2004-2009	-11.6	[-21.7; -0.0]	2004-2009
	7.9	[4.1; 11.9]	2009-2019	7.2	[1.4; 13.4]	2009-2019
DDLPS	5.2	[3.0; 7.5]	2004-2019	3.4	[-0.3; 7.2]	2004-2019
MLPS	-0.7	[-4.7; 3.6]	2004-2019	3.1	[-2.5; 9.0]	2004-2019
				31.4	[5.0; 64.4]	2004-2008
				0.0	[-14.2; 16.6]	2008-2013
				-10.1	[-21.6; 3.1]	2013-2019
Liposarcoma NOS & other	1.6	[-3.0; 6.4]	2004-2019	-6.0	[-13.4; 1.9]	2004-2019

Source: Belgian Cancer Registry

AAPC: average annual percentage change

ALT/WDLPS: Atypical lipomatous tumour/well differentiated liposarcoma

DDLPS: Dedifferentiated liposarcoma

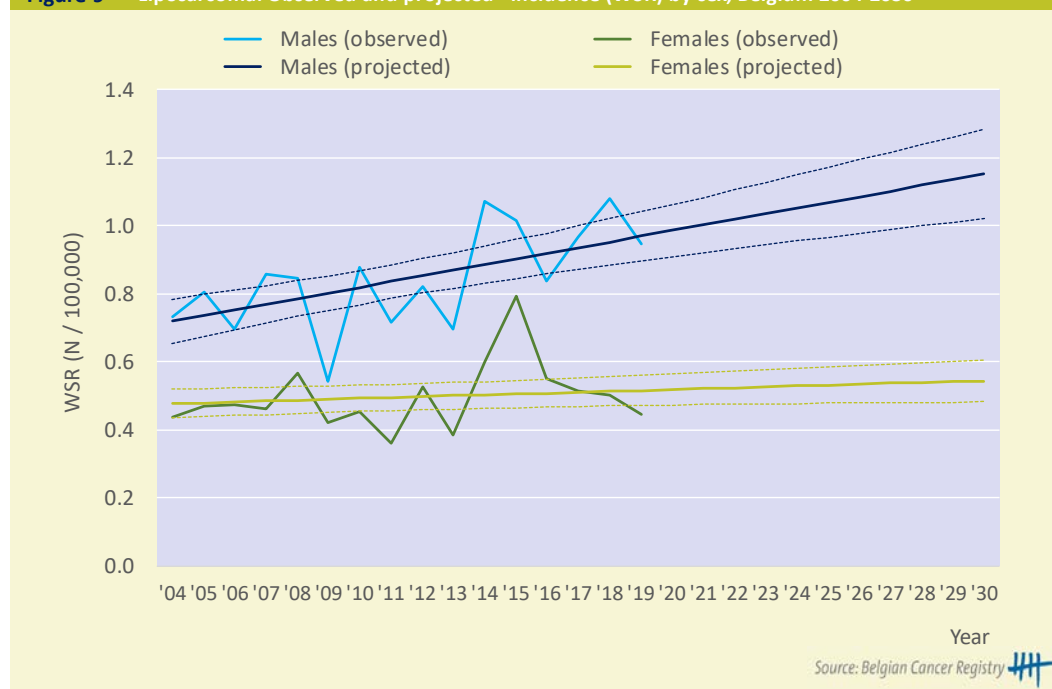
MLPS: Myxoid liposarcoma

NOS: not otherwise specified

Period: When a joinpoint occurred, APC's are calculated for the period before and after the joinpoint. This column represents the corresponding time interval. AAPC's are always calculated over the entire study-period.

Incidence projections

Figure 9 Liposarcoma: Observed and projected* incidence (WSR) by sex, Belgium 2004-2030

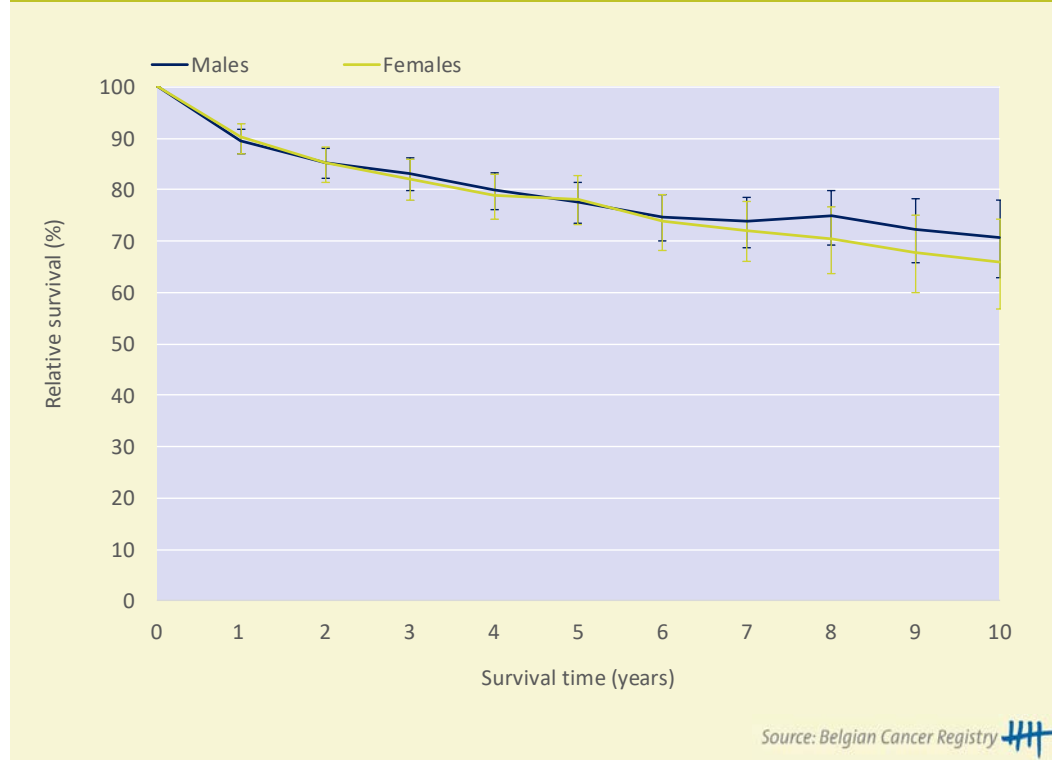


Source: Belgian Cancer Registry

WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

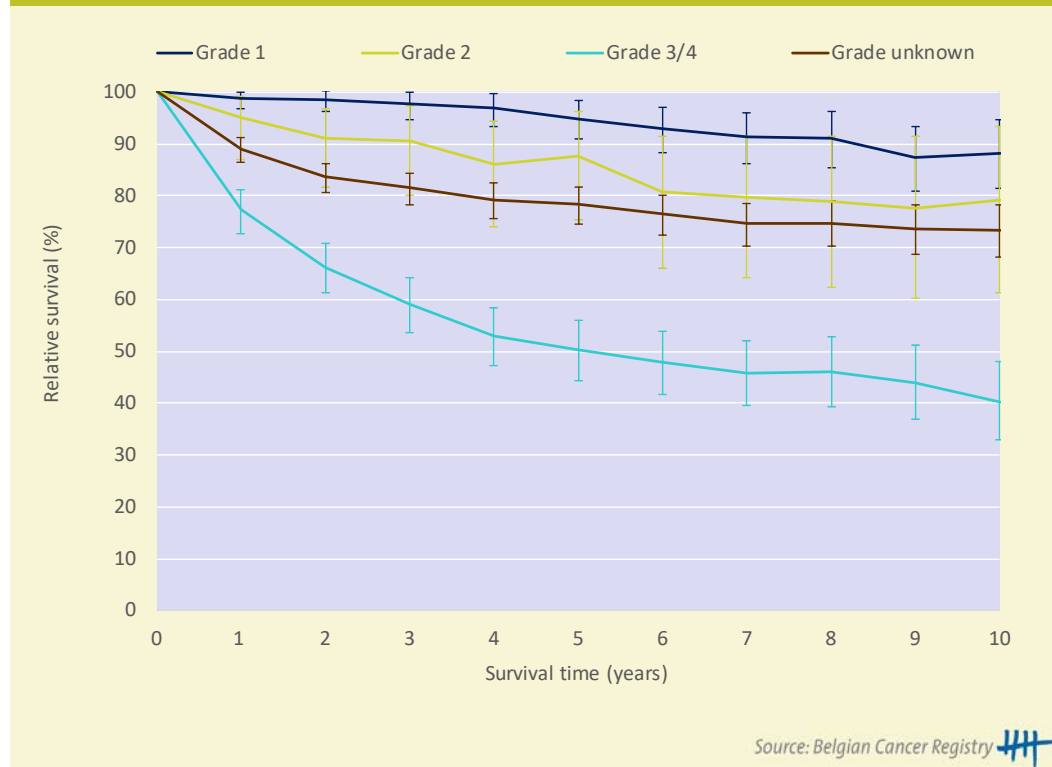
Survival

Figure 10 Liposarcoma: Relative survival* by sex, Belgium 2010-2019



* The relative survival values are represented with 95% Confidence Intervals

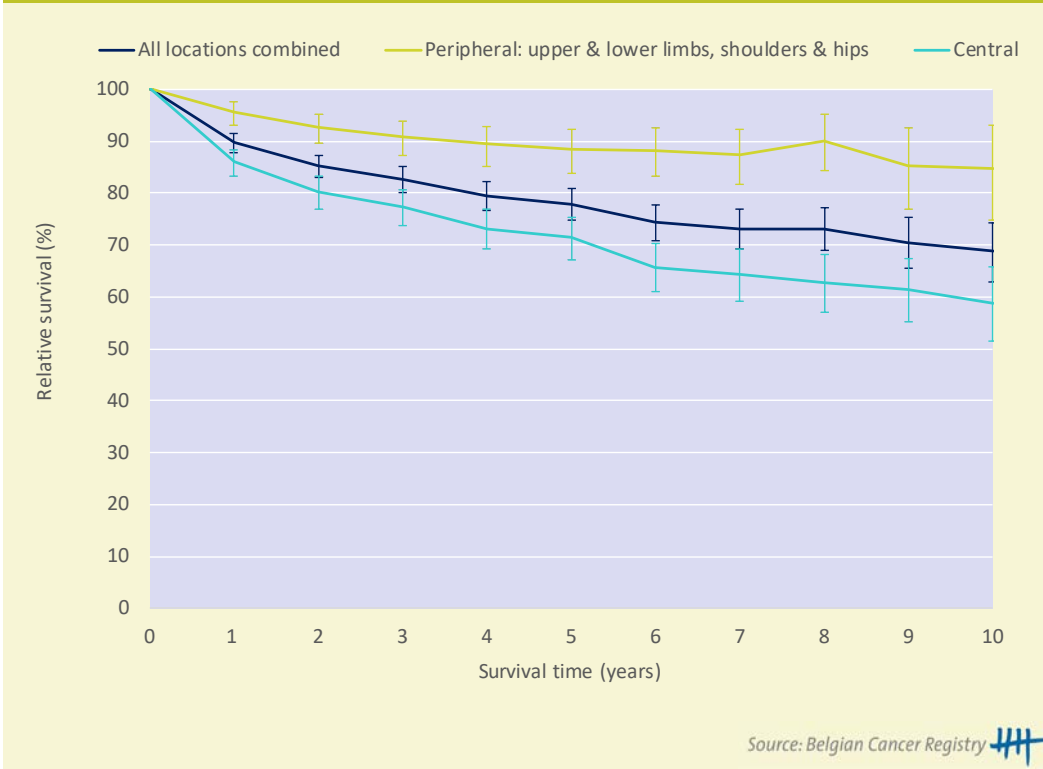
Figure 11 Liposarcoma: Relative survival* by grade, Belgium 2004-2019



Grade based on the FNCLCC grading or the differentiation grade (WHO). Therefore grade 3 and 4 (which is not included in FNCLCC) were combined. Note that 45% of cases were registered without information on tumour grade (i.e. grade unknown).

* The relative survival values are represented with 95% Confidence Intervals

Figure 12 Liposarcoma: Relative survival* by primary tumour location, Belgium 2004-2019



Survival data for unspecified primary tumour locations is not shown (N=10)
 * The relative survival values are represented with 95% Confidence Intervals

Figure 13 Liposarcoma: 5-year relative survival* by age and sex, Belgium 2010-2019

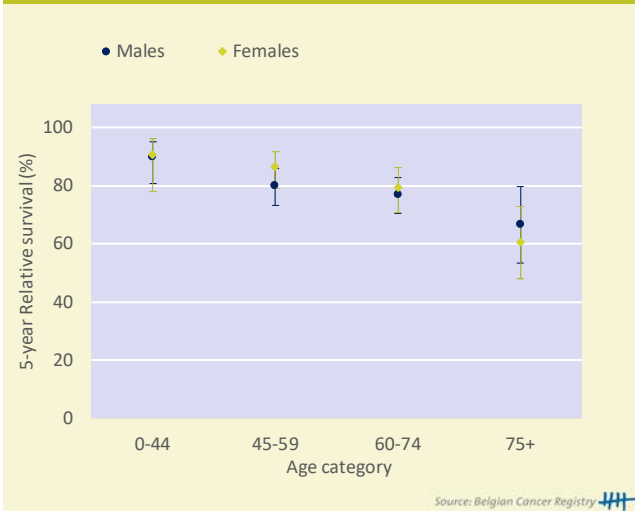
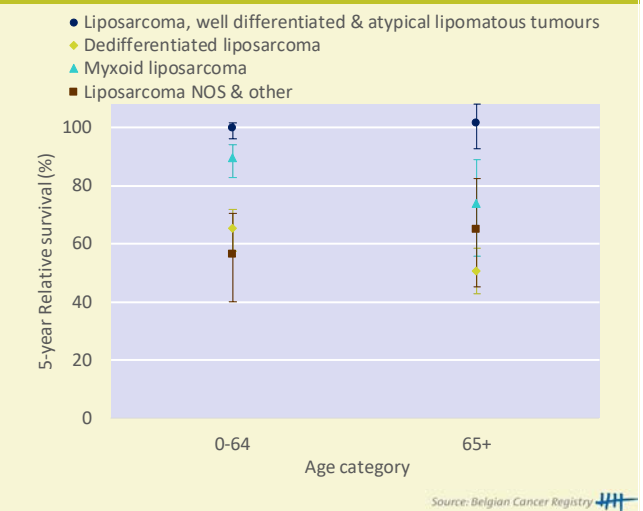


Figure 14 Liposarcoma: 5-year relative survival* by subtype, Belgium 2010-2019



* The relative survival values are represented with 95% Confidence Intervals

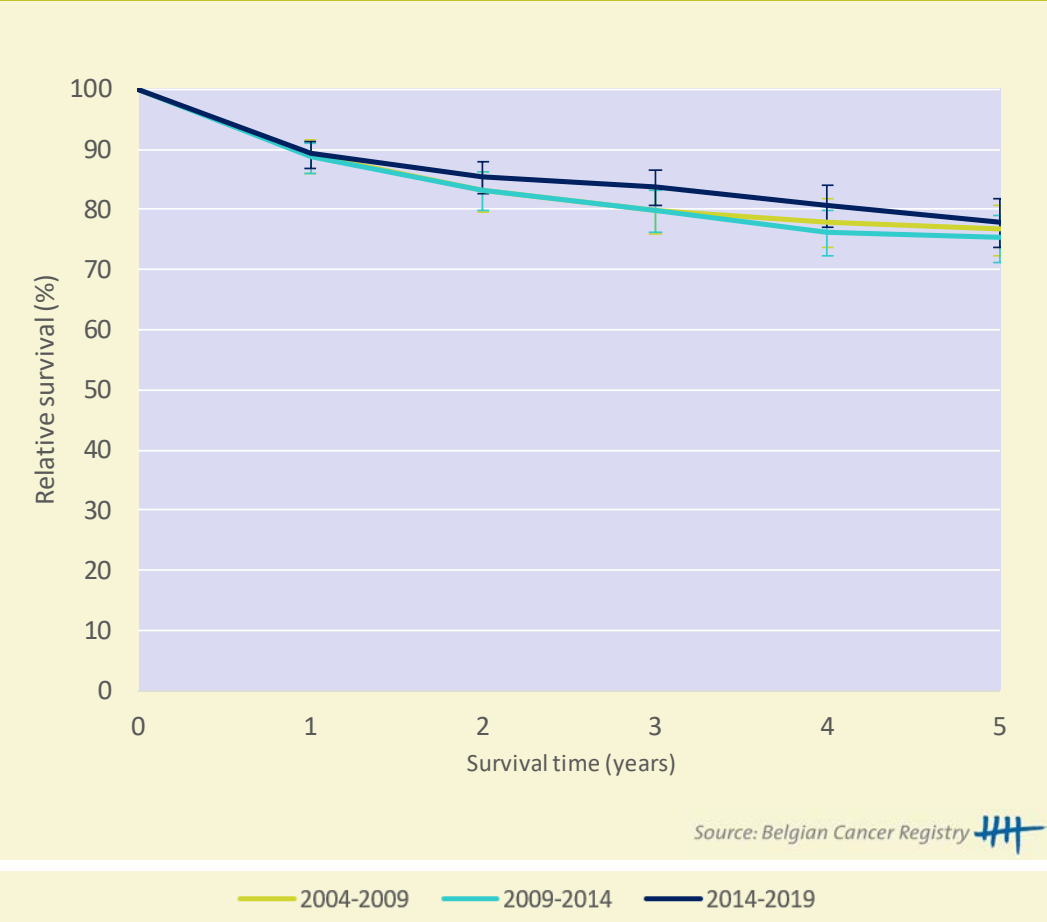
Table 3 Liposarcoma: Conditional 5-year relative survival* in Belgium, 2010-2019

X years since diagnosis	N at risk	%
1 year	1,177	82.8
2 year	998	85.8
3 year	820	88.5

* Unadjusted 5-yr relative survival probability conditional on surviving the first X years since diagnosis, %
 * Interpretation in lay-man's terms: Given that a patient has already survived X years, what is the relative survival probability 5 years later.

Survival trends

Figure 15 Liposarcoma: Relative survival* by cohort, Belgium 2004-2019



* The relative survival values are represented with 95% Confidence Intervals

3.2.2 MALIGNANT (MYO)FIBROBLASTIC TUMOURS AND SO-CALLED FIBROHISTIOCYTIC TUMOURS

MAIN SUBTYPES:

- *Dermatofibrosarcoma protuberans (DFSP)*
- *Solitary fibrous tumour, malignant (MSFT)*
- *Fibrosarcoma*
- *Myxofibrosarcoma*
- *Miscellaneous malignant (myo)fibroblastic tumours and so-called fibrohistiocytic tumours*

KEYNOTES

Incidence (table 1-2; figure 1-7)

- The incidence of malignant (myo)fibroblastic tumours and fibrohistiocytic tumours increases with age, especially in males who show a higher incidence than females after the age of 65 years.
- This category of sarcomas is the most frequent one diagnosed in the skin (see chapter 3.2).
- Dermatofibrosarcoma protuberans is the most frequently occurring subtype, specifically before the age of 60 years. In older patients, myxofibrosarcoma is the dominant subtype. Fibrosarcoma presents typically in two age-peaks, the so-called 'infant' variant in very young children (<1 year) and to the 'adult' variant in older patients (>50 years).

Survival (table 3; figure 8-11)

- The 5-year relative survival of patients with malignant (myo)fibroblastic tumours and fibrohistiocytic tumours lies around 85% and given that a patient survives the first three years, the (conditional) relative survival probability 5 years later is above 96%.
- The relative 5-year survival of the patients:
 - Does not differ between males and females.
 - Is dependent on subtype with the best prognosis for dermatofibrosarcoma protuberans (100%), an intermediate rate for myxofibrosarcoma (82%) and the worst for fibrosarcoma (60%) patients.
 - Decreases with age, probably partly because of the reducing proportion of dermatofibrosarcoma protuberans after 60 years of age.
 - Does not show an improvement over time (2004-2019).

Table 1 Malignant (myo)fibroblastic tumours & so-called fibrohistiocytic tumours: Overview of incidence, prevalence and survival by sex in Belgium, 2010-2019				
Males				
Incidence	N	CR	WSR	
Incidence, 2010-2019	639	1.2	0.8	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	270	4.8	3.2	
10-year prevalence, 31.12.2019	490	8.7	5.9	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	630	87.5	[83.2;91.2]	
10-year relative survival, 2010-2019	630	90.1	[83.9;95.7]	
Females				
Incidence	N	CR	WSR	
Incidence, 2010-2019	614	1.1	0.8	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	271	4.6	3.4	
10-year prevalence, 31.12.2019	512	8.8	6.4	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	610	90.2	[86.6;93.3]	
10-year relative survival, 2010-2019	610	86.1	[79.9;91.4]	
Median age at diagnosis, 2010-2019 (y)	55 [Q1: 39; Q3: 70]			
M/F-ratio	1.0			

Source: Belgian Cancer Registry 

N: number of new diagnoses

CR: crude (all ages) rate (N/100,000 person years)

WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

M/F-ratio: Male/Female ratio based on the age-standardised rates (2010-2019)

Incidence

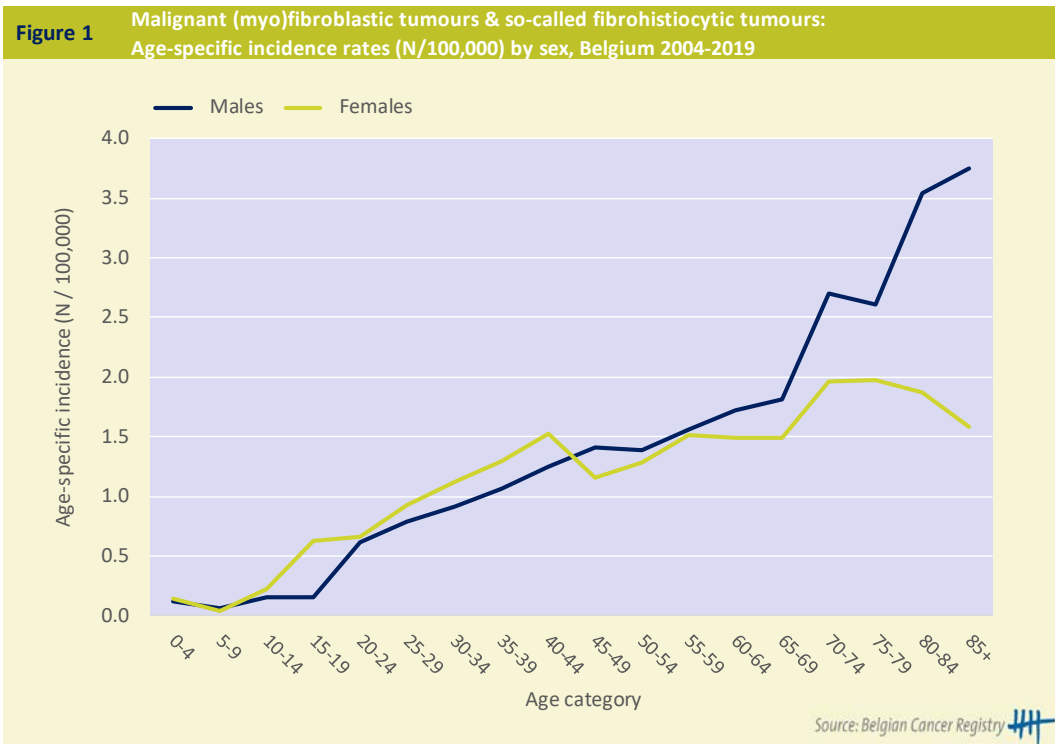
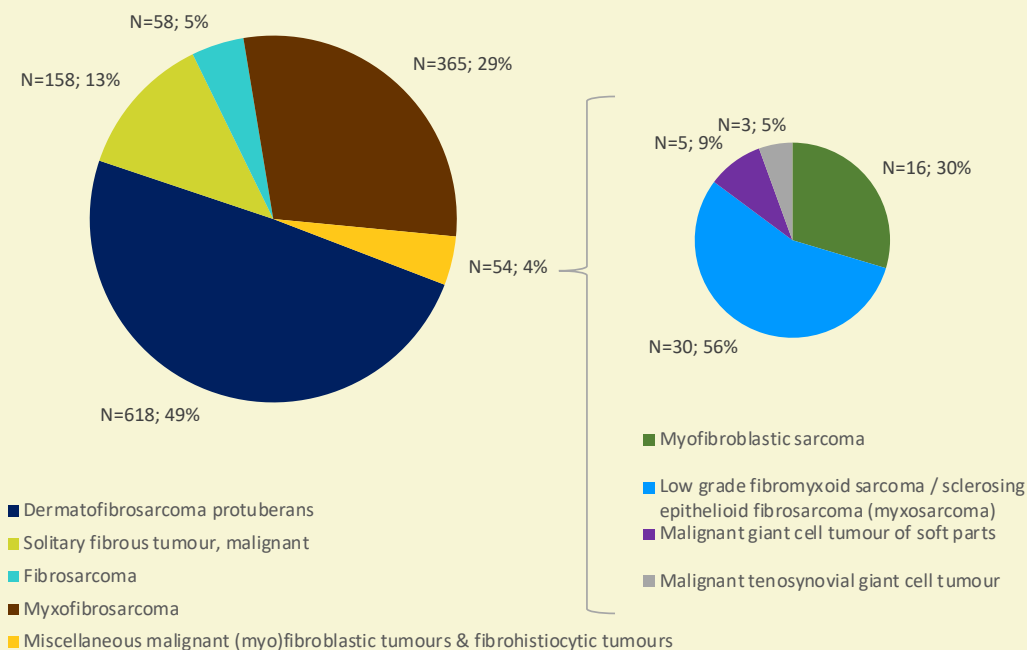
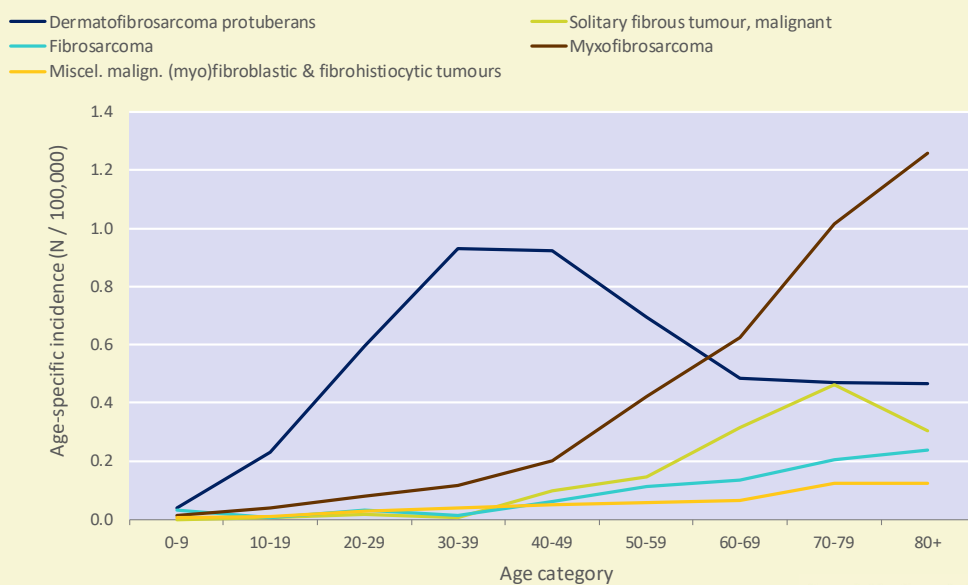


Figure 2 Malignant (myo)fibroblastic tumours & so-called fibrohistiocytic tumours: Incidence distribution by subtype, Belgium 2010-2019



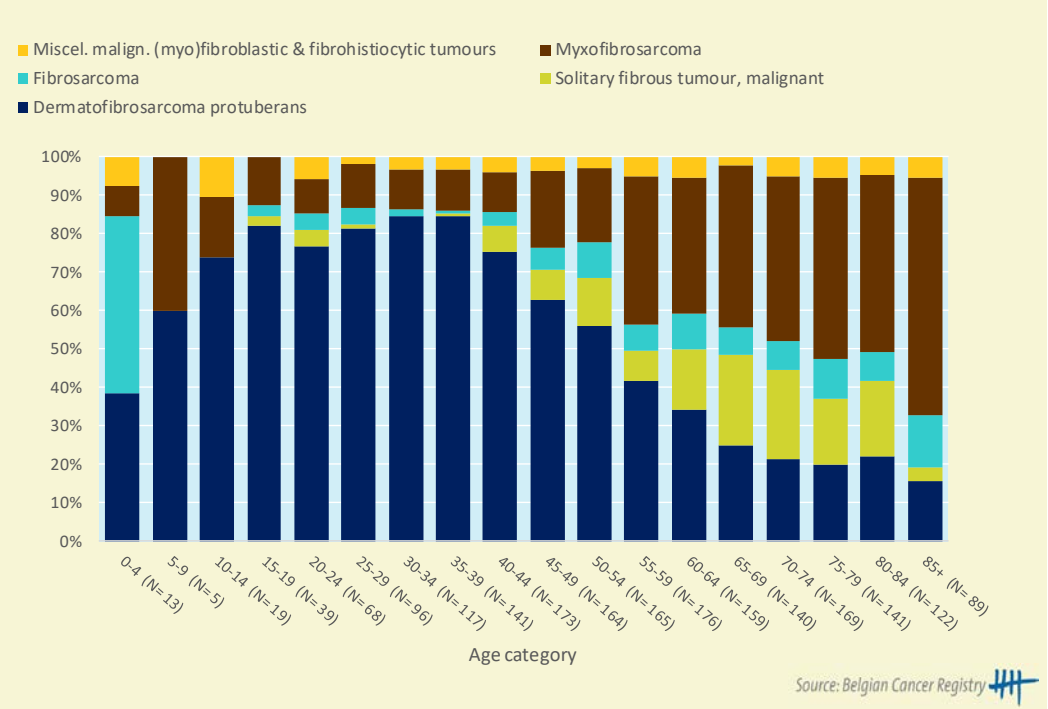
Source: Belgian Cancer Registry

Figure 3 Malignant (myo)fibroblastic tumours & so-called fibrohistiocytic tumours: Age-specific incidence by subtype, Belgium 2004-2019



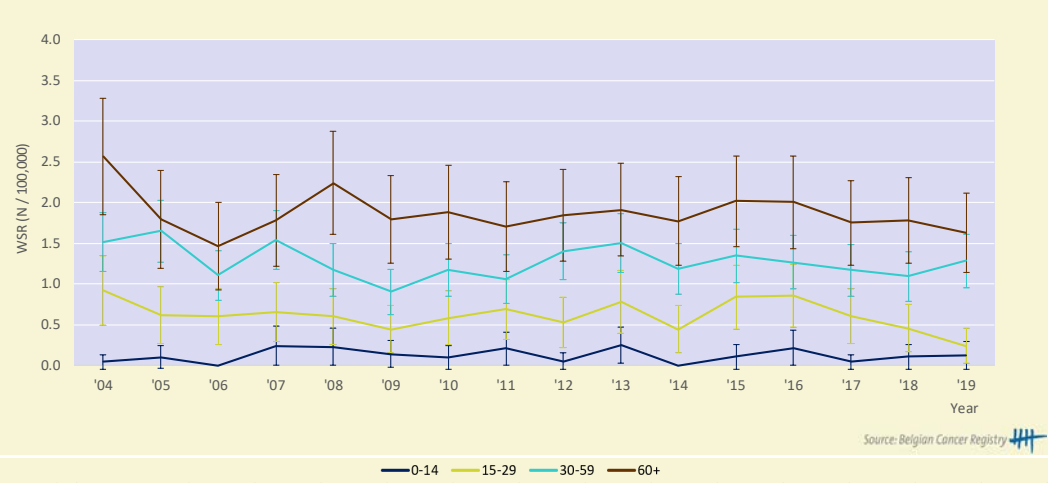
Source: Belgian Cancer Registry

Figure 4 Malignant (myo)fibroblastic tumours & so-called fibrohistiocytic tumours: Subtype incidence distribution (%) by age category, Belgium 2004-2019



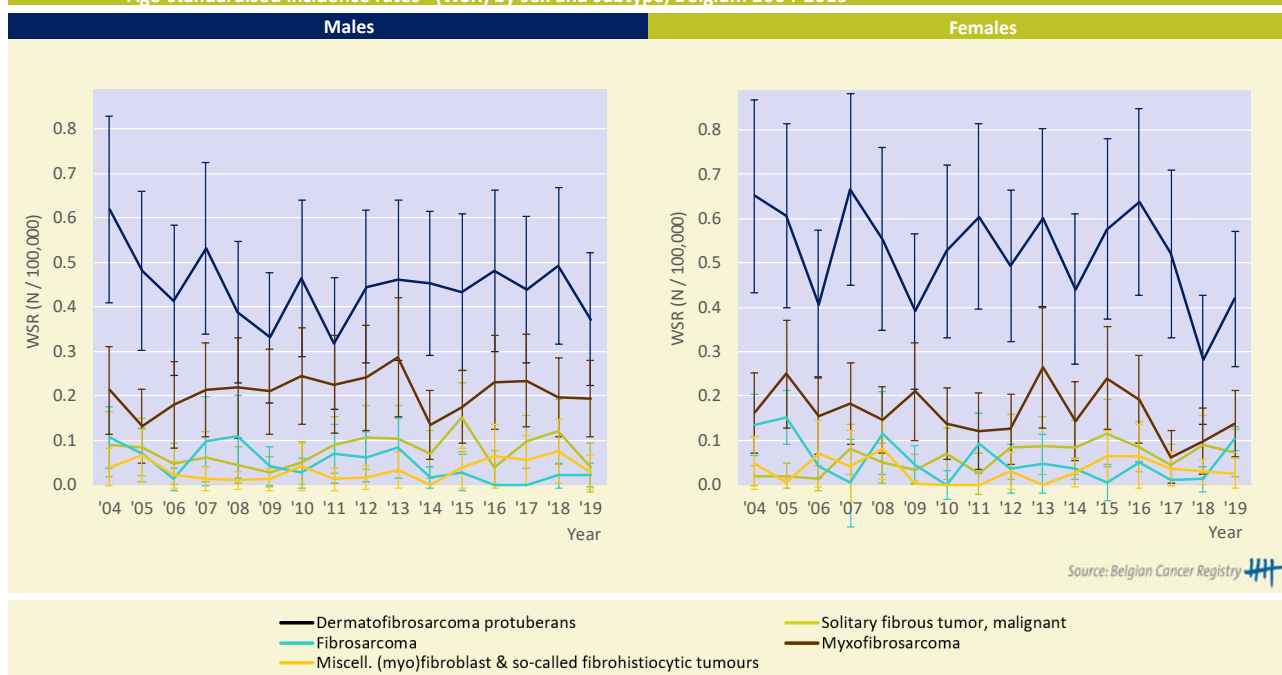
Incidence trends

Figure 5 Malignant (myo)fibroblastic tumours and so-called fibrohistiocytic tumours: Age-standardised incidence rates* (WSR) by age category, males and females, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals.

Figure 6 Malignant (myo)fibroblastic tumours and so-called fibrohistiocytic tumours:
Age-standardised incidence rates* (WSR) by sex and subtype, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals.

Table 2 Malignant (myo)fibroblastic tumours and so-called fibrohistiocytic tumours:
Incidence trends by sex and age category in Belgium, 2004-2019

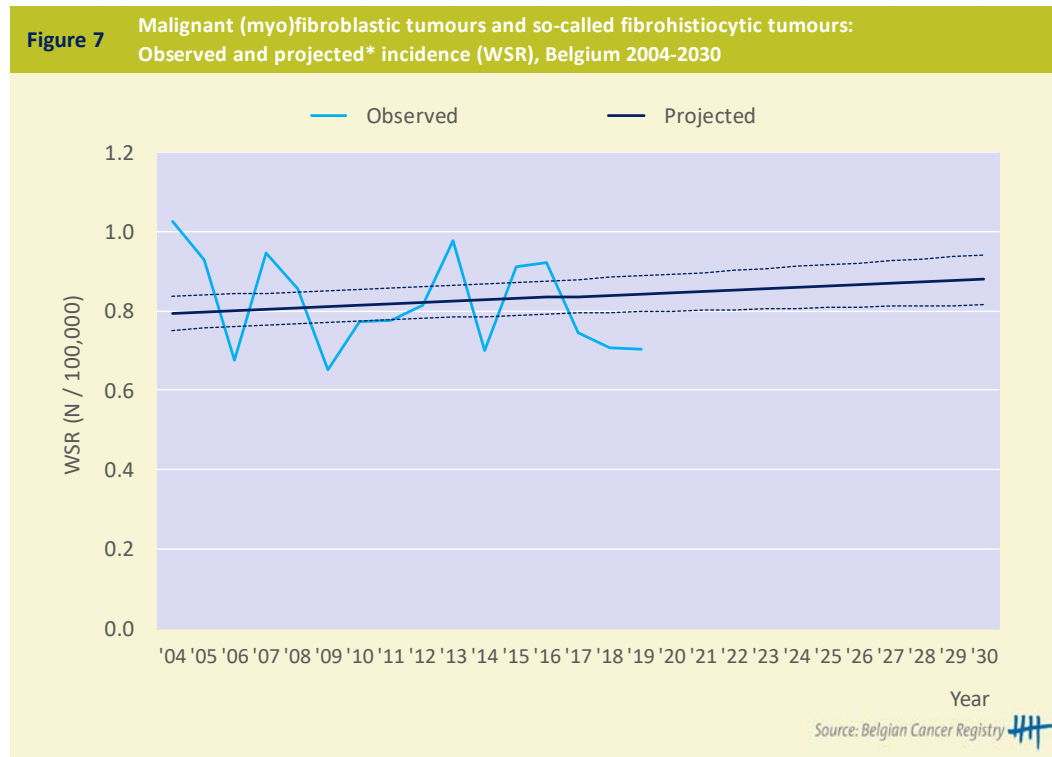
Age category	Males			Females		
	AAPC (%)	95%CI	Period	AAPC (%)	95%CI	Period
All ages	-0.5	[-2.3; 1.3]	2004-2019	-1.7	[-3.9; 0.6]	2004-2019
0 - 14 y	-	-	-	-	-	-
15 - 29 y	-1.0	[-5.6; 3.9]	2004-2019	-9.7	[-12.7; -6.6]	2004-2019
				-5.3	[-10.7; 0.5]	2004-2012
				20.8	[7.4; 36.0]	2012-2016
-46.0	[-55.6; -34.4]	2016-2019				
30 - 59 y	-0.2	[-1.8; 1.4]	2004-2019	-1.6	[-4.7; 1.5]	2004-2019
60+ y	-0.9	[-3.2; 1.5]	2004-2019	-1.6	[-4.1; 1.0]	2004-2019
				-8.4	[-17.0; 1.0]	2004-2008
				2.0	[-1.1; 5.3]	2008-2019

Source: Belgian Cancer Registry

AAPC: average annual percentage change

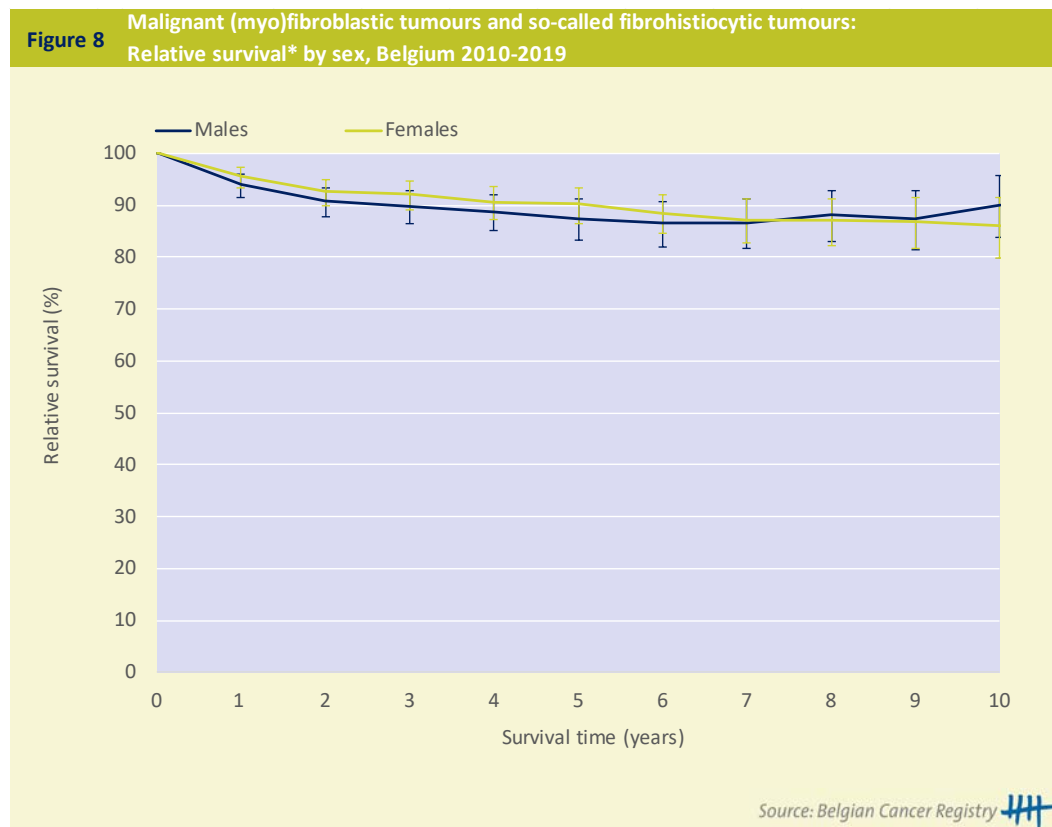
Period: When a joinpoint occurred, APC's are calculated for the period before and after the joinpoint. This column represents the corresponding time interval. AAPC's are always calculated over the entire study-period.

Incidence projections

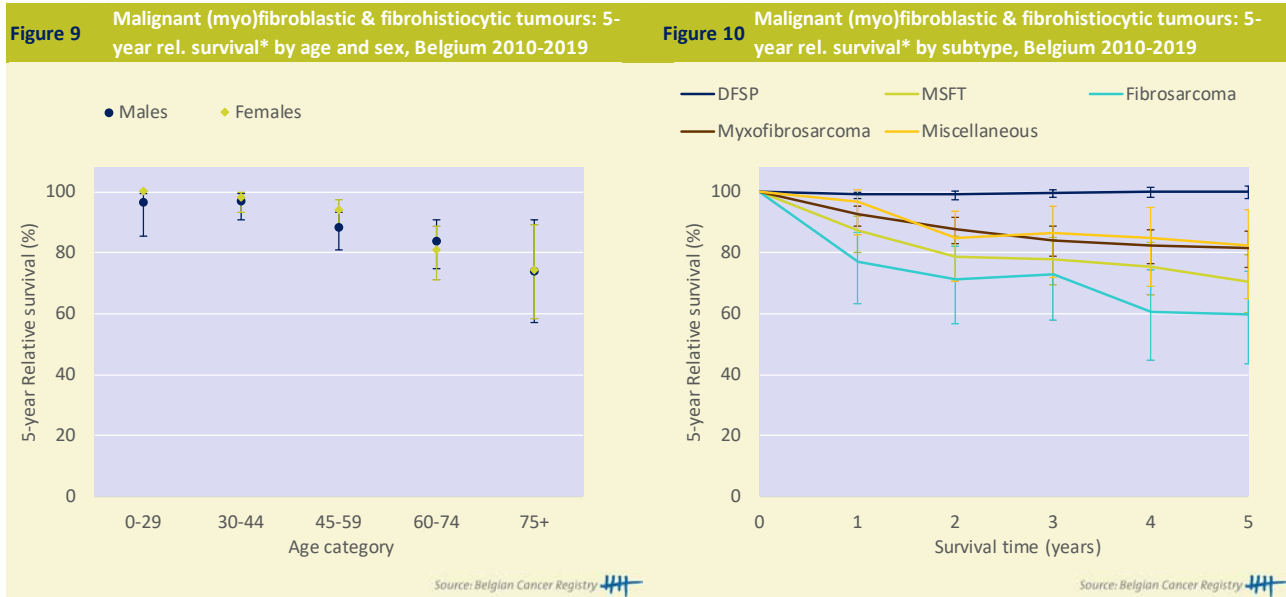


WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

Survival



* The relative survival values are represented with 95% Confidence Intervals



DFSP: Dermatofibrosarcoma protuberans

MSFT: Solitary fibrous tumour, malignant

* The relative survival values are represented with 95% Confidence Intervals

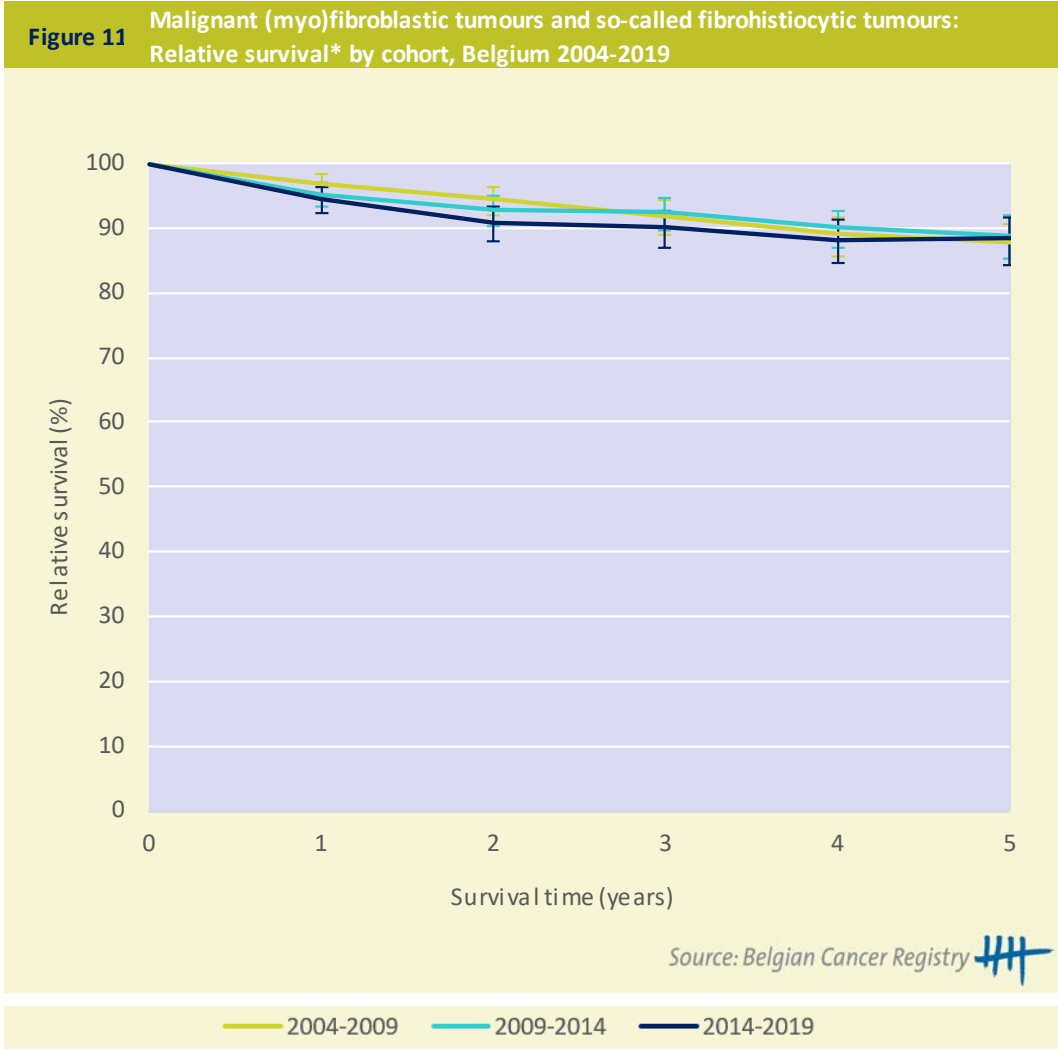
Table 3 Malignant (myo)fibroblastic tumours and so-called fibrohistiocytic tumours: Conditional 5-year relative survival* in Belgium, 2010-2019

X years since diagnosis	N at risk	%
1 year	1,151	92.3
2 year	1,006	94.8
3 year	884	96.3

* Unadjusted 5-yr relative survival probability conditional on surviving the first X years since diagnosis, %

* Interpretation in lay-man's terms: Given that a patient has already survived X years, what is the relative survival probability 5 years later.

Survival trends



* The relative survival values are represented with 95% Confidence Intervals

3.2.2.1 DERMATOFIBROSARCOMA PROTUBERANS

MAIN SUBTYPES:

- *Dermatofibrosarcoma protuberans (DFSP)*
- *Pigmented dermatofibrosarcoma protuberans*
- *Dermatofibrosarcoma protuberans, fibrosarcomatous*

KEYNOTES

Incidence (table 1-2; figure 1-4)

- The age-specific incidence of dermatofibrosarcoma protuberans shows a peak incidence around 40 years.
- Almost half of the dermatofibrosarcoma protuberans originate in the trunk.

Survival (table 3; figure 5-7)

- Dermatofibrosarcoma protuberans patients have the best prognosis compared to all other patients with bone and soft tissue tumours described in this report. The 5-year relative survival remains stable around 100%, regardless of sex or age group. In males, relative survival marginally exceeds 100% which occurs when survival in the patient population slightly exceeds the survival in the general population matched for ages, sex and region. This occurs for some cancers with excellent prognosis (methodology described in section 2.3.5).

Table 1 Dermatofibrosarcoma protuberans: Overview of incidence, prevalence and survival by sex in Belgium, 2010-2019				
Males				
Incidence	N	CR	WSR	
Incidence, 2010-2019	289	0.5	0.4	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	138	2.4	2.0	
10-year prevalence, 31.12.2019	268	4.7	3.6	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	284	102.0	[98.2;104.2]	
10-year relative survival, 2010-2019	284	109.1	[103.8;112.2]	
Females				
Incidence	N	CR	WSR	
Incidence, 2010-2019	329	0.6	0.5	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	151	2.6	2.2	
10-year prevalence, 31.12.2019	313	5.4	4.4	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	328	98.8	[95.7;100.7]	
10-year relative survival, 2010-2019	328	96.4	[89.5;100.6]	
Median age at diagnosis, 2010-2019 (y)	43 [Q1: 33; Q3: 55]			
M/F-ratio	0.9			

Source: Belgian Cancer Registry 

N: number of new diagnoses

CR: crude (all ages) rate (N/100,000 person years)

WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

M/F-ratio: Male/Female ratio based on the age-standardised rates (2010-2019)

Incidence

Figure 1 Dermatofibrosarcoma protuberans:
Age-specific incidence rates (N/100,000) by sex, Belgium 2004-2019

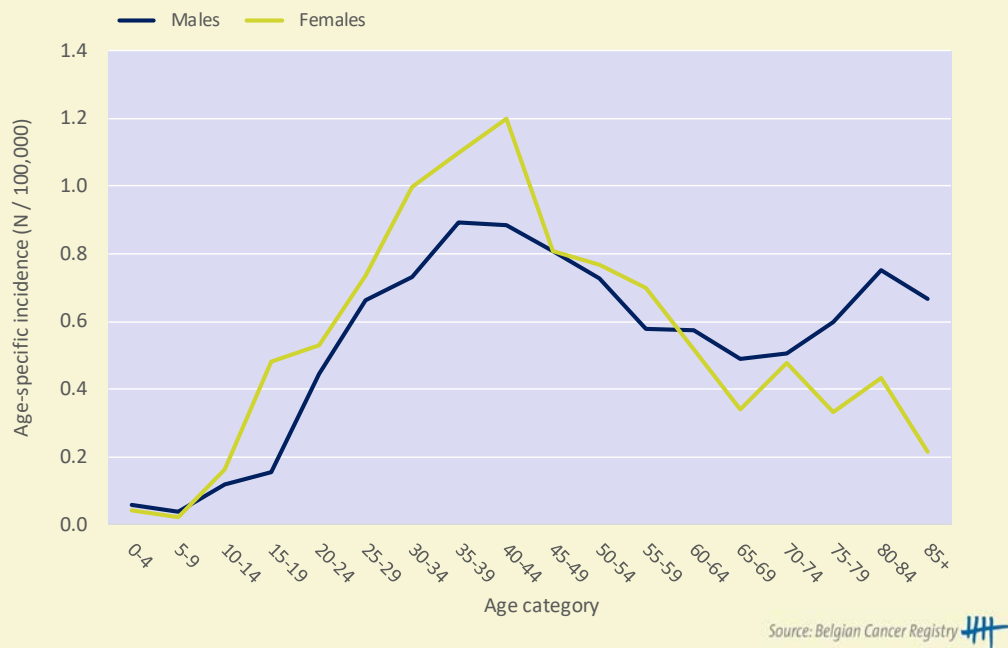
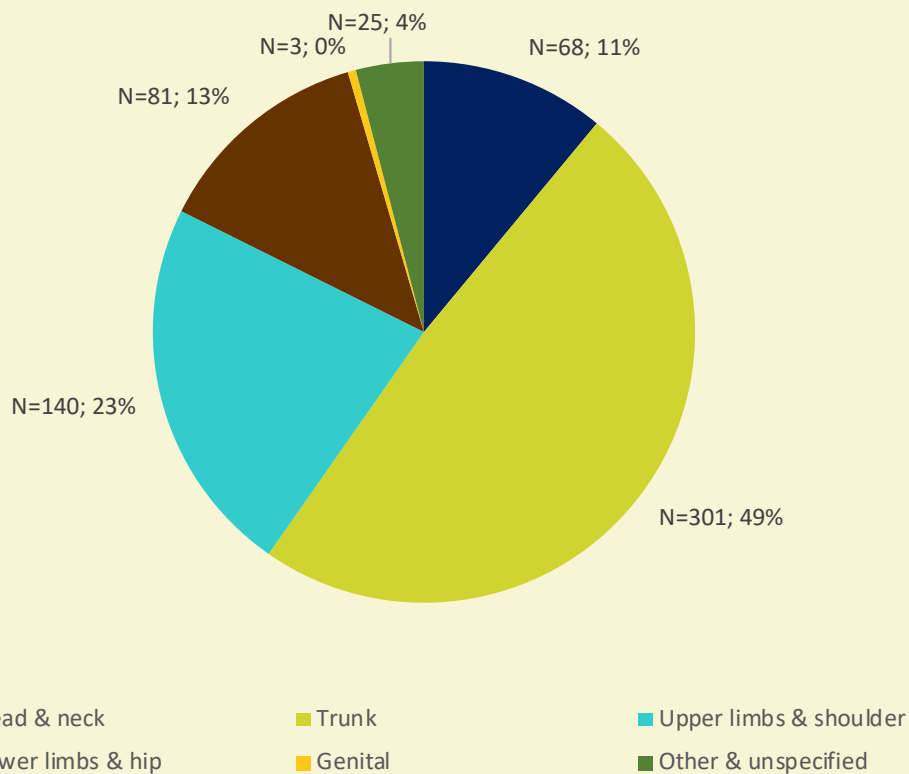
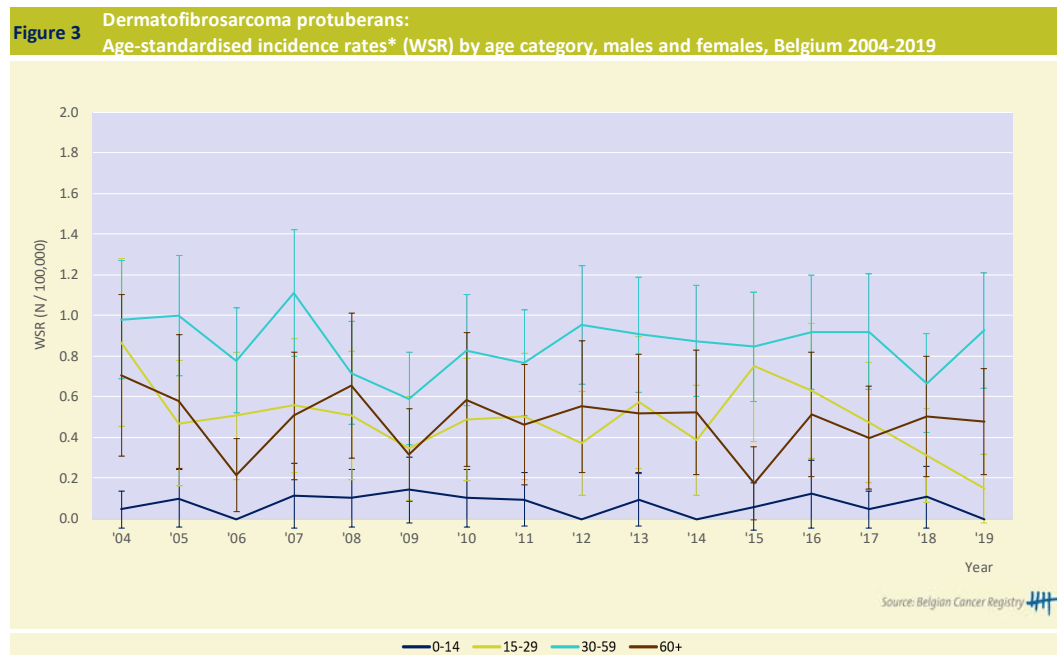


Figure 2 Dermatofibrosarcoma protuberans:
Incidence distribution by primary tumour location, Belgium 2010-2019



Incidence trends



* The age-standardised incidence rates are represented with 95% Confidence Intervals

Table 2 Dermatofibrosarcoma protuberans:
Incidence trends by sex and age category in Belgium, 2004-2019

Age category	Males			Females		
	AAPC (%)	95%CI	Period	AAPC (%)	95%CI	Period
All ages	-1.3	[-3.0; 0.4]	2004-2019	-1.9	[-4.4; 0.6]	2004-2019
	-7.3	[-12.6; -1.7]	2004-2009			
	1.8	[-0.9; 4.6]	2009-2019			
0 - 14 y	-	-	-	-	-	-
15 - 29 y	-2.4	[-6.7; 2.1]	2004-2019	-10.0	[-15.2; -4.4]	2004-2019
				1.1	[-5.7; 8.3]	2004-2016
30 - 59 y	-0.4	[-2.3; 1.5]	2004-2019	-43.3	[-59.3; -21.1]	2016-2019
				-1.4	[-5.3; 2.6]	2004-2019
				-7.5	[-13.2; -1.4]	2004-2009
60+ y	3.3	[0.3; 6.3]	2009-2019	-1.2	[-5.3; 3.1]	2004-2019

Source: Belgian Cancer Registry

AAPC: average annual percentage change

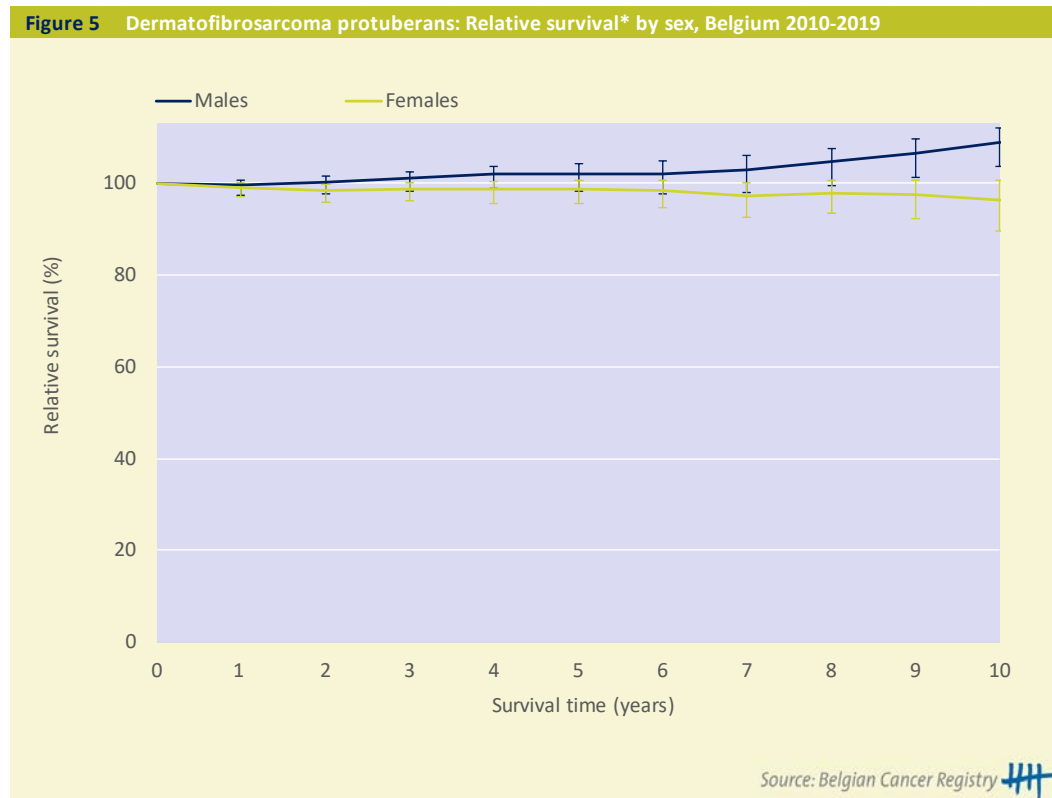
Period: When a joinpoint occurred, APC's are calculated for the period before and after the joinpoint. This column represents the corresponding time interval. AAPC's are always calculated over the entire study-period.

Incidence projections



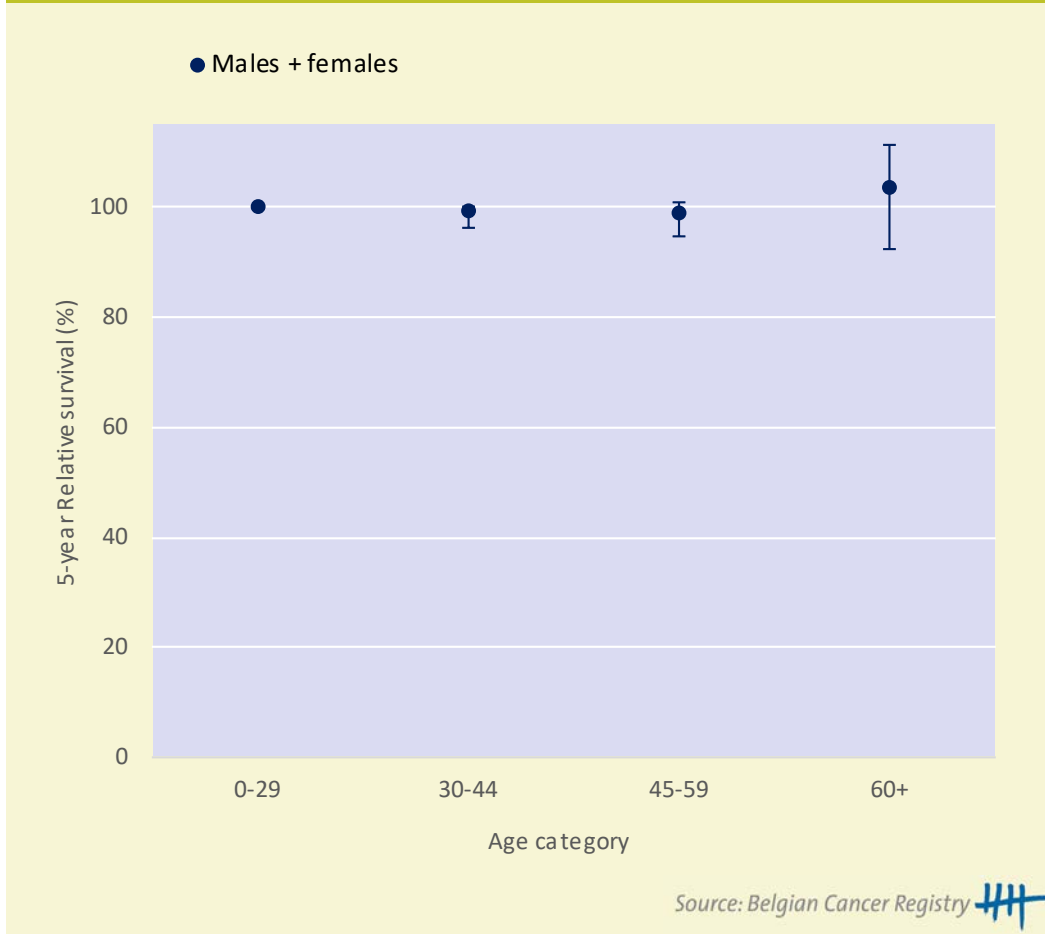
WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

Survival



* The relative survival values are represented with 95% Confidence Intervals

Figure 6 Dermatofibrosarcoma protuberans: 5-year relative survival* by age, Belgium 2010-2019



* The relative survival values are represented with 95% Confidence Intervals

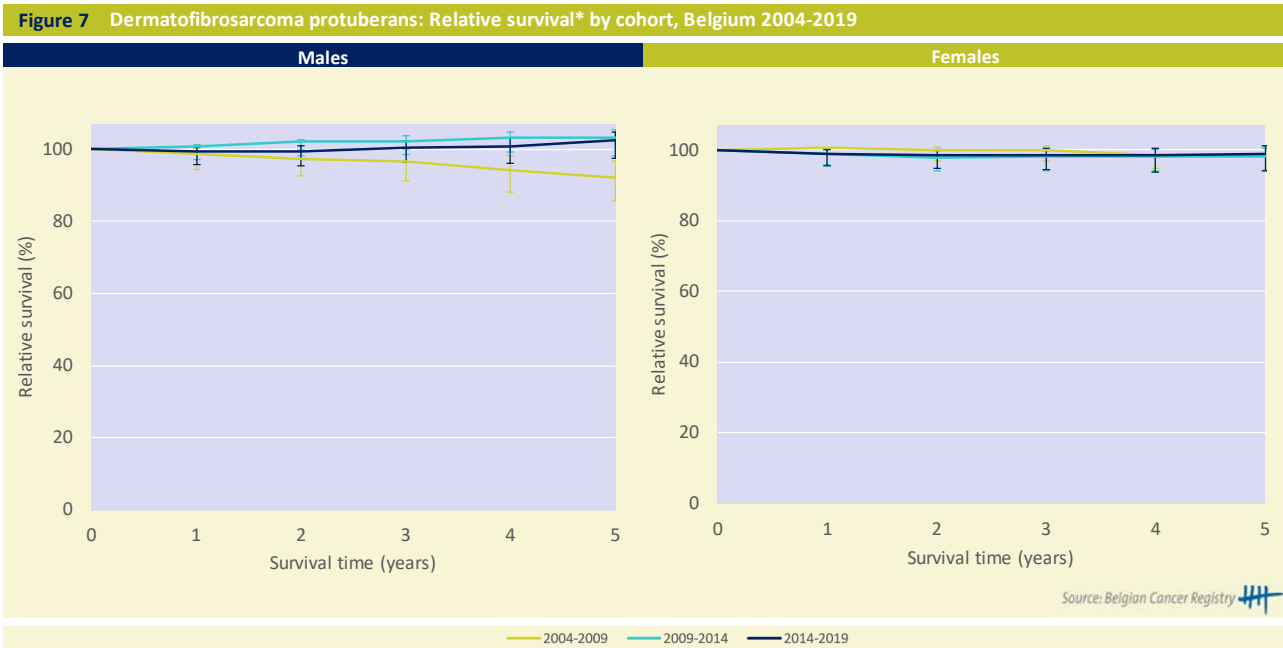
Table 3 Dermatofibrosarcoma protuberans: Conditional 5-year relative survival* in Belgium, 2010-2019

X years since diagnosis	N at risk	%
1 year	602	100.7
2 year	546	100.5
3 year	503	101.1

* Unadjusted 5-yr relative survival probability conditional on surviving the first X years since diagnosis, %

* Interpretation in lay-man's terms: Given that a patient has already survived X years, what is the relative survival probability 5 years later.

Survival trends



* The relative survival values are represented with 95% Confidence Intervals

3.2.2.2 SOLITARY FIBROUS TUMOUR

KEYNOTES

Incidence (table 1-2; figure 1-4)

- Malignant solitary fibrous tumours are most often diagnosed in the older population (very rarely under the age of 40 years).
- These tumours can occur at any anatomical site, most frequently in subcutaneous connective and other soft tissues or in visceral organs (including lung, heart etc.).
- Between 2004 and 2019, an increasing trend is observed in females (average annual percentage change of 9.9%).
- Over time, coding and differentiation of malignant and non-malignant tumours evolved. Data include those tumours considered and reported as malignant at the time of diagnosis. Based on pathology, the biological behaviour of solitary fibrous tumours cannot be predicted. They can metastasize (even if it cannot be labelled “malignant”) and thus show its malignant character.

Survival (table 3; figure 5-7)

- The 5-year relative survival of these patients decreases with age: from 89% in patients younger than 60 years old to less than 60% in patients of 60+.
- The 10-year relative survival in males (75%) is better than in females (62%).
- No consistent improvement in survival is observed over time.

Solitary fibrous tumour: Overview of incidence, prevalence and survival by sex in Belgium, 2010-2019				
Males				
Incidence	N	CR	WSR	
Incidence, 2010-2019	83	0.2	0.1	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	30	0.5	0.3	
10-year prevalence, 31.12.2019	51	0.9	0.5	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	81	73.1	[58.7;85.1]	
10-year relative survival, 2010-2019	81	74.6	[54.5;92.3]	
Females				
Incidence	N	CR	WSR	
Incidence, 2010-2019	75	0.1	0.1	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	34	0.6	0.3	
10-year prevalence, 31.12.2019	48	0.8	0.5	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	73	67.9	[53.0;80.0]	
10-year relative survival, 2010-2019	73	61.8	[42.7;78.4]	
Median age at diagnosis, 2010-2019 (y)	66 [Q1: 53; Q3: 75]			
M/F-ratio	1.1			

Source: Belgian Cancer Registry 

N: number of new diagnoses

CR: crude (all ages) rate (N/100,000 person years)

WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

M/F-ratio: Male/Female ratio based on the age-standardised rates (2010-2019)

Incidence

Figure 1 Solitary fibrous tumour: Age-specific incidence rates (N/100,000) by sex, Belgium 2004-2019

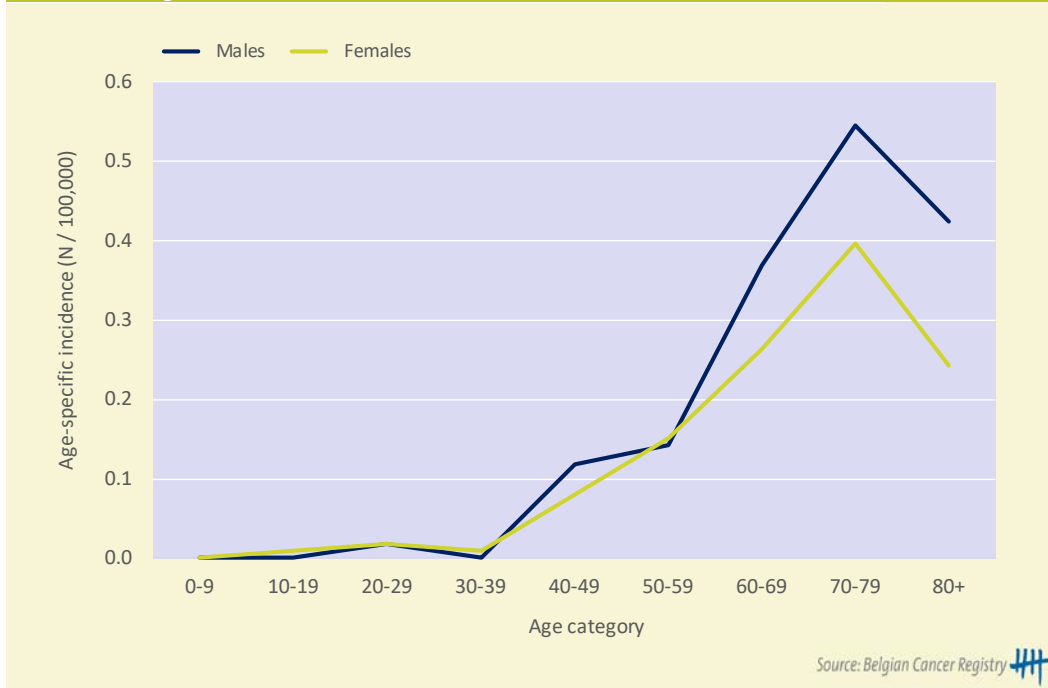
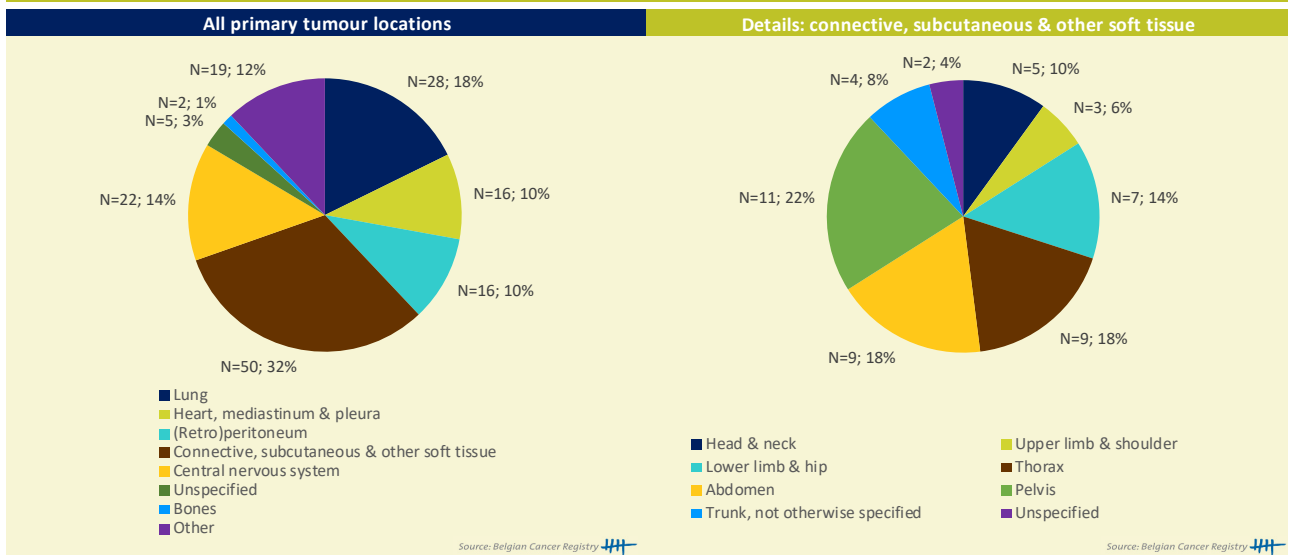
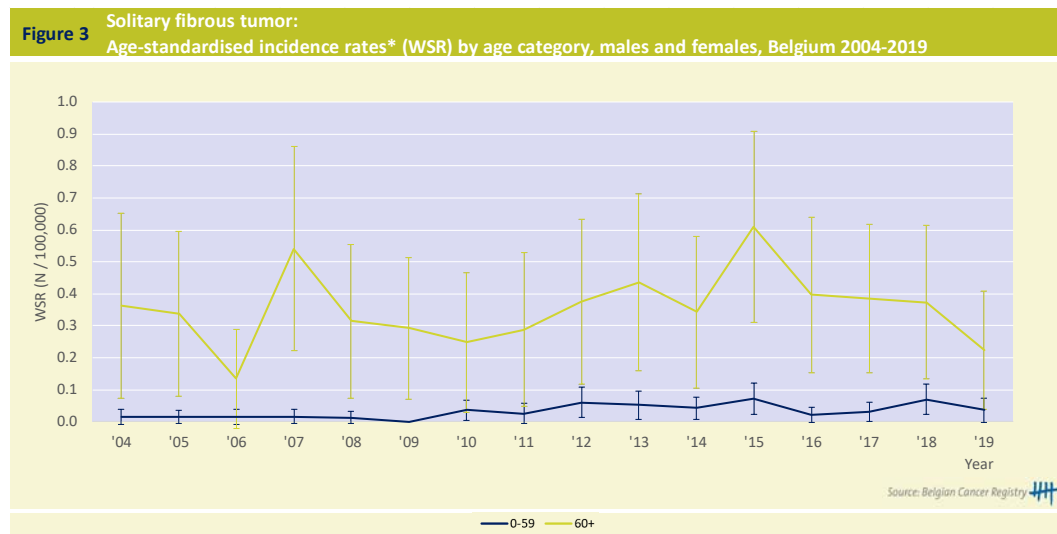


Figure 2 Solitary fibrous tumour: Incidence distribution by primary tumour location, Belgium 2010-2019



Incidence trends



* The age-standardised incidence rates are represented with 95% Confidence Intervals

Table 2 Solitary fibrous tumour: Incidence trends by sex in Belgium, 2004-2019

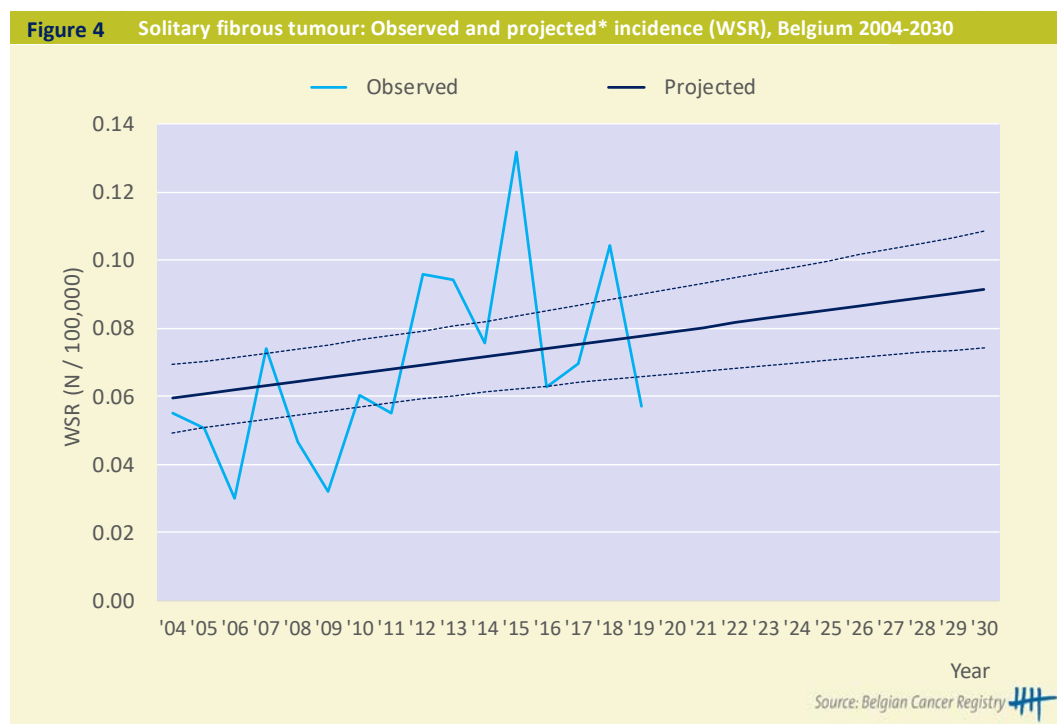
Age category	Males			Females		
	AAPC (%)	95%CI	Period	AAPC (%)	95%CI	Period
All ages	-2.8	[-8.2; 2.8]	2004-2019	9.9	[3.7; 16.5]	2004-2019
	-16.6	[-30.7; 0.5]	2004-2009			
	33.6	[9.1; 63.5]	2009-2013			
	-10.8	[-23.1; 3.4]	2013-2019			

Source: Belgian Cancer Registry

AAPC: average annual percentage change.

Period: When a joinpoint occurred, APC's are calculated for the period before and after the joinpoint. This column represents the corresponding time interval. AAPC's are always calculated over the entire study-period.

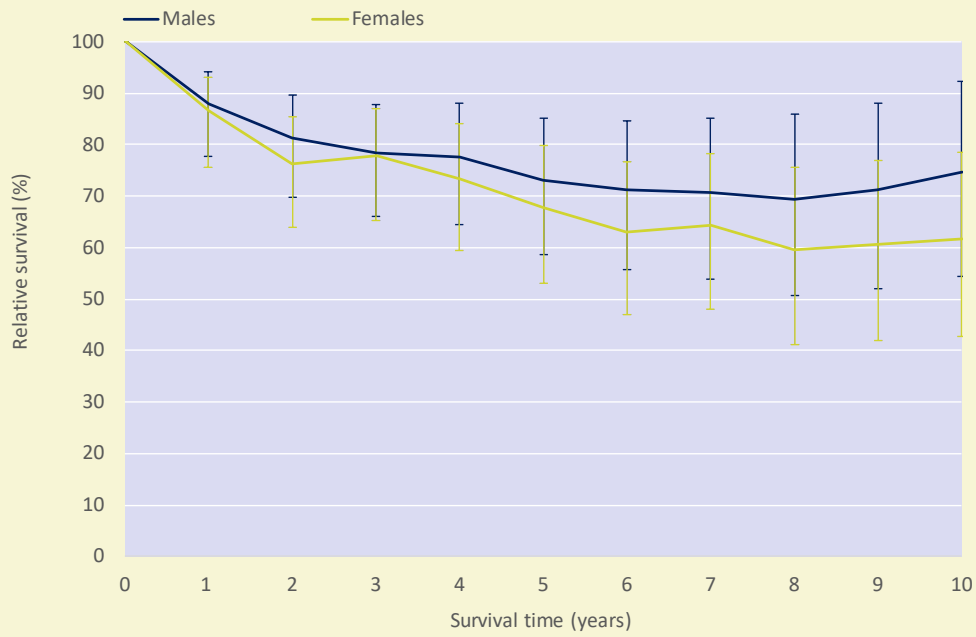
Incidence projections



WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

Survival

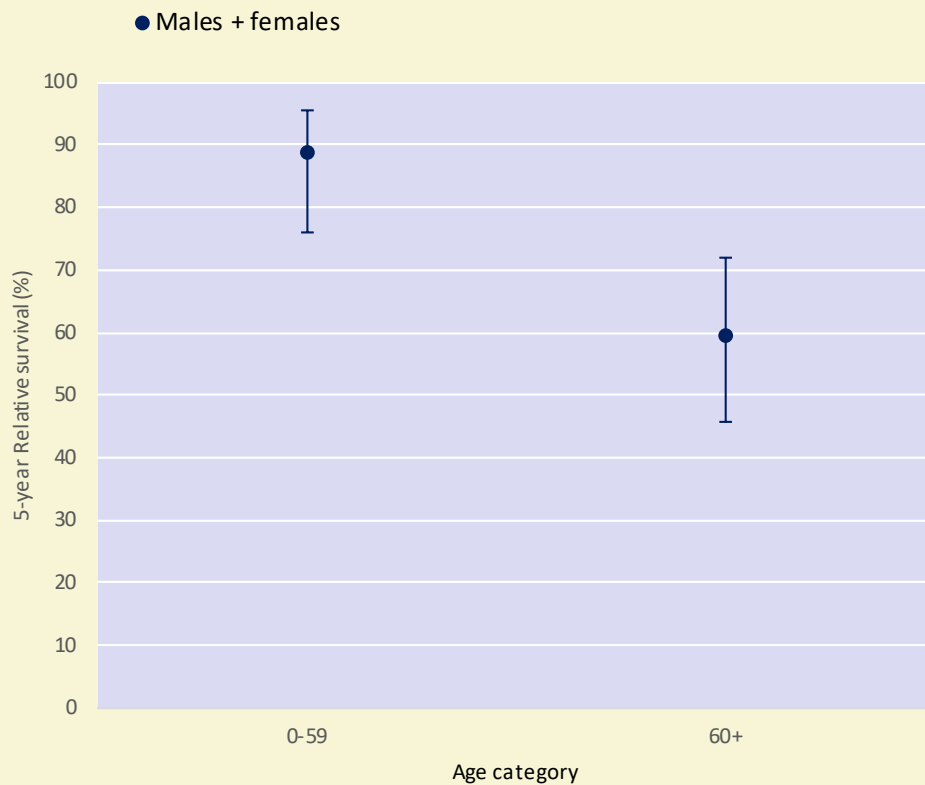
Figure 5 Solitary fibrous tumour: Relative survival* by sex, Belgium 2010-2019



Source: Belgian Cancer Registry

* The relative survival values are represented with 95% Confidence Intervals

Figure 6 Solitary fibrous tumour: 5-year relative survival* by age, Belgium 2010-2019



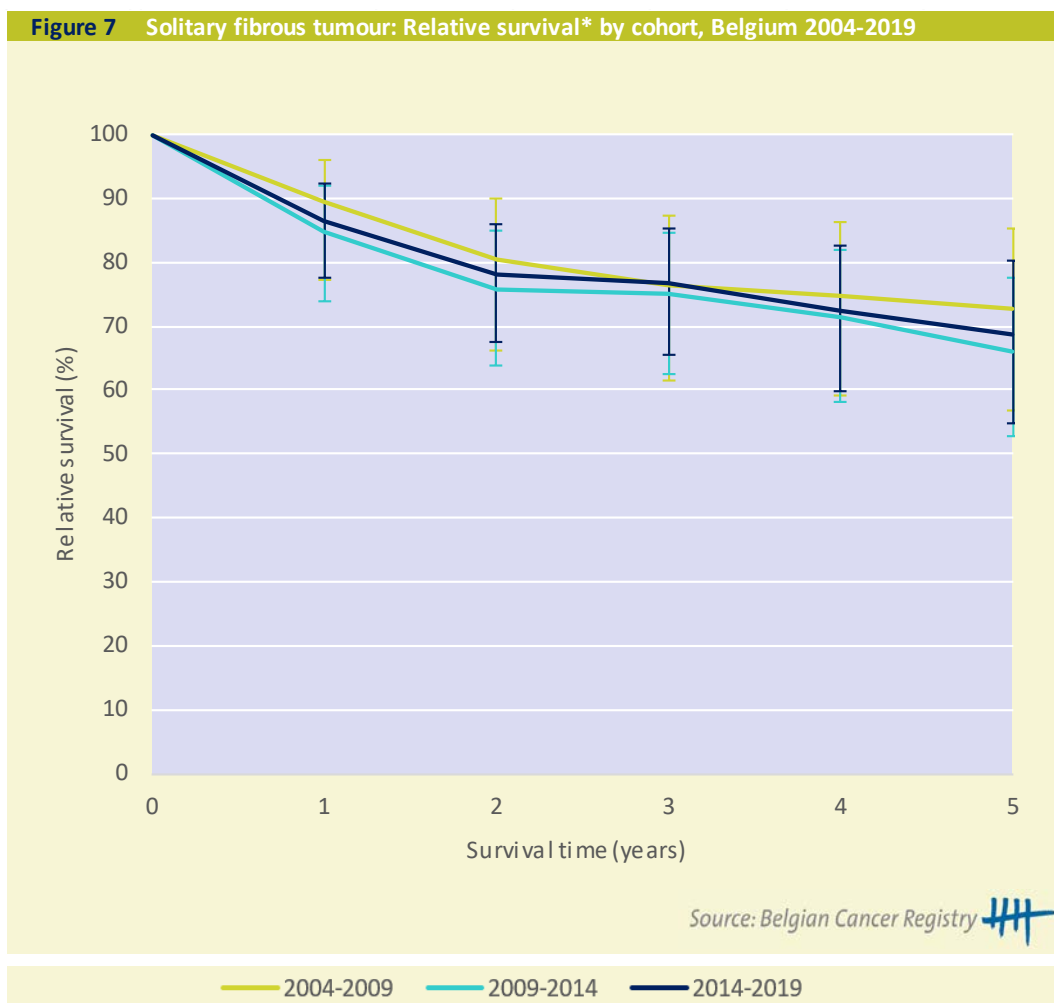
Source: Belgian Cancer Registry

* The relative survival values are represented with 95% Confidence Intervals

Solitary fibrous tumour: Conditional 5-year relative survival* in Belgium, 2010-2019		
X years since diagnosis	N at risk	%
1 year	131	77.4
2 year	110	85.7
3 year	93	83.2

* Unadjusted 5-yr relative survival probability conditional on surviving the first X years since diagnosis, %
 * Interpretation in lay-man's terms: Given that a patient has already survived X years, what is the relative survival probability 5 years later.

Survival trends



* The relative survival values are represented with 95% Confidence Intervals

3.2.2.3 FIBROSARCOMA

MAIN SUBTYPES:

- *Adult fibrosarcoma*
- *Infantile fibrosarcoma*

KEYNOTES

Incidence (table 1-2; figure 1-4)

- Fibrosarcoma is most often diagnosed in the older population with increasing incidence from 30 years onwards and a median age at diagnosis of 62 years. A small incidence peak is observed in young children corresponding to infantile fibrosarcoma which is very rare and diagnosed in the first year of life (only 6 diagnoses in 2004-2019).
- More than half of the fibrosarcoma occur in abdomen, pelvis, trunk and thorax.
- The decreasing incidence of fibrosarcoma between 2004 and 2019 (average annual percentage change of -9.5%), especially in the patients older than 60 years at diagnosis, may be partly explained by classification changes and better registration.
- Based on the incidence projections, the incidence rate (WSR) is expected to remain stable in the nearby future.

Survival (table 3; figure 5-7)

- The 5-year relative survival of fibrosarcoma patients is better in females (74%) than in males (56%). Remark: the 5-year relative survival for infantile fibrosarcoma patients (N=6 only) is 100%.
- The apparent decrease of the 5-year relative survival rate in the period 2004-2009 compared to the period 2010-2019 may be partly due to a more accurate diagnosis and registration of sarcomas.

Males				
	N	CR	WSR	
Incidence				
Incidence, 2010-2019	29	0.05	0.03	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	3	0.05	0.03	
10-year prevalence, 31.12.2019	13	0.23	0.14	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2004-2019	61	55.6	[40.4;69.5]	
10-year relative survival, 2004-2019	61	62.3	[43.9;79.6]	
Females				
Incidence	N	CR	WSR	
Incidence, 2010-2019	29	0.1	0.0	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	8	0.1	0.2	
10-year prevalence, 31.12.2019	19	0.3	0.3	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2004-2019	69	74.4	[59.6;86.0]	
10-year relative survival, 2004-2019	69	71.4	[54.8;85.5]	
Median age at diagnosis, 2010-2019 (y)	62 [Q1: 48; Q3: 76]			
M/F-ratio	0.9			

Source: Belgian Cancer Registry 

N: number of new diagnoses

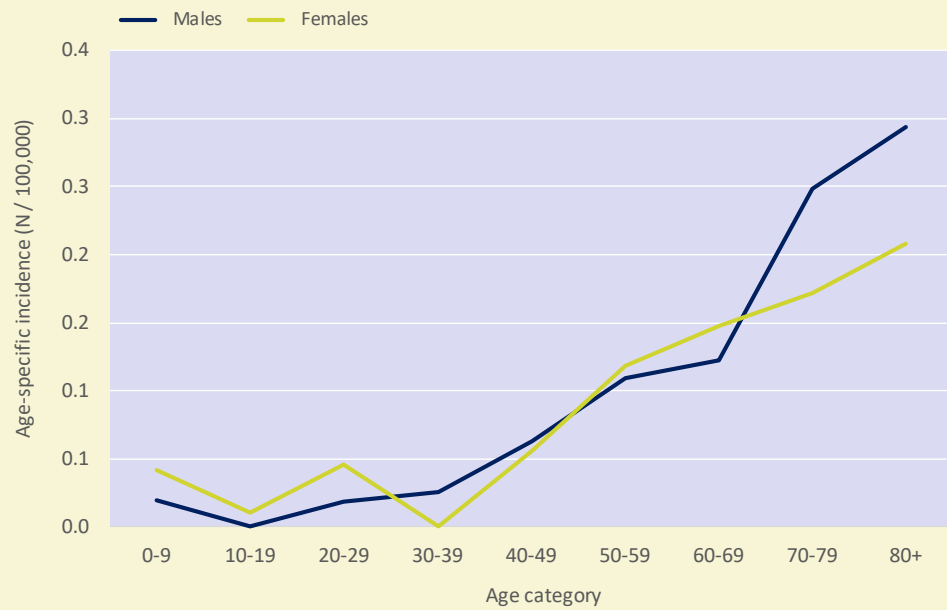
CR: crude (all ages) rate (N/100,000 person years)

WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

M/F-ratio: Male/Female ratio based on the age-standardised rates (2010-2019)

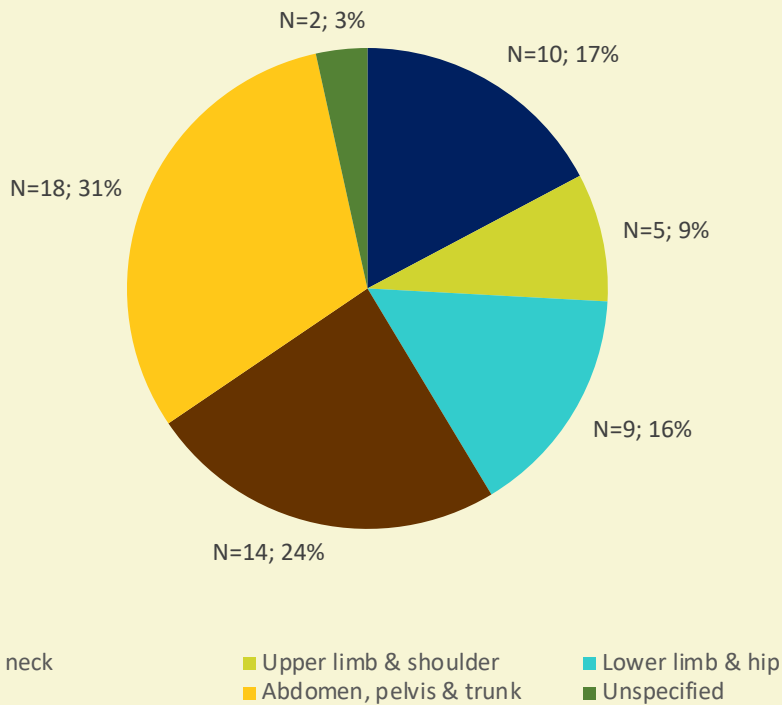
Incidence

Figure 1 Fibrosarcoma: Age-specific incidence rates (N/100,000) by sex, Belgium 2004-2019



Source: Belgian Cancer Registry

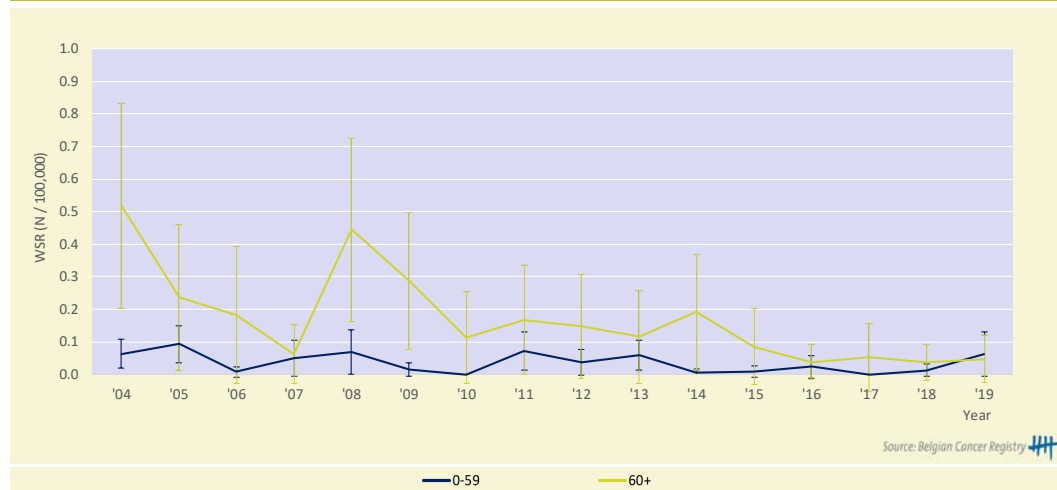
Figure 2 Fibrosarcoma: Incidence distribution by primary tumour location, Belgium 2010-2019



Source: Belgian Cancer Registry

Incidence trends

Figure 3 Fibrosarcoma: Age-standardised incidence rates* (WSR) by age category, males and females, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals

Table 2 Fibrosarcoma: Incidence trends in Belgium, 2004-2019

Males + females		
AAPC (%)	95%CI	Period
-9.5	[-17.0; -1.3]	2004-2019

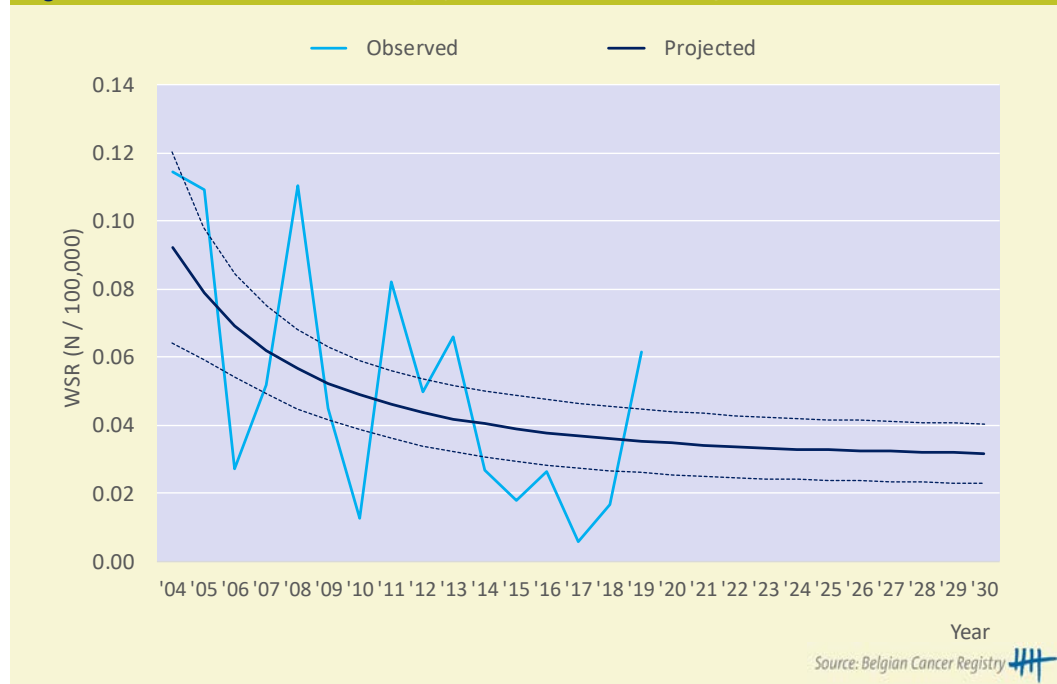
Source: Belgian Cancer Registry

AAPC: average annual percentage change

Period: When a joinpoint occurred, APC's are calculated for the period before and after the joinpoint. This column represents the corresponding time interval. AAPC's are always calculated over the entire study-period.

Incidence projections

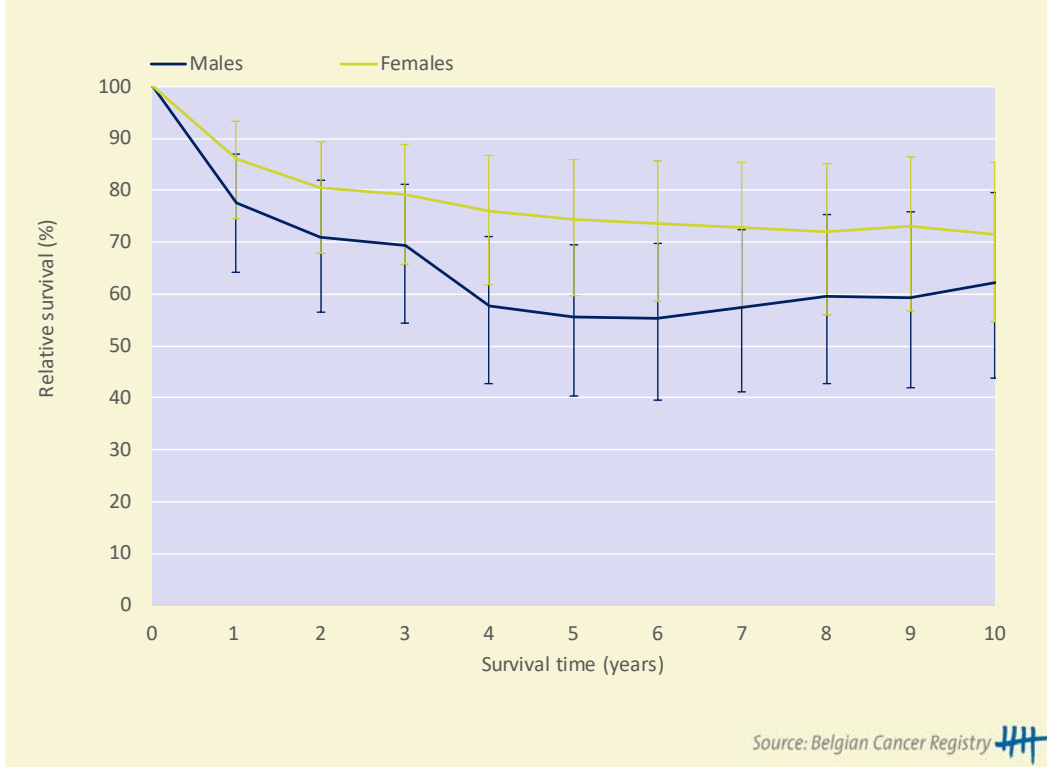
Figure 4 Fibrosarcoma: Observed and projected* incidence (WSR), Belgium 2004-2030



WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

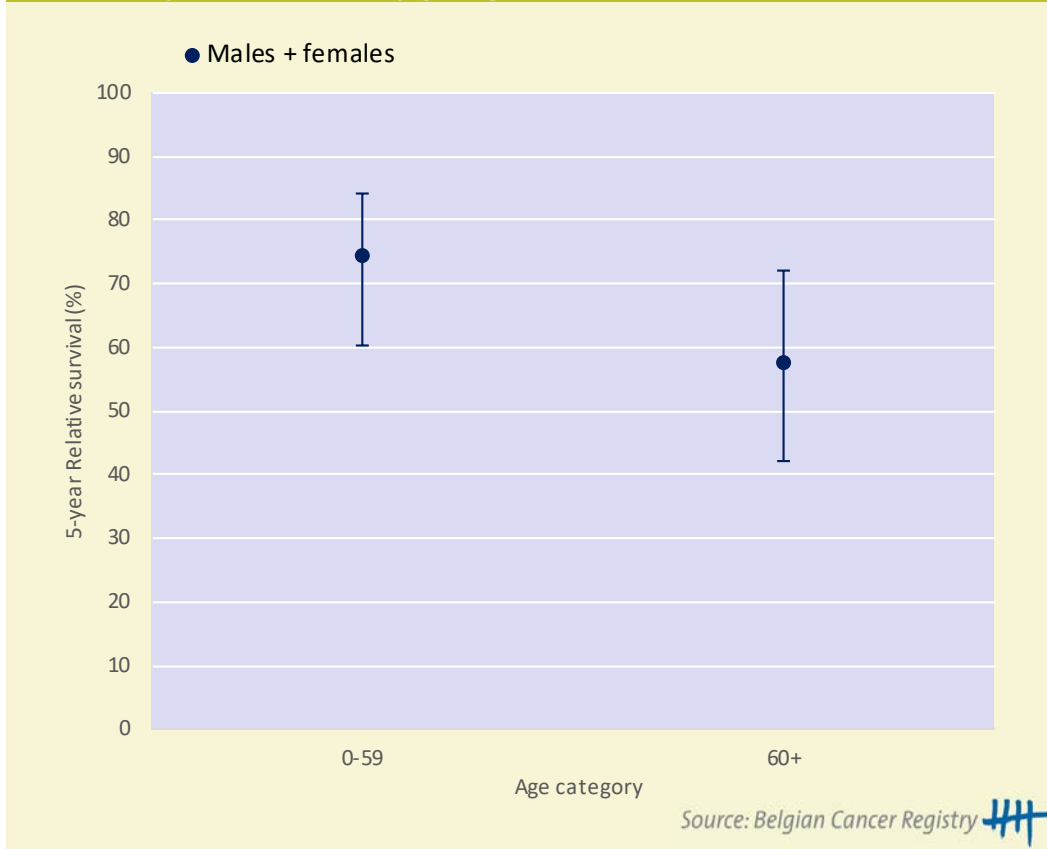
Survival

Figure 5 Fibrosarcoma: Relative survival* by sex, Belgium 2004-2019



* The relative survival values are represented with 95% Confidence Intervals

Figure 6 Fibrosarcoma: 5-year relative survival* by age, Belgium 2004-2019

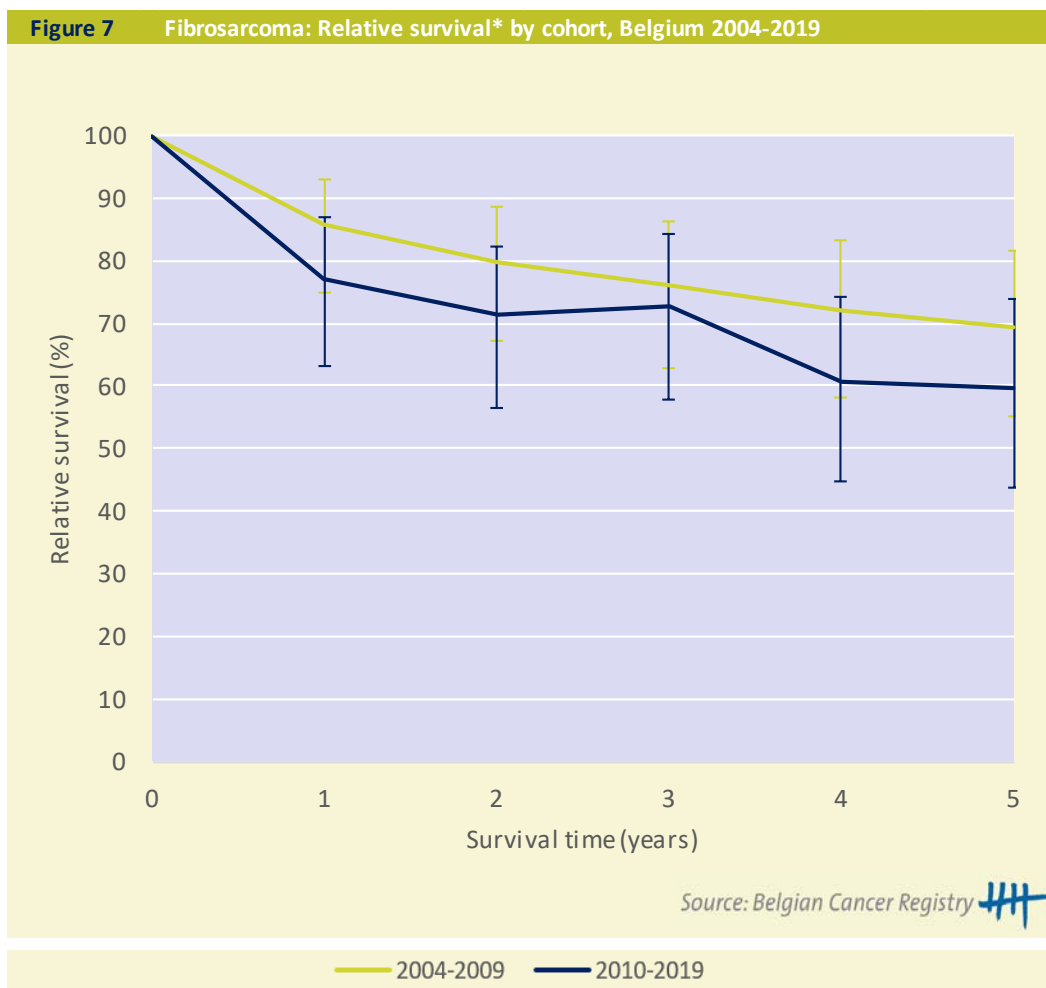


* The relative survival values are represented with 95% Confidence Intervals

Fibrosarcoma: Conditional 5-year relative survival* in Belgium, 2004-2019		
X years since diagnosis	N at risk	%
1 year	103	79.0
2 year	89	86.2
3 year	83	88.8

* Unadjusted 5-yr relative survival probability conditional on surviving the first X years since diagnosis, %
 * Interpretation in lay-man's terms: Given that a patient has already survived X years, what is the relative survival probability 5 years later.

Survival trends



* The relative survival values are represented with 95% Confidence Intervals

3.2.2.4 MYXOFIBROSARCOMA

KEYNOTES

Incidence (table 1-2; figure 1-4)

- Myxofibrosarcoma occurs more frequently in the older population (rarely under the age of 40 years) with a peak incidence above 80 years.
- Myxofibrosarcomas are more common in males than in females (M/F-ratio of 1.4), especially in the older population.
- Almost half of the myxofibrosarcomas occur in the lower limbs and hip.

Survival (table 3; figure 5-7)

- The 5-year relative survival of myxofibrosarcoma patients remains stable across age groups (i.e. younger versus older than 60 years), and lies around 80%.
- The 10-year relative survival does not seem to differ between males and females, and equals circa 75%.
- The apparent decrease of the 5-year relative survival rate in the period 2014-2019 compared to the period 2004-2014 may be partly due to a more accurate diagnosis and registration of sarcomas.

Table 1 Myxofibrosarcoma: Overview of incidence, prevalence and survival by sex in Belgium, 2010-2019				
Males				
Incidence	N	CR	WSR	
Incidence, 2010-2019	211	0.4	0.2	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	81	1.4	0.7	
10-year prevalence, 31.12.2019	136	2.4	1.3	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	210	78.2	[69.1;86.2]	
10-year relative survival, 2010-2019	210	75.3	[61.5;87.9]	
Females				
Incidence	N	CR	WSR	
Incidence, 2010-2019	154	0.3	0.2	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	62	1.1	0.6	
10-year prevalence, 31.12.2019	112	1.9	1.0	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	154	86.0	[76.5;93.6]	
10-year relative survival, 2010-2019	154	75.4	[57.8;90.8]	
Median age at diagnosis, 2010-2019 (y)	67 [Q1: 56; Q3: 78]			
M/F-ratio	1.4			

Source: Belgian Cancer Registry 

N: number of new diagnoses

CR: crude (all ages) rate (N/100,000 person years)

WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

M/F-ratio: Male/Female ratio based on the age-standardised rates (2010-2019)

Incidence

Figure 1 Myxofibrosarcoma: Age-specific incidence rates (N/100,000) by sex, Belgium 2004-2019

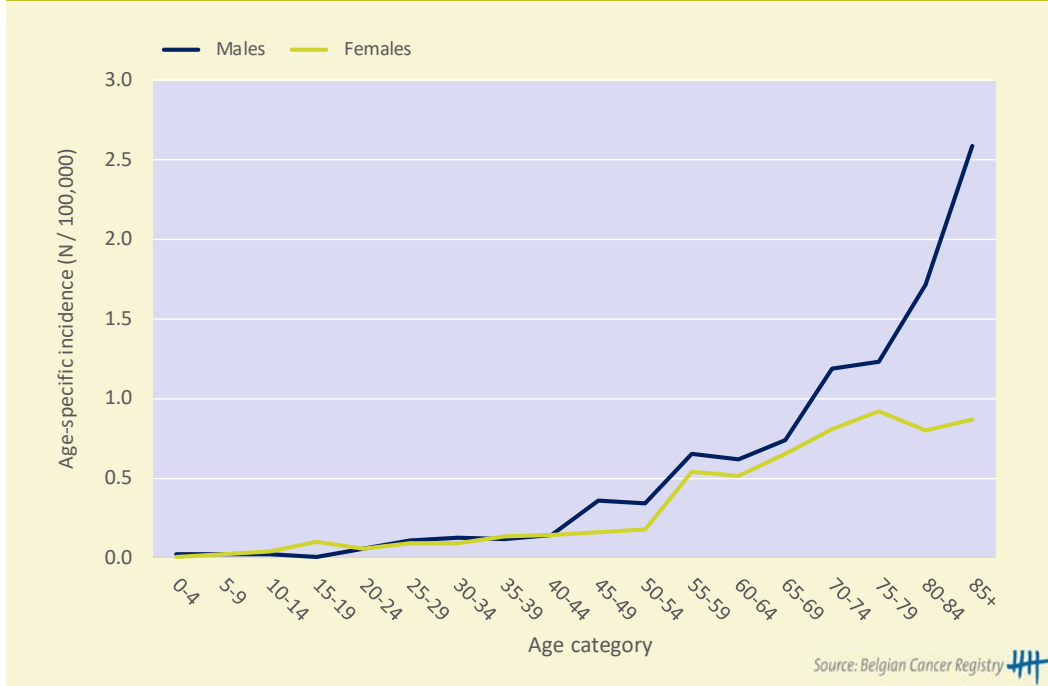
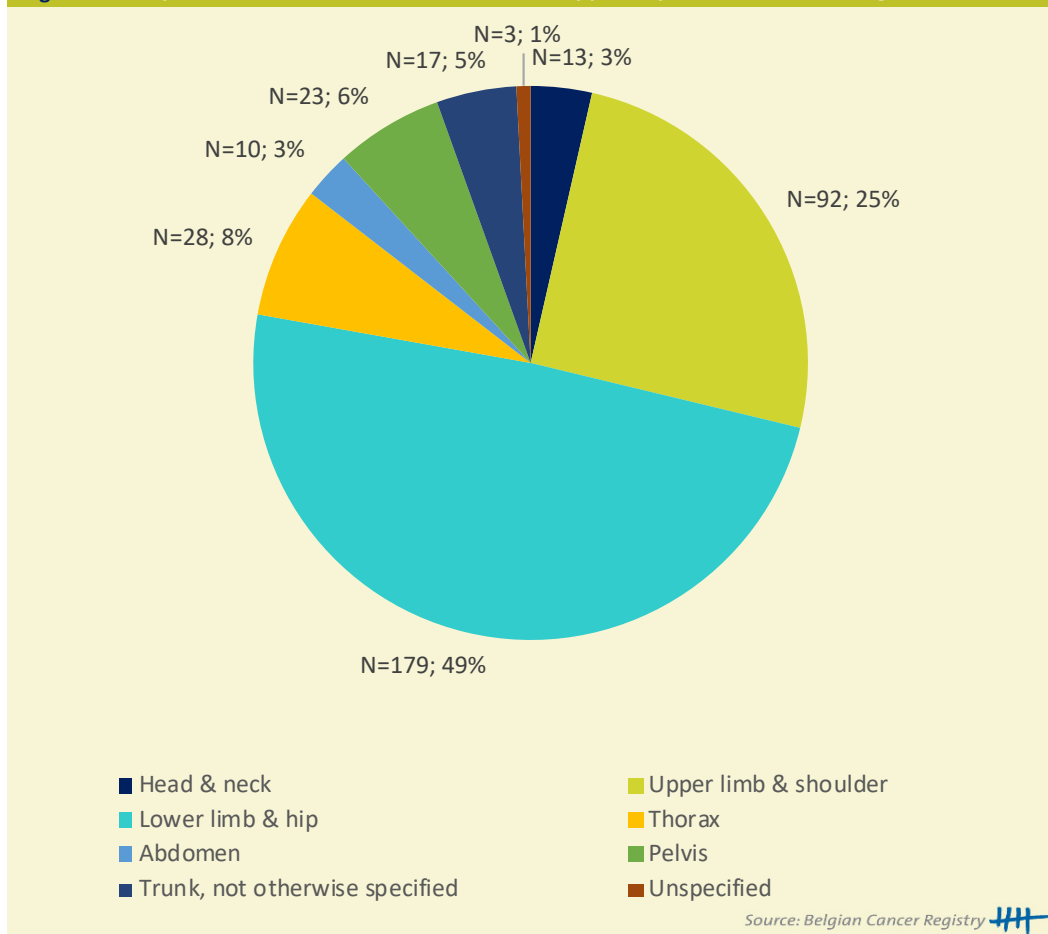
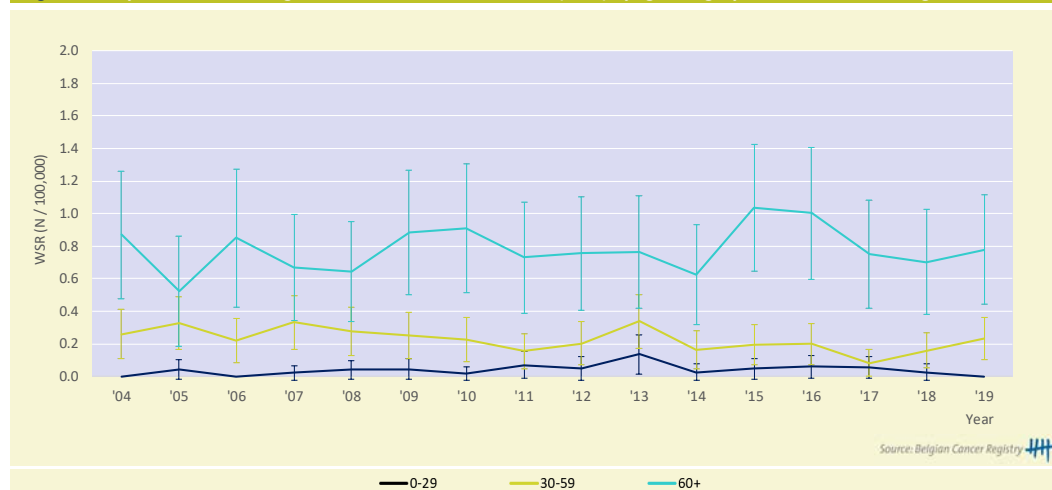


Figure 2 Myxofibrosarcoma: Incidence distribution by primary tumour location, Belgium 2010-2019



Incidence trends

Figure 3 Myxofibrosarcoma: Age-standardised incidence rates* (WSR) by age category, males & females, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals

Table 2 Myxofibrosarcoma: Incidence trends by sex and age category in Belgium, 2004-2019

Age category	Males			Females		
	AAPC (%)	95%CI	Period	AAPC (%)	95%CI	Period
All ages	0.6	[-1.8; 3.1]	2004-2019	-3.0	[-6.9; 1.0]	2004-2019
0 - 29 yrs	-	-	-	-	-	-
30 - 59 yrs	-2.4	[-5.5; 0.8]	2004-2019	-6.2	[-12.5; 0.6]	2004-2019
60+ yrs	2.8	[-0.9; 6.5]	2004-2019	-2.6	[-7.1; 2.0]	2004-2019

Source: Belgian Cancer Registry

AAPC: average annual percentage change

Period: When a joinpoint occurred, APC's are calculated for the period before and after the joinpoint. This column represents the corresponding time interval. AAPC's are always calculated over the entire study-period.

Incidence projections

Figure 4 Myxofibrosarcoma: Observed and projected* incidence (WSR), Belgium 2004-2030



WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

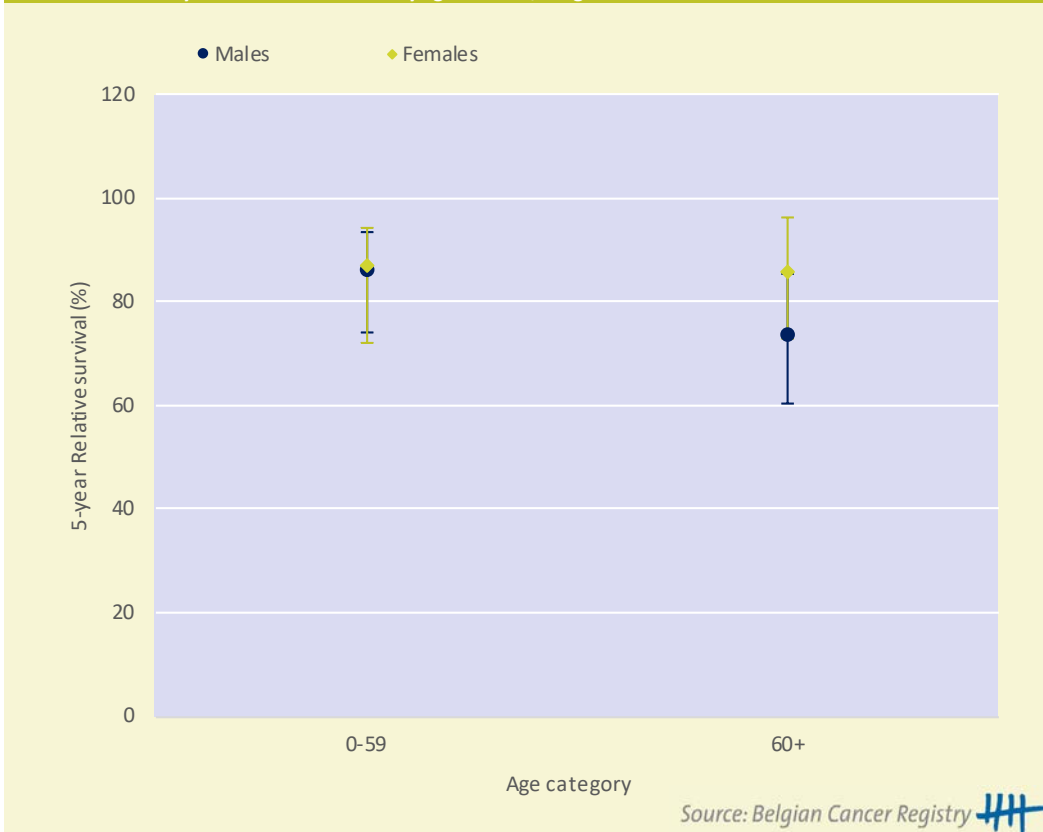
Survival

Figure 5 Myxofibrosarcoma: Relative survival* by sex, Belgium 2010-2019



* The relative survival values are represented with 95% Confidence Intervals

Figure 6 Myxofibrosarcoma: 5-year relative survival* by age and sex, Belgium 2010-2019



* The relative survival values are represented with 95% Confidence Intervals

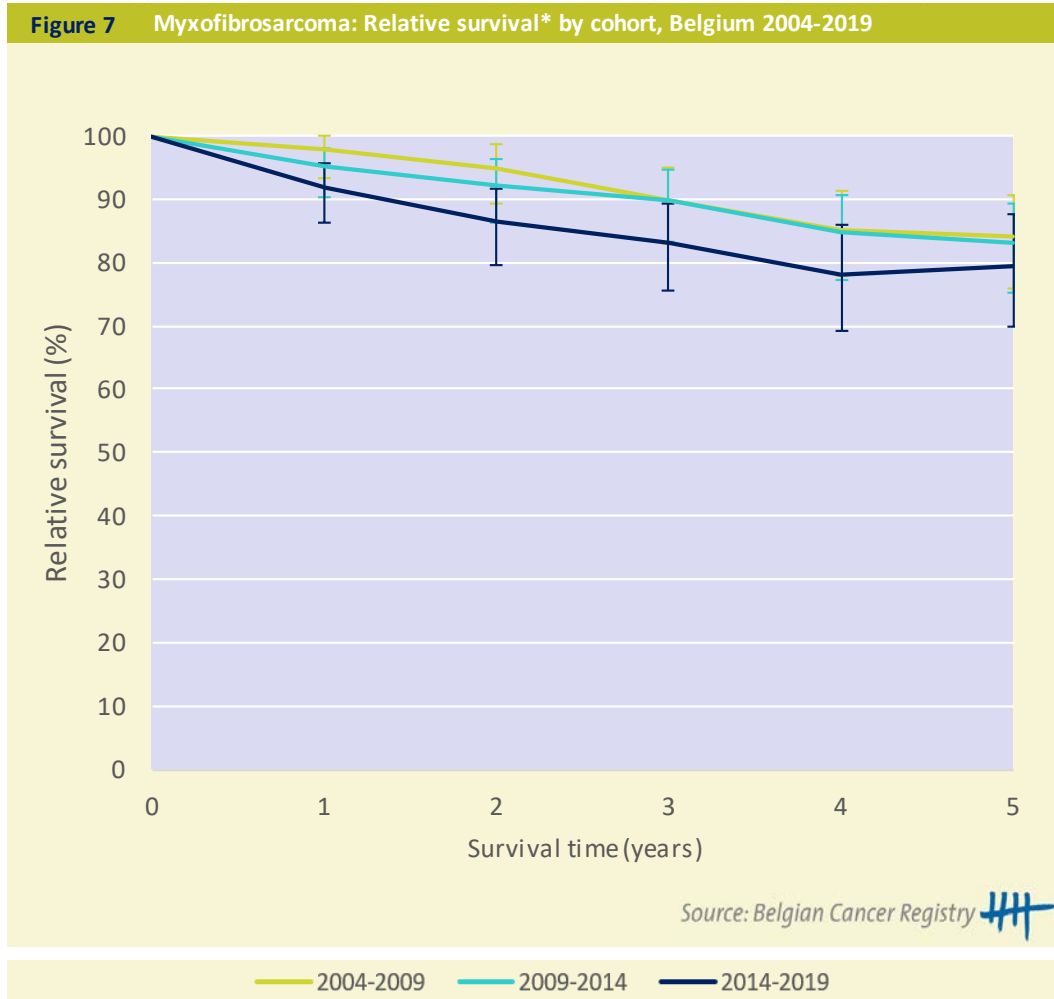
Table 3 Myxofibrosarcoma: Conditional 5-year relative survival* in Belgium, 2010-2019

X years since diagnosis	N at risk	%
1 year	325	85.0
2 year	275	86.9
3 year	223	91.3

* Unadjusted 5-yr relative survival probability conditional on surviving the first X years since diagnosis, %

* Interpretation in lay-man's terms: Given that a patient has already survived X years, what is the relative survival probability 5 years later.

Survival trends



* The relative survival values are represented with 95% Confidence Intervals

3.2.3 VASCULAR SARCOMA

MAIN SUBTYPES:

- *Kaposi sarcoma*
- *Epithelioid haemangioendothelioma*
- *Angiosarcoma*

KEYNOTES

Incidence (table 1-2; figure 1-7)

- Vascular sarcomas are mainly observed in adults, with a peak incidence in the older patients. Vascular sarcomas are much more often diagnosed in males than in females (male/female ratio: 1.9).
- Vascular sarcoma is the most frequent sarcoma type that develops in the breast, heart, mediastinum and pleura (See chapter 3.2).
- Kaposi sarcoma develops more often in males and represents the most frequent vascular tumour between age 20 and 50. Angiosarcoma is more frequent in the female population and is the most frequent vascular tumour after age 60 and -to a much lesser extent- before the age of 20 years. Epithelioid haemangioendothelioma is rare (0.05/100,000 person years) and occurs more frequent in older patients.
- Over time, the incidence of vascular tumours is increasing in males, especially after the age of 60 years.

Survival (table 3; figure 8-11)

- The 10-year relative survival of patients with vascular sarcoma is slightly better in males (55%) than in females (45%). This difference could be explained by the fact that angiosarcomas, which are observed more often in females, are the most aggressive subtype with the worst relative survival of all sarcomas (See chapter 3.2).
- The 5-year relative survival decreases with age, from 76% before the age of 45 years to 39% after the age of 75 years.
- The rare subgroup of haemangioendotheliomas has a 5-year relative survival of 51%.
- No improvement is observed over time.

Table 1 Vascular sarcoma: Overview of incidence, prevalence and survival by sex, Belgium, 2010-2019

Males				
Incidence	N	CR	WSR	
Incidence, 2009-2019	577	1.1	0.7	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	180	3.2	2.0	
10-year prevalence, 31.12.2019	309	5.5	3.4	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	564	59.3	[54.1;64.2]	
10-year relative survival, 2010-2019	564	55.2	[47.6;62.5]	
Females				
Incidence	N	CR	WSR	
Incidence, 2010-2019	417	0.7	0.4	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	120	2.1	1.0	
10-year prevalence, 31.12.2019	197	3.4	1.7	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	412	51.8	[45.7;57.8]	
10-year relative survival, 2010-2019	412	45.0	[35.1;55.1]	
Median age at diagnosis, 2010-2019 (y)	67 [Q1: 50; Q3: 77]			
M/F-ratio	1.9			

Source: Belgian Cancer Registry 

N: number of new diagnoses

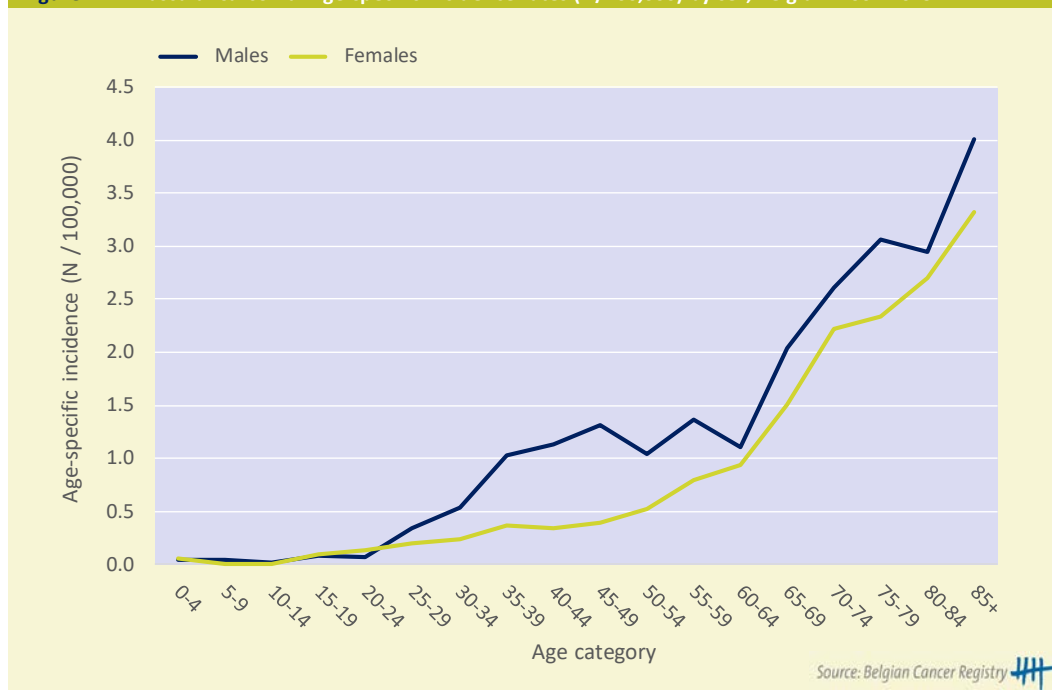
CR: crude (all ages) rate (N/100,000 person years)

WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

M/F-ratio: Male/Female ratio based on the age-standardised rates (2010-2019)

Incidence

Figure 1 Vascular sarcoma: Age-specific incidence rates (N/100,000) by sex, Belgium 2004-2019



Source: Belgian Cancer Registry 

Figure 2 Vascular sarcoma: Incidence distribution by subtype, Belgium 2010-2019

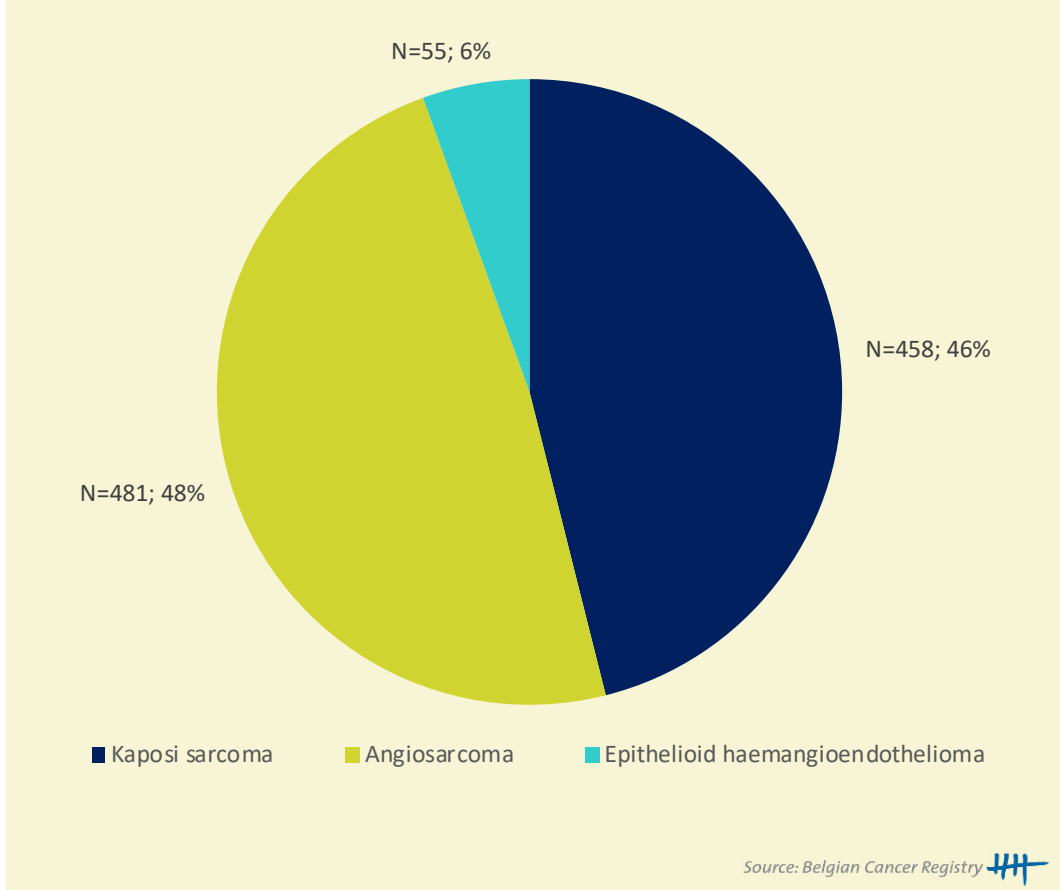


Figure 3 Vascular sarcoma: Age-specific incidence rates (N/100,000) by subtype, Belgium 2004-2019

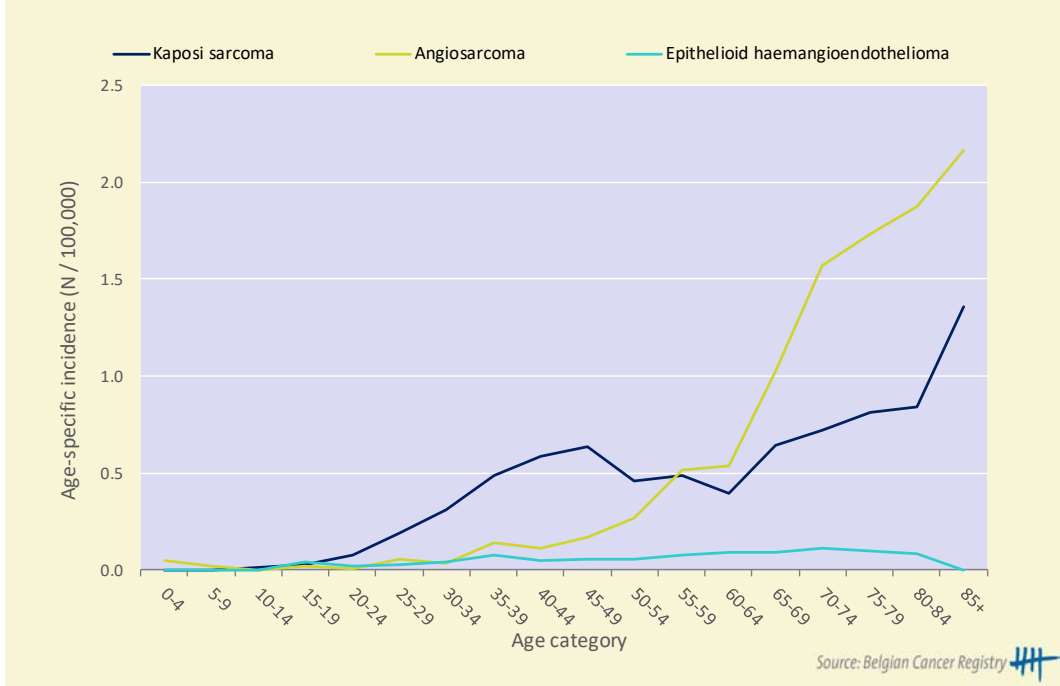
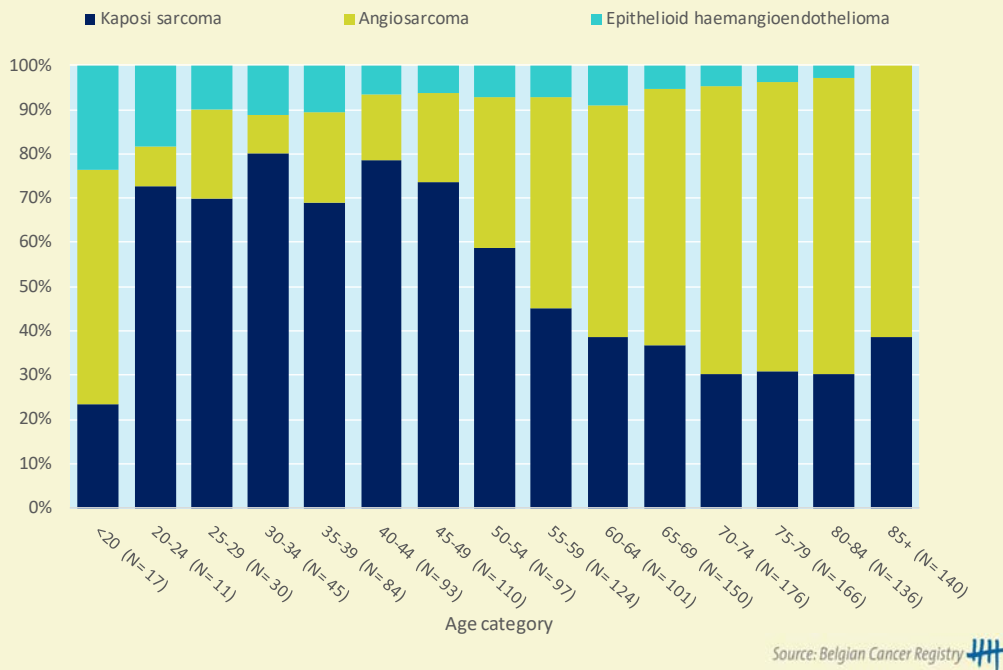
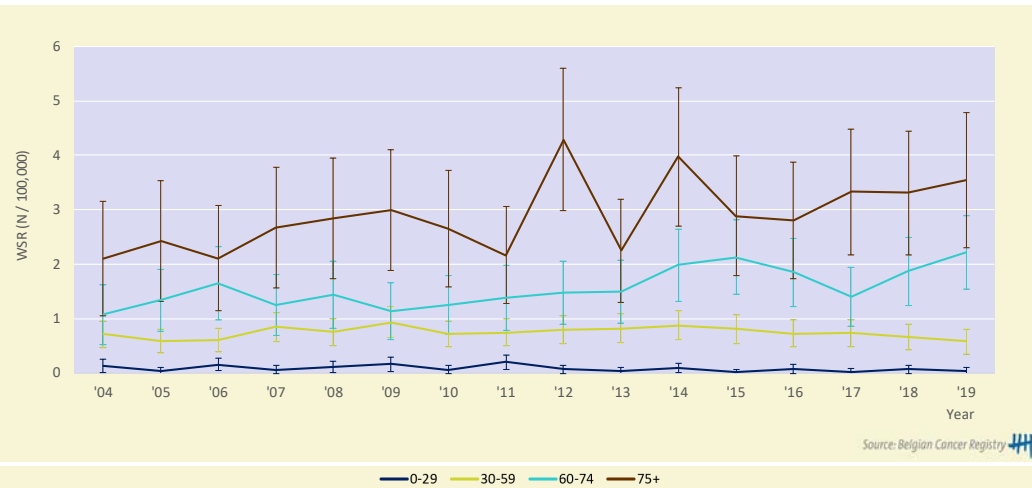


Figure 4 Vascular sarcoma: Subtype incidence distribution (%) by age category, Belgium 2004-2019



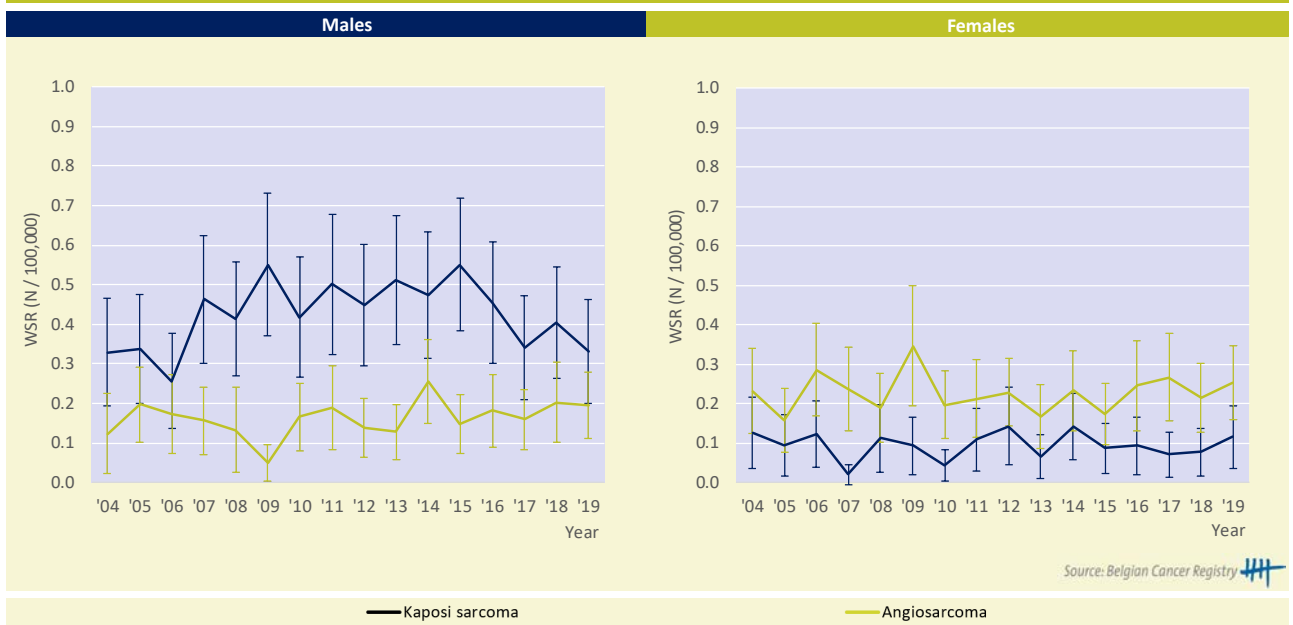
Incidence trends

Figure 5 Vascular sarcoma: Age-standardised incidence rates* (WSR) by age category, males and females, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals.

Figure 6 Vascular sarcoma: Age-standardised incidence rates* (WSR) by sex and subtype, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals.

Table 2 Vascular sarcoma: Incidence trends by sex and age category in Belgium, 2004-2019

Age category	Males			Females		
	AAPC (%)	95%CI	Period	AAPC (%)	95%CI	Period
All ages	1.3	[0.0; 2.7]	2004-2019	0.0	[-2.3; 2.2]	2004-2019
	5.0	[2.2; 7.9]	2004-2012			
	-2.7	[-5.7; 0.4]	2012-2019			
0 - 29 y	-	-	-	-	-	-
30 - 59 y	-0.5	[-2.2; 1.2]	2004-2019	-0.6	[-3.3; 2.2]	2004-2019
	7.9	[-0.6; 17.2]	2004-2007			
	2.5	[-0.2; 5.2]	2007-2015			
	-11.8	[-16.8; -6.4]	2015-2019			
60 - 74 y	4.8	[1.7; 8.0]	2004-2019	2.3	[-0.1; 4.8]	2004-2019
75+ y	4.3	[0.3; 8.5]	2004-2019	2.7	[-0.5; 5.9]	2004-2019

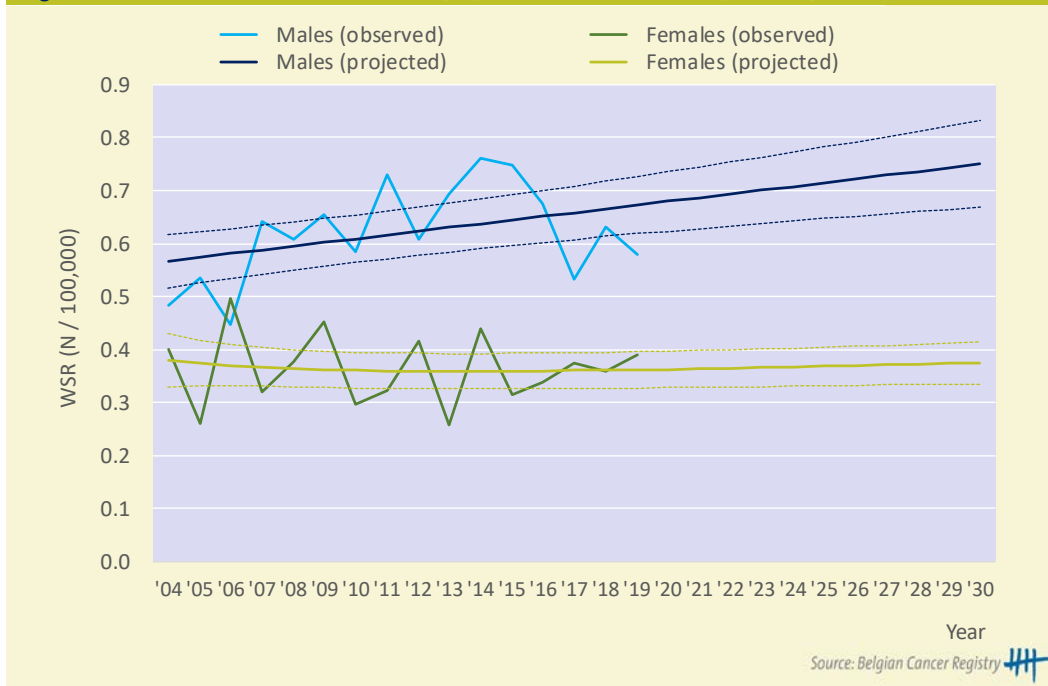
Source: Belgian Cancer Registry

AAPC: average annual percentage change

Period: When a joinpoint occurred, APC's are calculated for the period before and after the joinpoint. This column represents the corresponding time interval. AAPC's are always calculated over the entire study-period.

Incidence projections

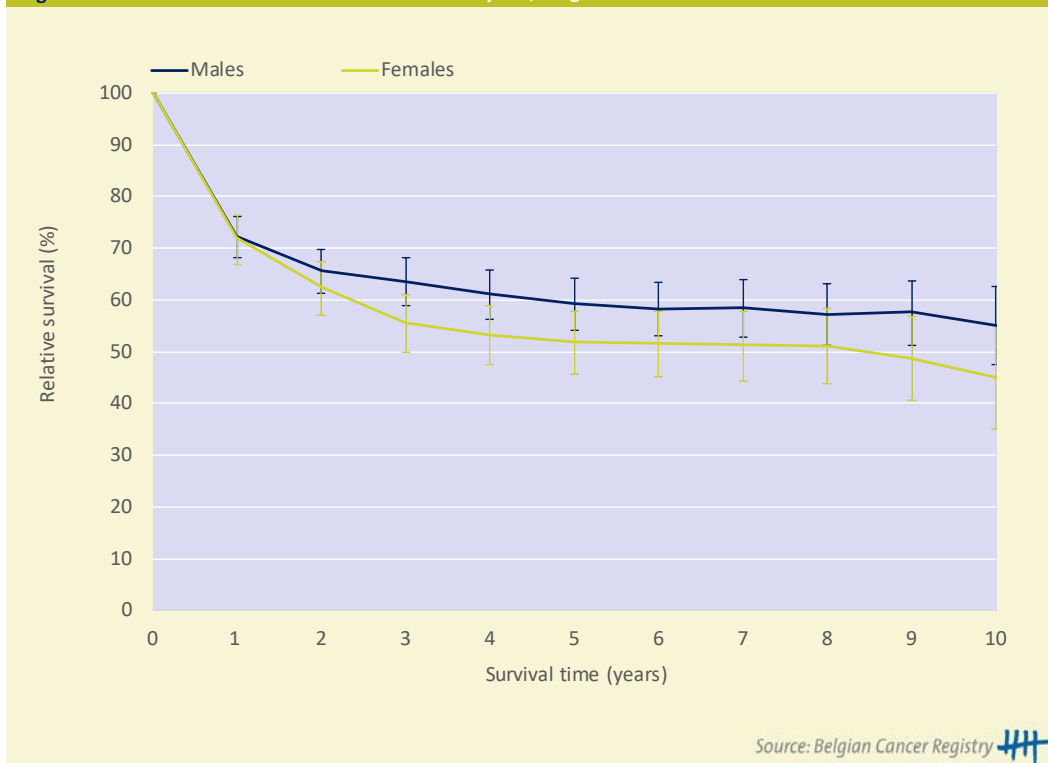
Figure 7 Vascular sarcoma: Observed and projected* incidence (WSR) by sex, Belgium 2004-2030



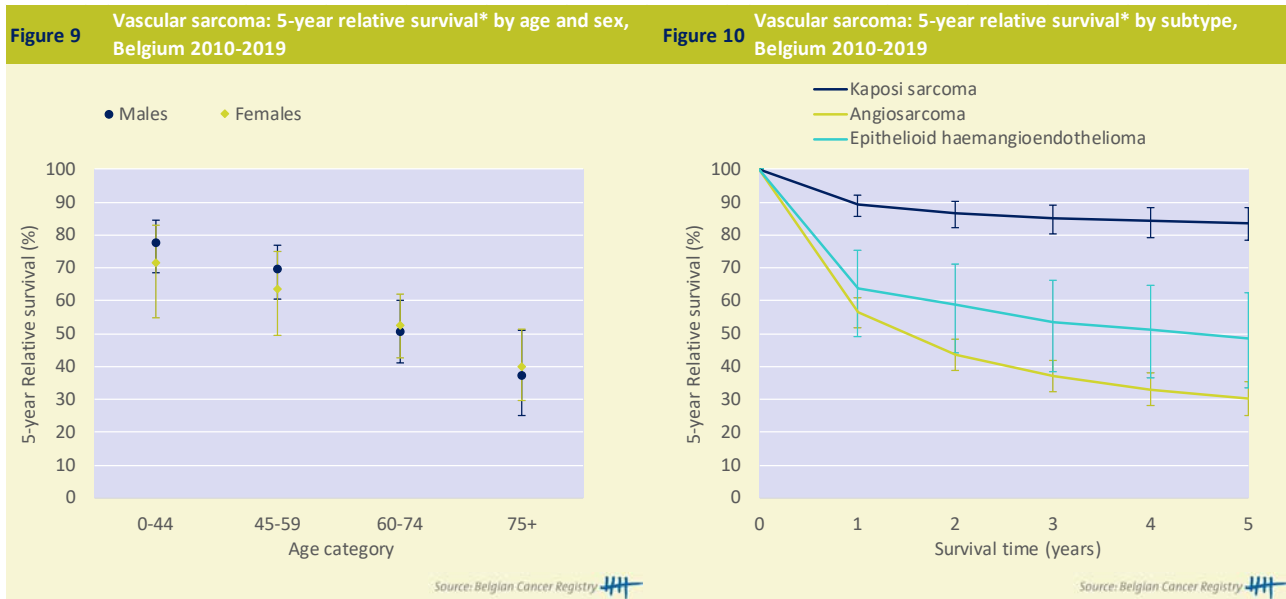
WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

Survival

Figure 8 Vascular sarcoma: Relative survival* by sex, Belgium 2010-2019



* The relative survival values are represented with 95% Confidence Intervals



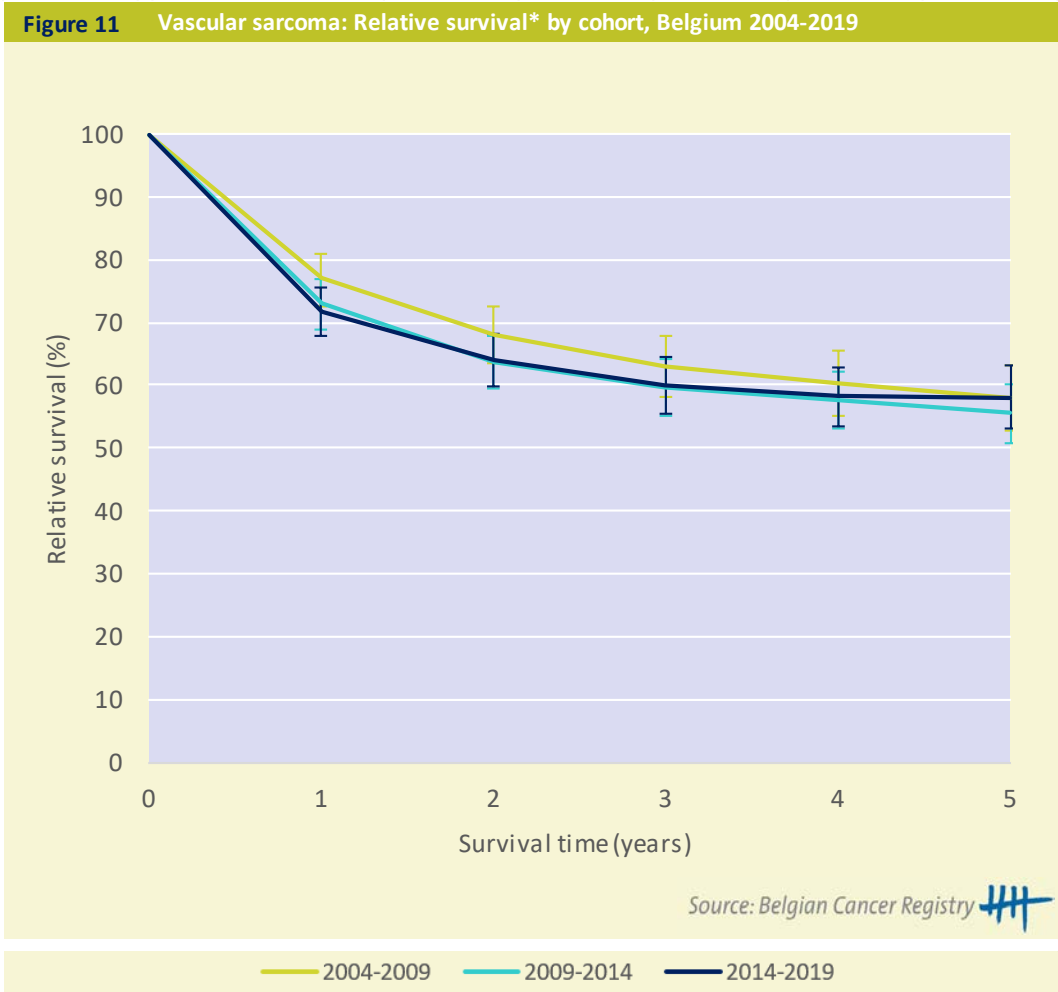
* The relative survival values are represented with 95% Confidence Intervals

Table 3 Vascular sarcoma: Conditional 5-year relative survival* in Belgium, 2010-2019

X years since diagnosis	N at risk	%
1 year	686	76.9
2 year	545	86.1
3 year	439	90.6

* Unadjusted 5-yr relative survival probability conditional on surviving the first X years since diagnosis, %
 * Interpretation in lay-man's terms: Given that a patient has already survived X years, what is the relative survival probability 5 years later.

Survival trends



* The relative survival values are represented with 95% Confidence Intervals

3.2.3.1 KAPOSI SARCOMA

KEYNOTES

Incidence (table 1-2; figure 1-4)

- Kaposi sarcoma is mainly observed in adult males older than 35 years, with a peak incidence above 80 years (male/female ratio: 4.7).
- AIDS-associated Kaposi sarcoma, the most aggressive form when untreated, is found in HIV-1–infected individuals. Its incidence has been reduced with the introduction of highly active antiretroviral therapy in the 1990's (before the start of Belgian Cancer Registry)^(1, 33).
- Almost half of the Kaposi sarcomas are observed in the skin of the lower limbs and hip.

Survival (table 3; figure 5-7)

- The relative survival of patients with Kaposi sarcoma is similar for males and females, and reaches a plateau two years after diagnosis around 87%.
- Given that a patient survives the first two years after diagnosis, the conditional relative survival 5 years later is more than 97%.
- The 5-year relative survival of patients with Kaposi sarcoma:
 - slightly decreases with age (from nearly 90% before 45 years to circa 75% after 75 years).
 - is improving over time: from 77% in 2004-2009 to 86% in 2014-2019.

Table 1 Kaposi sarcoma: Overview of incidence, prevalence and survival by sex in Belgium, 2010-2019

Males				
Incidence	N	CR	WSR	
Incidence, 2010-2019	362	0.7	0.4	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	143	2.5	1.6	
10-year prevalence, 31.12.2019	261	4.6	3.0	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	356	84.6	[78.7;89.6]	
10-year relative survival, 2010-2019	356	79.2	[68.3;88.6]	
Females				
Incidence	N	CR	WSR	
Incidence, 2010-2019	96	0.2	0.1	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	34	0.6	0.3	
10-year prevalence, 31.12.2019	66	1.1	0.6	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	95	80.0	[66.5;90.9]	
10-year relative survival, 2010-2019	95	71.7	[39.2;99.0]	
Median age at diagnosis, 2010-2019 (y)	56 [Q1: 44; Q3: 72]			
M/F-ratio	4.7			

Source: Belgian Cancer Registry 

N: number of new diagnoses

CR: crude (all ages) rate (N/100,000 person years)

WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

M/F-ratio: Male/Female ratio based on the age-standardised rates (2010-2019)

Incidence

Figure 1 Kaposi sarcoma: Age-specific incidence rates (N/100,000) by sex, Belgium 2004-2019

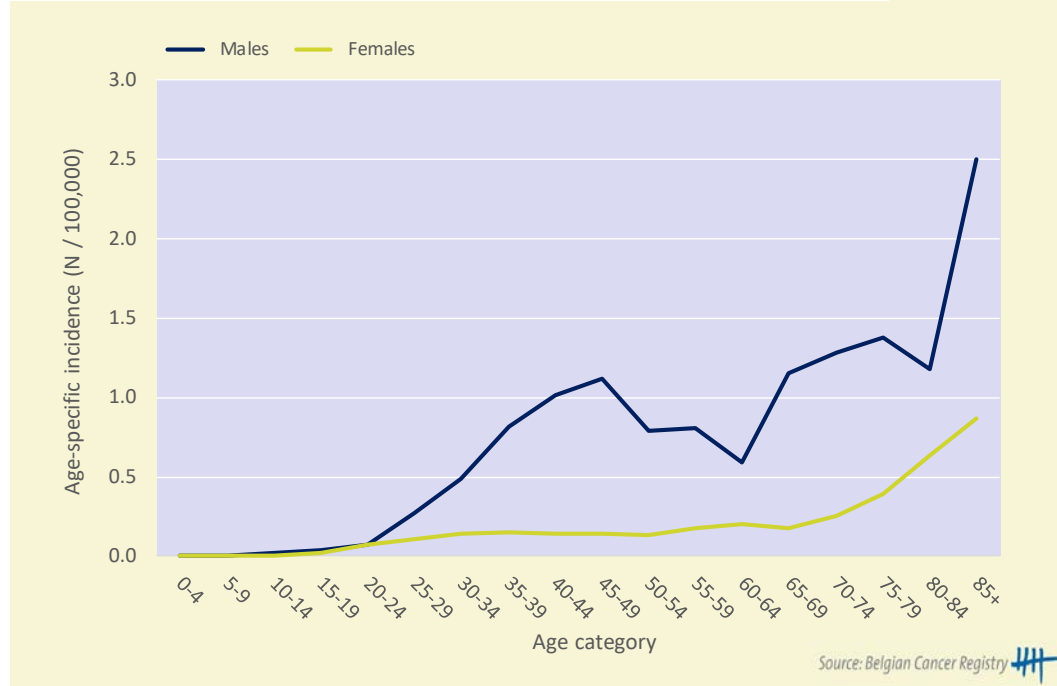
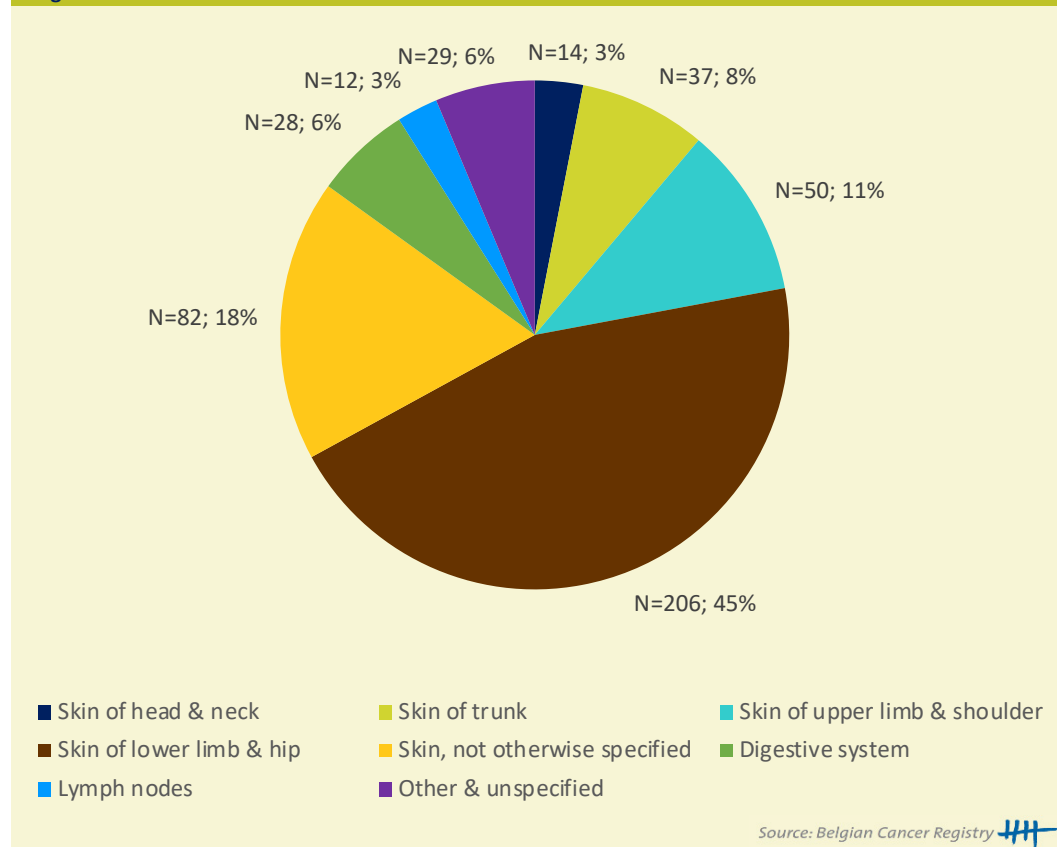
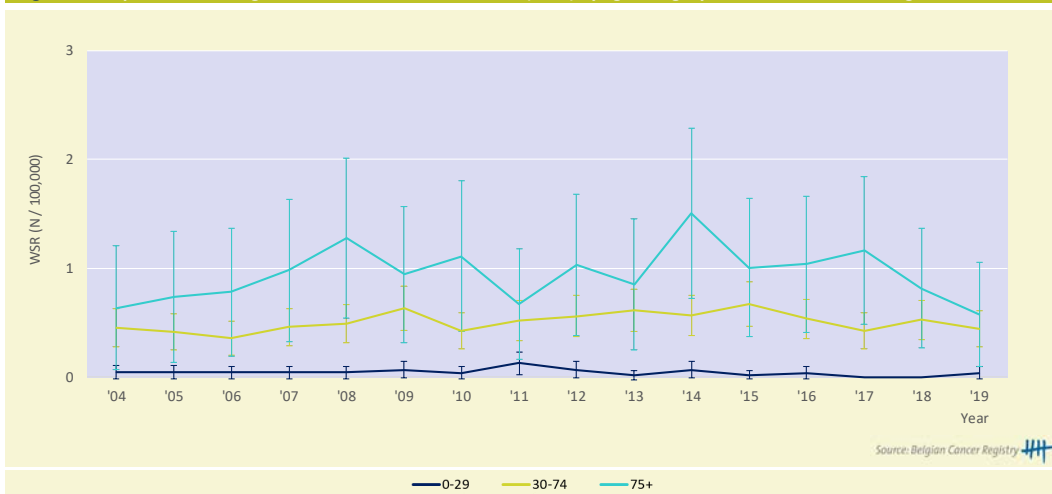


Figure 2 Kaposi sarcoma: Incidence distribution by primary tumour location, Belgium 2010-2019



Incidence trends

Figure 3 Kaposi sarcoma: Age-standardised incidence rates* (WSR) by age category, males and females, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals

Table 2 Kaposi sarcoma: Incidence trends by sex and age category in Belgium, 2004-2019

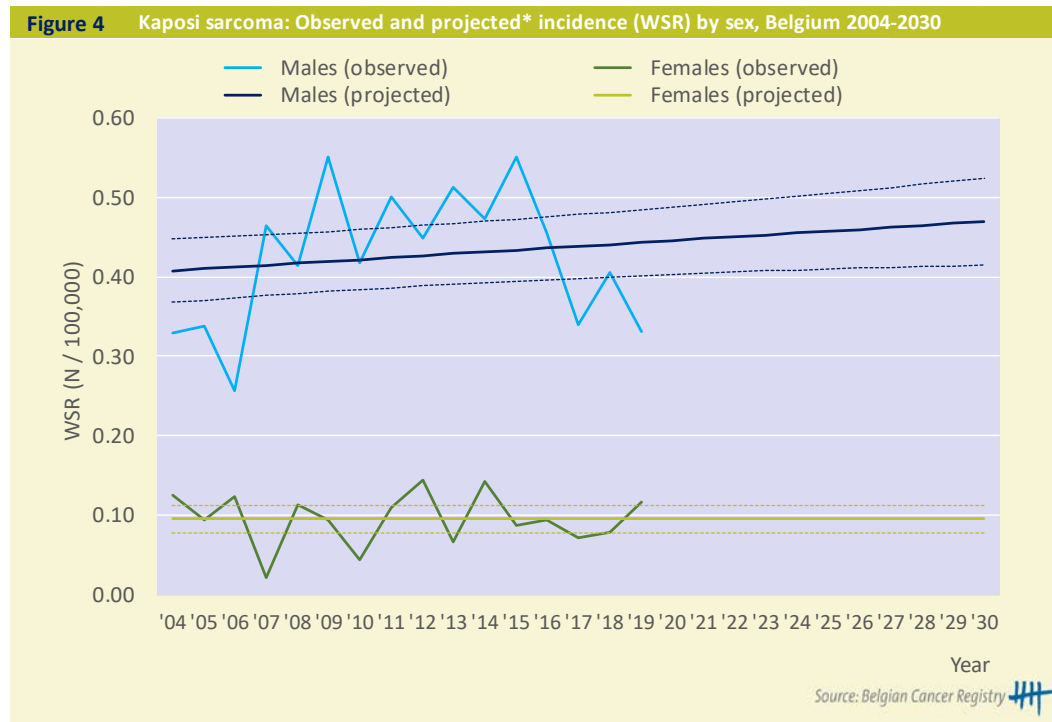
Age category	Males			Females		
	AAPC (%)	95%CI	Period	AAPC (%)	95%CI	Period
All ages	0.9	[-1.0; 2.8]	2004-2019	0.8	[-5.0; 6.9]	2004-2019
	7.0	[2.9; 11.2]	2004-2012			
	-5.6	[-9.8; -1.3]	2012-2019			
0 - 29 y	-	-	-	-	-	-
30 - 74 y	2.0	[-0.3; 4.3]	2004-2019	3.4	[-2.6; 9.8]	2004-2019
	10.9	[2.7; 19.8]	2004-2009			
	-2.2	[-5.6; 1.3]	2009-2019			
75+ y	2.2	[-3.3; 8.0]	2004-2019	-1.4	[-6.6; 4.2]	2004-2019

Source: Belgian Cancer Registry

AAPC: average annual percentage change

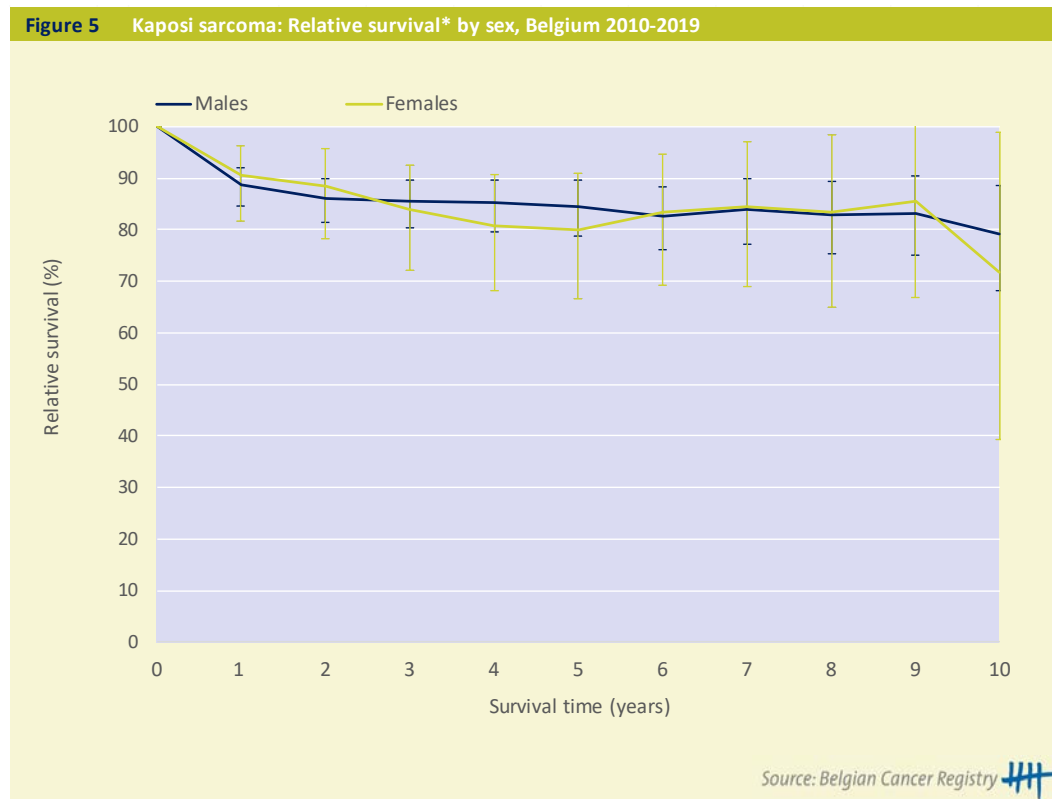
Period: When a joinpoint occurred, APC's are calculated for the period before and after the joinpoint. This column represents the corresponding time interval. AAPC's are always calculated over the entire study-period.

Incidence projections



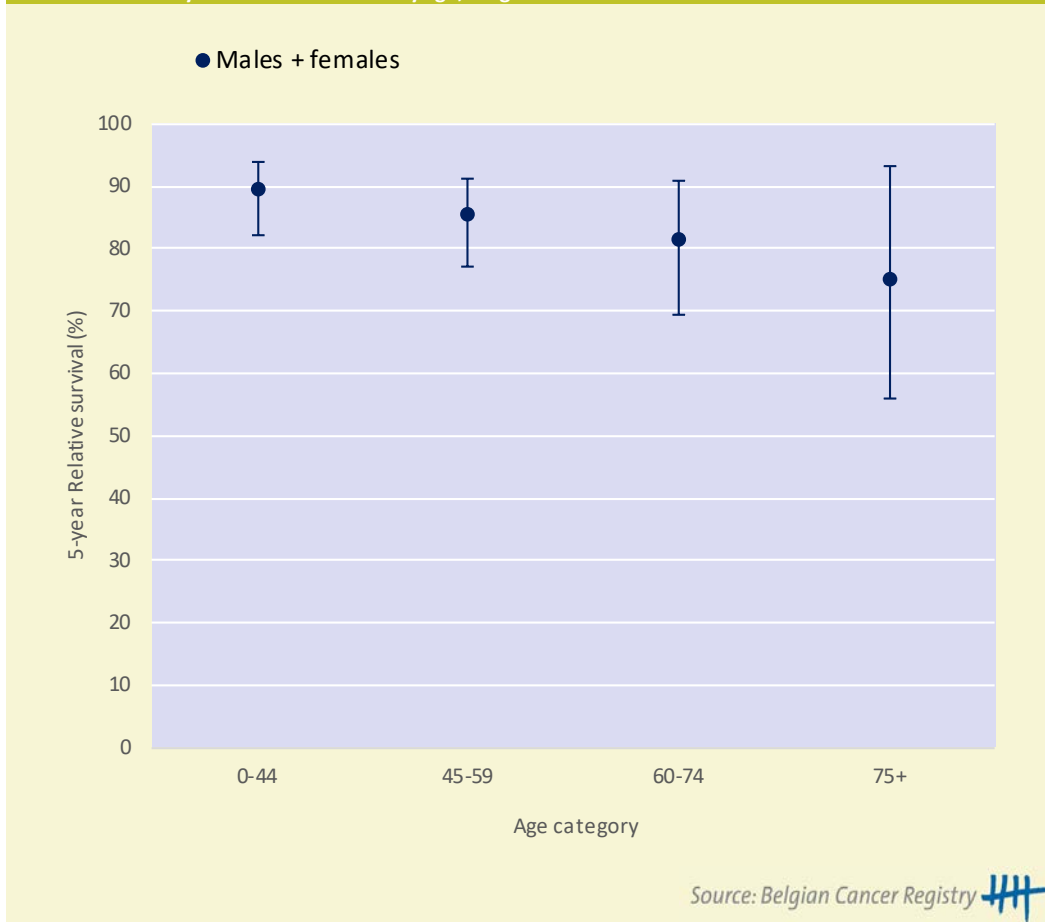
WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

Survival



* The relative survival values are represented with 95% Confidence Intervals

Figure 6 Kaposi sarcoma:
5-year relative survival* by age, Belgium 2010-2019



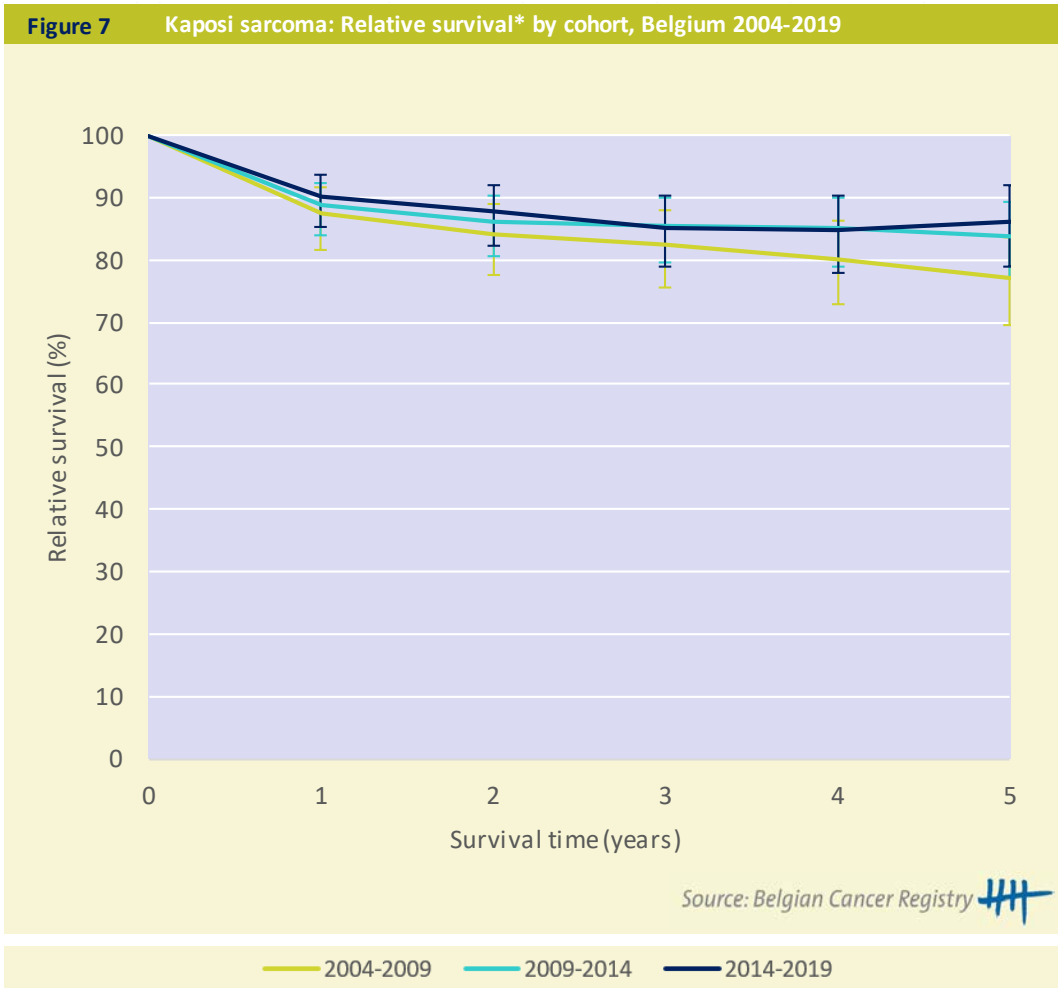
* The relative survival values are represented with 95% Confidence Intervals

Kaposi sarcoma: Conditional 5-year relative survival* in Belgium, 2010-2019		
X years since diagnosis	N at risk	%
1 year	388	92.9
2 year	334	97.1
3 year	291	97.5

* Unadjusted 5-yr relative survival probability conditional on surviving the first X years since diagnosis, %

* Interpretation in lay-man's terms: Given that a patient has already survived X years, what is the relative survival probability 5 years later.

Survival trends



* The relative survival values are represented with 95% Confidence Intervals

3.2.3.2 ANGIOSARCOMA

KEYNOTES

Incidence (table 1-2; figure 1-4)

- Angiosarcoma is slightly more frequent in females than in males (male/female ratio of 0.8) and is mostly diagnosed in the older population (peak incidence above 70 years of age).
- The incidence of angiosarcoma tends to increase in women older than 75 years (average annual percentage change: +3.4%).
- One out of three angiosarcomas is diagnosed in breast tissue and often therapy related. Of 135 patients with a breast angiosarcoma diagnosis in 2012-2019, 62% previously received radiotherapy for a prior breast cancer*.

Survival (table 3; figure 5-8)

- Angiosarcoma is the most aggressive sarcoma subtype, with the worst 5-year relative survival rate (30% - see chapter 3.2).
- The 5-year relative survival of angiosarcoma patients is significantly higher in females (41%) than in males (14%).
- While the 5-year relative survival decreases with age in women (from nearly 50% under 65 years of age to 30% after the age of 75 years), it remains constant between 10 and 20% in males whatever the age category.
- The apparent decrease of the 5-year relative survival rate observed in the period 2009-2019, compared to the period 2004-2009 may be partly due to the more accurate diagnosis and registration.

* Breast cancers treated with radiotherapy (based on nomenclature codes using data from the Inter-mutualistic Agency (IMA)) diagnosed in the period 2004 until one year prior to the breast angiosarcoma diagnosis. Data not shown.

Table 1 Angiosarcoma: Overview of incidence, prevalence and survival by sex in Belgium, 2010-2019				
Males				
Incidence	N	CR	WSR	
Incidence, 2010-2019	189	0.3	0.2	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	30	0.5	0.3	
10-year prevalence, 31.12.2019	39	0.7	0.3	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	183	14.4	[8.9;21.2]	
10-year relative survival, 2004-2019	251	13.1	[7.6;20.6]	
Females				
Incidence	N	CR	WSR	
Incidence, 2010-2019	292	0.5	0.2	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	76	1.3	0.5	
10-year prevalence, 31.12.2019	112	1.9	0.8	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	288	40.8	[33.9;47.9]	
10-year relative survival, 2004-2019	444	34.6	[23.0;43.5]	
Median age at diagnosis, 2010-2019 (y)	72 [Q1: 63; Q3: 80]			
M/F-ratio	0.8			

Source: Belgian Cancer Registry 

N: number of new diagnoses

CR: crude (all ages) rate (N/100,000 person years)

WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

M/F-ratio: Male/Female ratio based on the age-standardised rates (2010-2019)

Incidence

Figure 1 Angiosarcoma: Age-specific incidence rates (N/100,000) by sex, Belgium 2004-2019

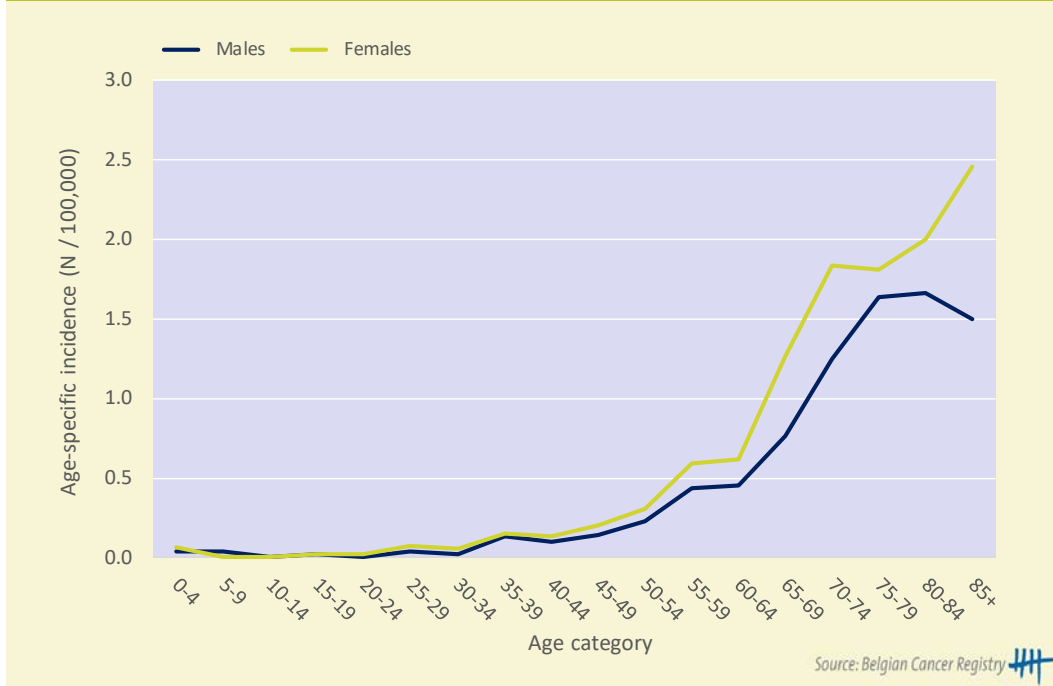
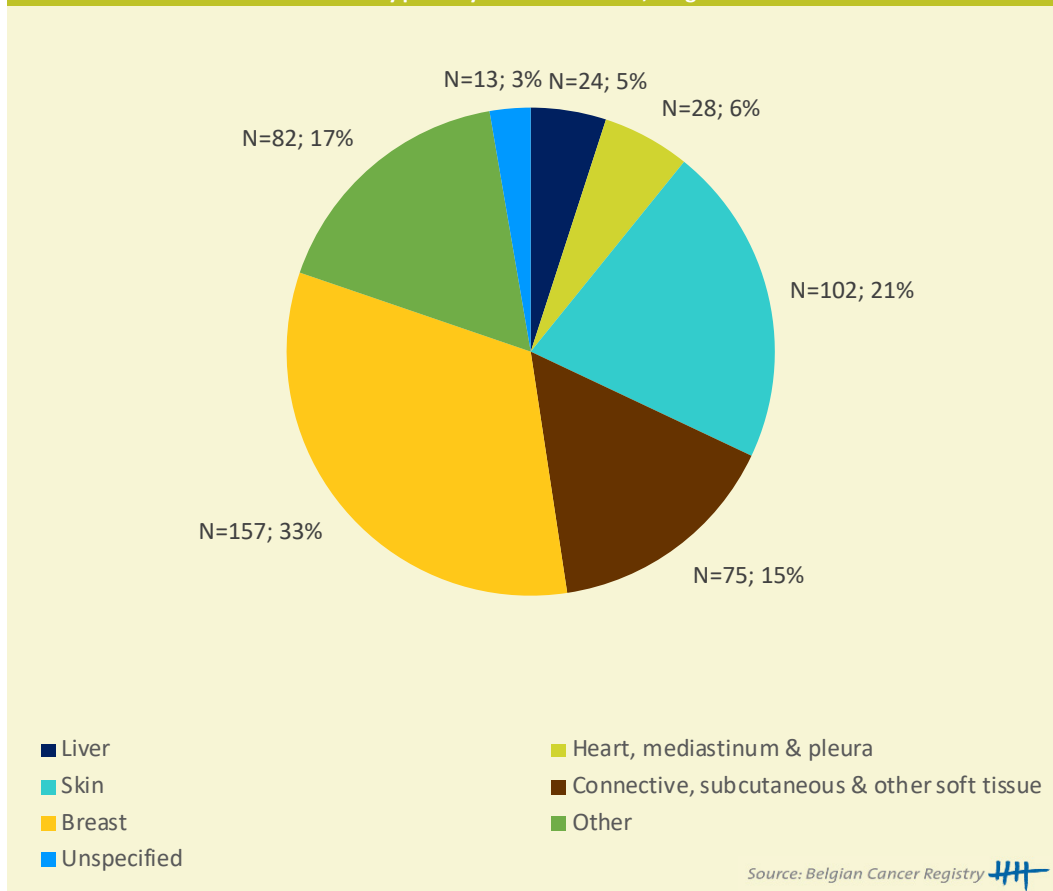
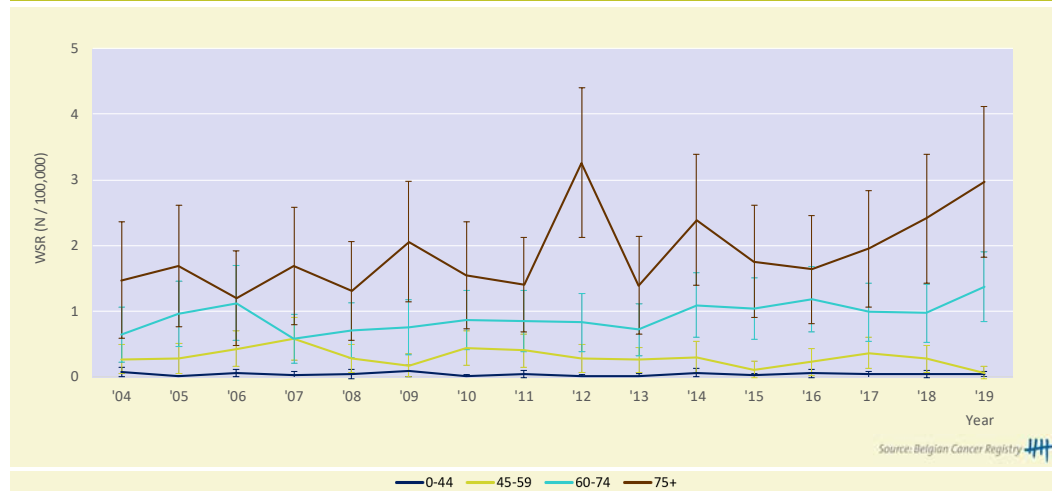


Figure 2 Angiosarcoma: Incidence distribution by primary tumour location, Belgium 2010-2019



Incidence trends

Figure 3 Angiosarcoma: Age-standardised incidence rates* (WSR) by age category, males and females, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals

Table 2 Angiosarcoma: Incidence trends by sex and age category in Belgium, 2004-2019

Age category	Males			Females		
	AAPC (%)	95%CI	Period	AAPC (%)	95%CI	Period
All ages	2.3	[-1.7; 6.5]	2004-2019	0.3	[-2.1; 2.8]	2004-2019
0 - 44 y	-	-	-	-	-	-
45 - 59 y	-	-	-	-9.0	[-15.0; -2.6]	2004-2019
60 - 74 y	4.4	[-1.6; 10.7]	2004-2019	3.0	[-0.6; 6.7]	2004-2019
75+ y	6.2	[-0.1; 12.9]	2004-2019	3.4	[0.2; 6.6]	2004-2019

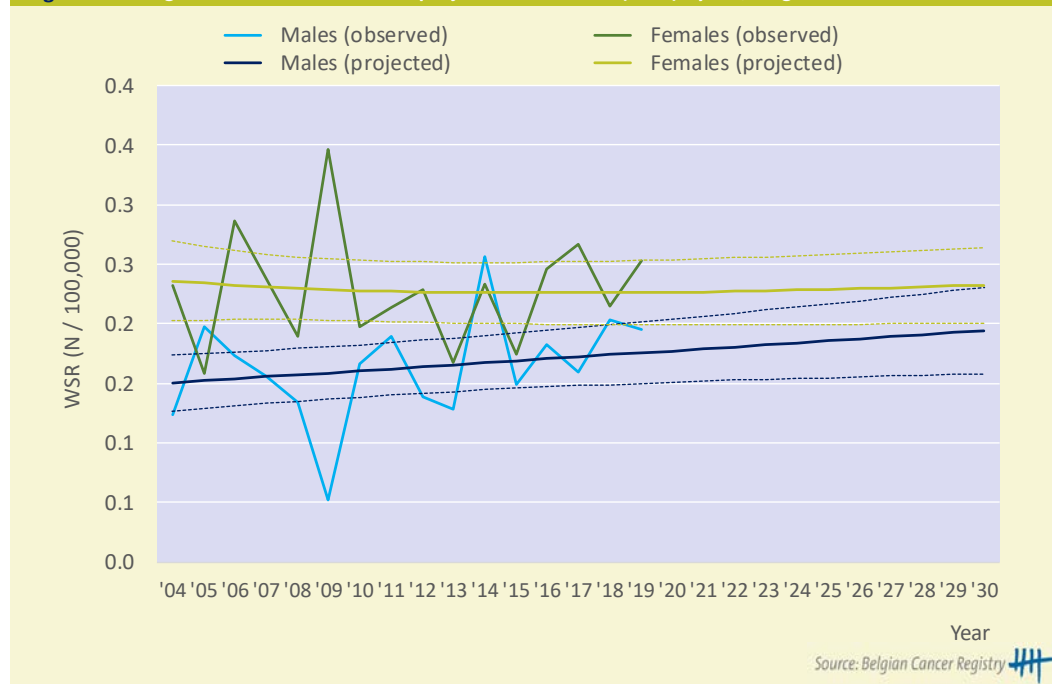
Source: Belgian Cancer Registry

AAPC: average annual percentage change

Period: When a joinpoint occurred, APC's are calculated for the period before and after the joinpoint. This column represents the corresponding time interval. AAPC's are always calculated over the entire study-period.

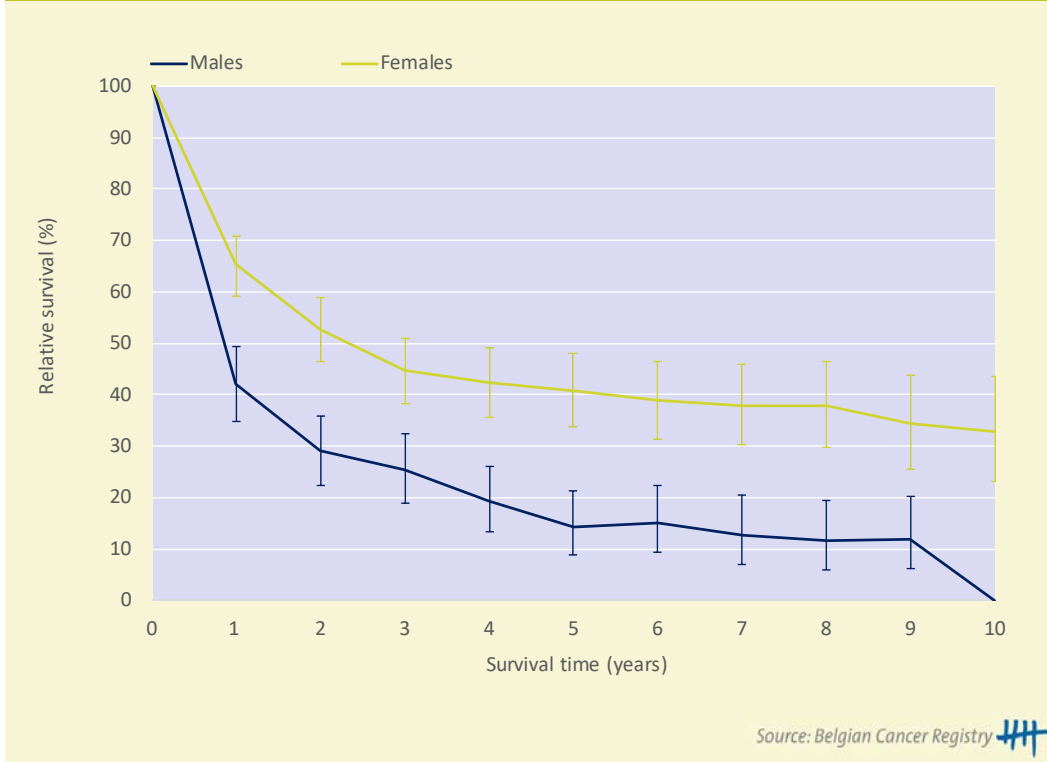
Incidence projections

Figure 4 Angiosarcoma: Observed and projected* incidence (WSR) by sex, Belgium 2004-2030



Survival

Figure 5 Angiosarcoma: Relative survival* by sex, Belgium 2010-2019



* The relative survival values are represented with 95% Confidence Intervals

Figure 6 Angiosarcoma: 5-year relative survival* by age and sex, Belgium 2010-2019

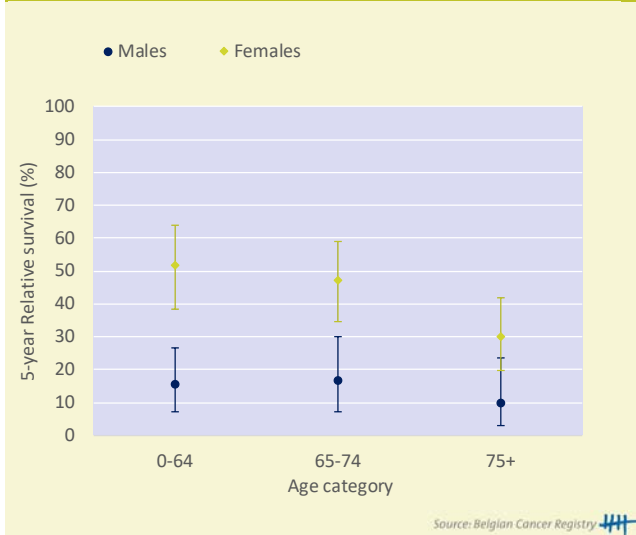
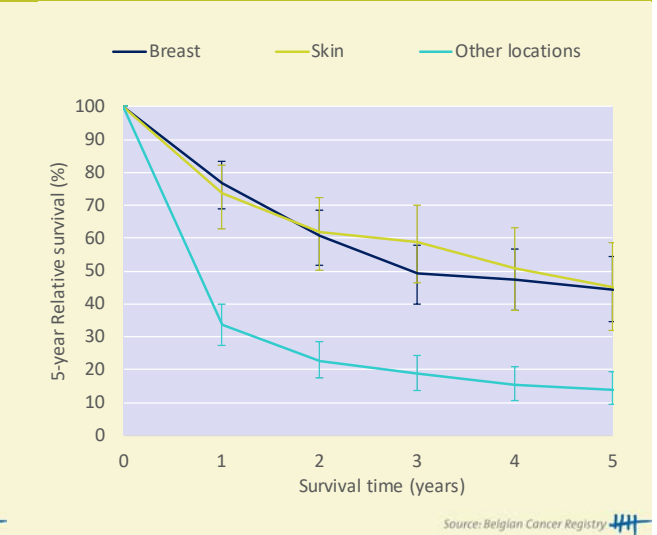


Figure 7 Angiosarcoma: 5-year relative survival by primary tumour location, Belgium 2010-2019

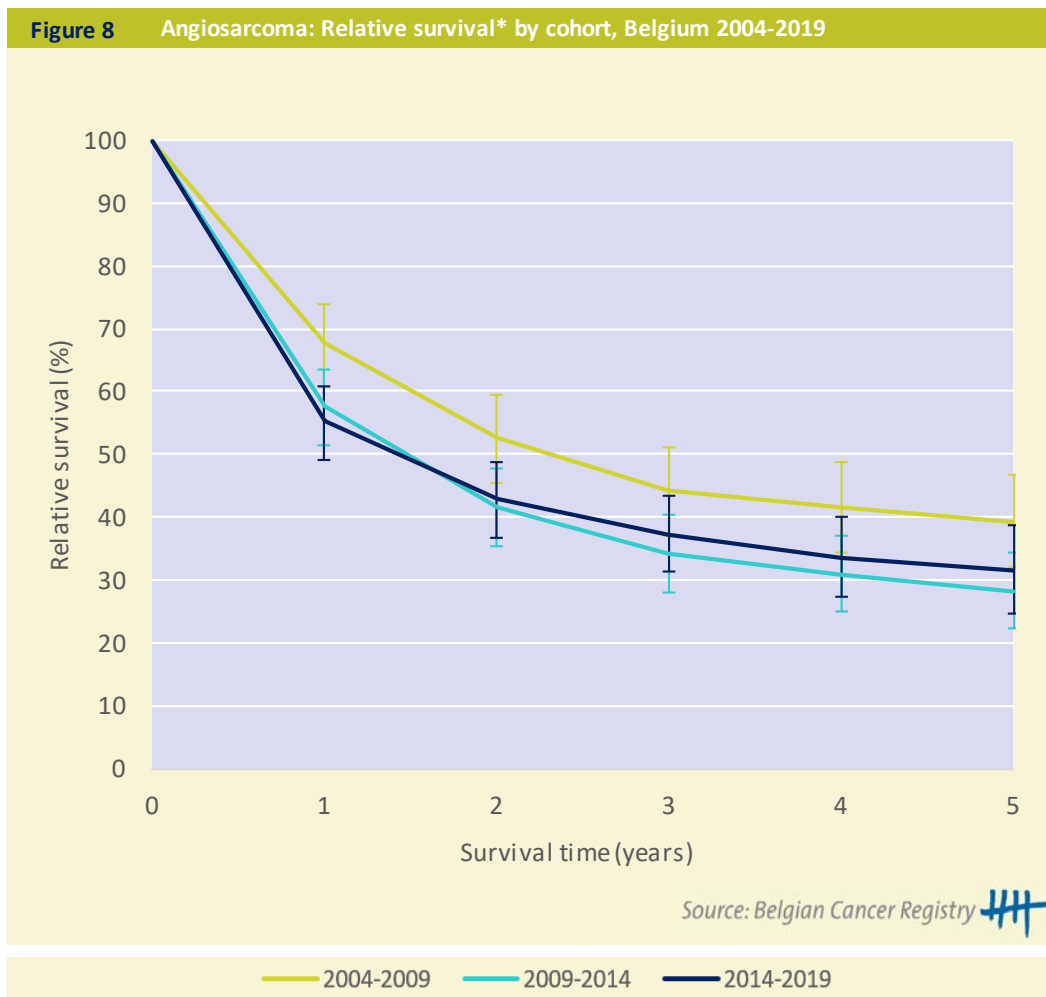


* The relative survival values are represented with 95% Confidence Intervals

Table 3 Angiosarcoma: Conditional 5-year relative survival* in Belgium, 2010-2019			
X years since diagnosis	N at risk	%	
1 year	262	51.7	
2 year	180	63.8	
3 year	126	73.1	

* Unadjusted 5-yr relative survival probability conditional on surviving the first X years since diagnosis, %
 * Interpretation in lay-man's terms: Given that a patient has already survived X years, what is the relative survival probability 5 years later.

Survival trends



* The relative survival values are represented with 95% Confidence Intervals

KEYNOTES

Incidence (table 1-2; figure 1-6)

- Leiomyosarcoma is more frequently diagnosed in females than in males (male/female ratio of 0.6), except for the oldest age group (75+ years).
- Leiomyosarcoma is the third most frequently occurring sarcoma overall, and in females the second most frequent.
- Leiomyosarcoma is the most frequent histological subtype of sarcomas arising in the female genital organs and in the urinary system (See chapter 3.2).
- The primary tumour location of leiomyosarcoma differs between males and females:
 - In males the skin (26%) and connective and soft tissues of limbs, head & neck and trunk (44%) represent more than 2/3. Note most leiomyosarcomas of the skin are no longer considered malignant but still included in the incidence data conform the classification applicable in the respective incidence years.
 - In females nearly half of leiomyosarcomas are diagnosed in genital organs (mostly corpus uteri).
- The majority of the leiomyosarcomas that present in the female genital organs are stage I (62%) at diagnosis, followed by stage IV tumours (29%).
- The apparent decrease of leiomyosarcoma incidence over time may be partly explained by:
 - More accurate diagnosis and registration. The increasing use of immunostainings and increased awareness for gastrointestinal stromal tumours could lead to reclassification of cases.
 - Changes in the classification of skin leiomyosarcoma. From 2019 onwards, most leiomyosarcoma are no longer considered malignant.
- Based on incidence projections, the incidence rates (WSR) are expected to remain stable in the future.

Survival (table 3; figure 7-9)

- Note that conform changes in the classification, most leiomyosarcomas of the skin are no longer considered malignant and therefore their survival is represented separately.
- The 5-year relative survival rate:
 - Decreases with age (from 74% under the age of 50 years to 40% after the age of 70 years).
 - Does not show a clear improvement over time (2004-2019).

Table 1 Leiomyosarcoma: Overview of incidence, prevalence and survival by sex, Belgium, 2010-2019

Males				
Incidence	N	CR	WSR	
Incidence, 2010-2019	470	0.9	0.5	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	169	3.0	1.5	
10-year prevalence, 31.12.2019	264	4.7	2.4	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	467	63.2	[56.9;69.2]	
10-year relative survival, 2010-2019	467	58.3	[47.7;69.0]	
Females				
Incidence	N	CR	WSR	
Incidence, 2010-2019	759	1.3	0.8	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	232	4.0	2.2	
10-year prevalence, 31.12.2019	400	6.9	3.7	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	747	57.2	[52.9;61.4]	
10-year relative survival, 2010-2019	747	49.3	[43.1;55.4]	
Median age at diagnosis, 2010-2019 (y)	66 [Q1: 53; Q3: 77]			
M/F-ratio	0.6			

Source: Belgian Cancer Registry 

N: number of new diagnoses

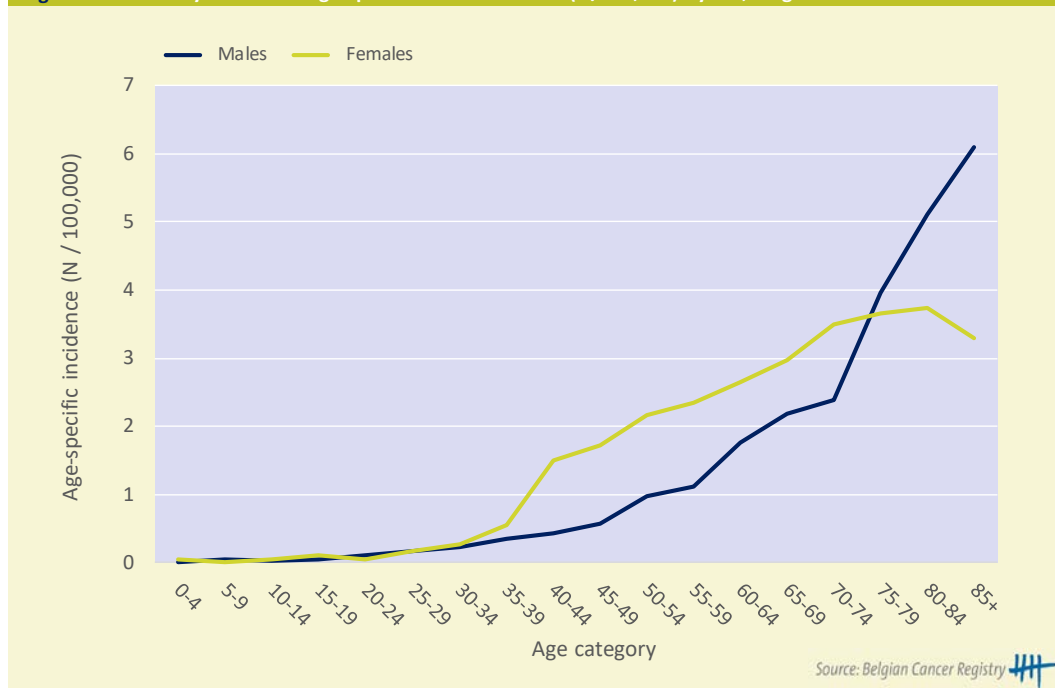
CR: crude (all ages) rate (N/100,000 person years)

WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

M/F-ratio: Male/Female ratio based on the age-standardised rates (2010-2019)

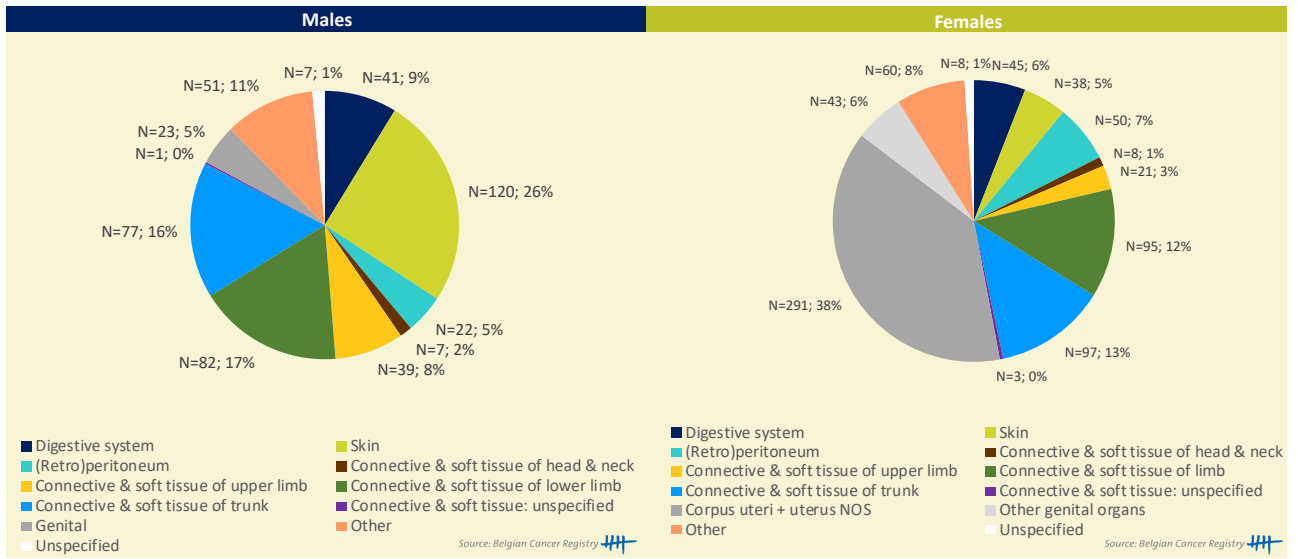
Incidence

Figure 1 Leiomyosarcoma: Age-specific incidence rates (N/100,000) by sex, Belgium 2004-2019



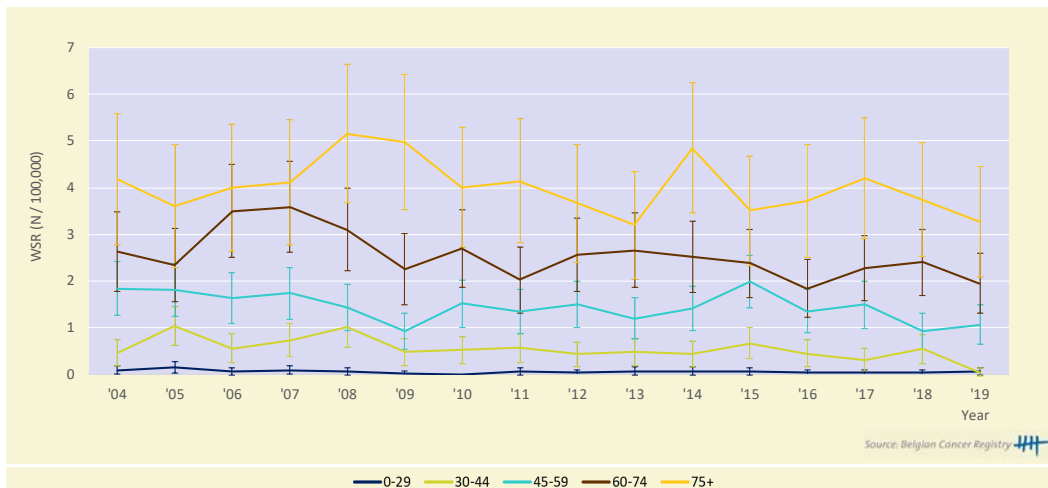
Source: Belgian Cancer Registry 

Figure 2 Leiomyosarcoma: Incidence distribution by primary tumour location, Belgium 2010-2019



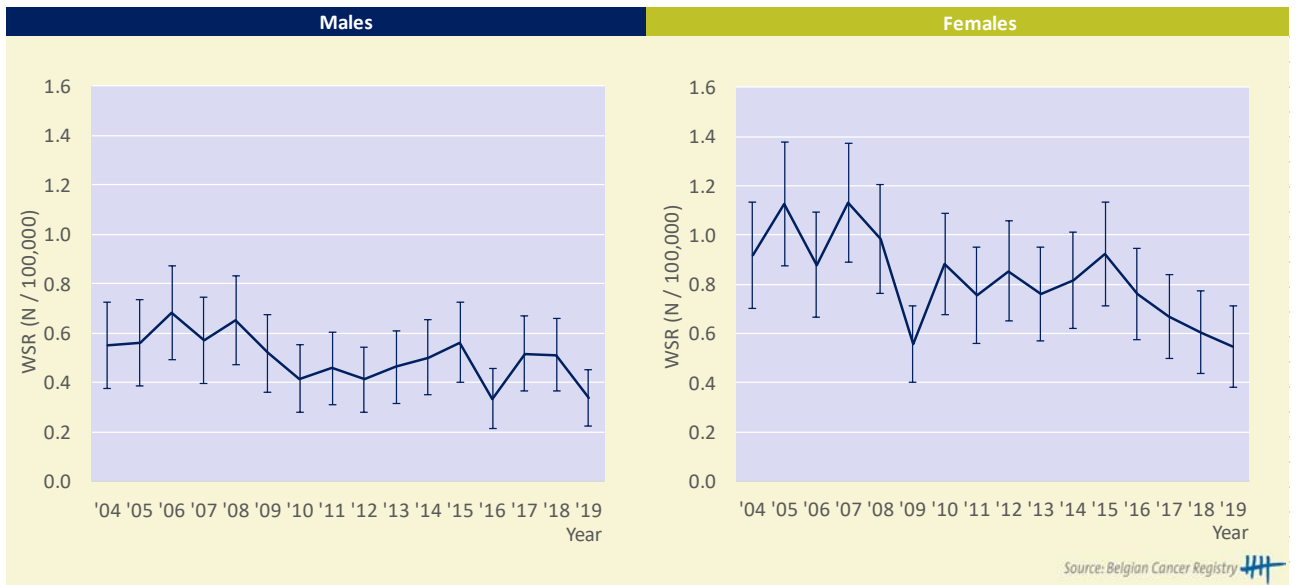
Incidence trends

Figure 3 Leiomyosarcoma: Age-standardised incidence rates* (WSR) by age category, males and females, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals.

Figure 4 Leiomyosarcoma: Age-standardised incidence rates* (WSR) by sex, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals.

Table 2 Leiomyosarcoma: Incidence trends by sex and age category in Belgium, 2004-2019

Age category	Males			Females		
	AAPC (%)	95%CI	Period	AAPC (%)	95%CI	Period
All ages	-2.5	[-4.4; -0.6]	2004-2019	-3.1	[-4.9; -1.2]	2004-2019
0 - 29 y	-	-	-	-	-	-
30 - 44 y	-	-	-	-7.4	[-13.5; -0.8]	2004-2019
45 - 59 y	-2.6	[-6.2; 1.1]	2004-2019	-2.4	[-5.4; 0.7]	2004-2019
60 - 74 y	-2.2	[-4.9; 0.7]	2004-2019	-2.4	[-4.4; -0.4]	2004-2019
75+ y	-0.3	[-1.9; 1.4]	2004-2019	-2.4	[-5.1; 0.5]	2004-2019

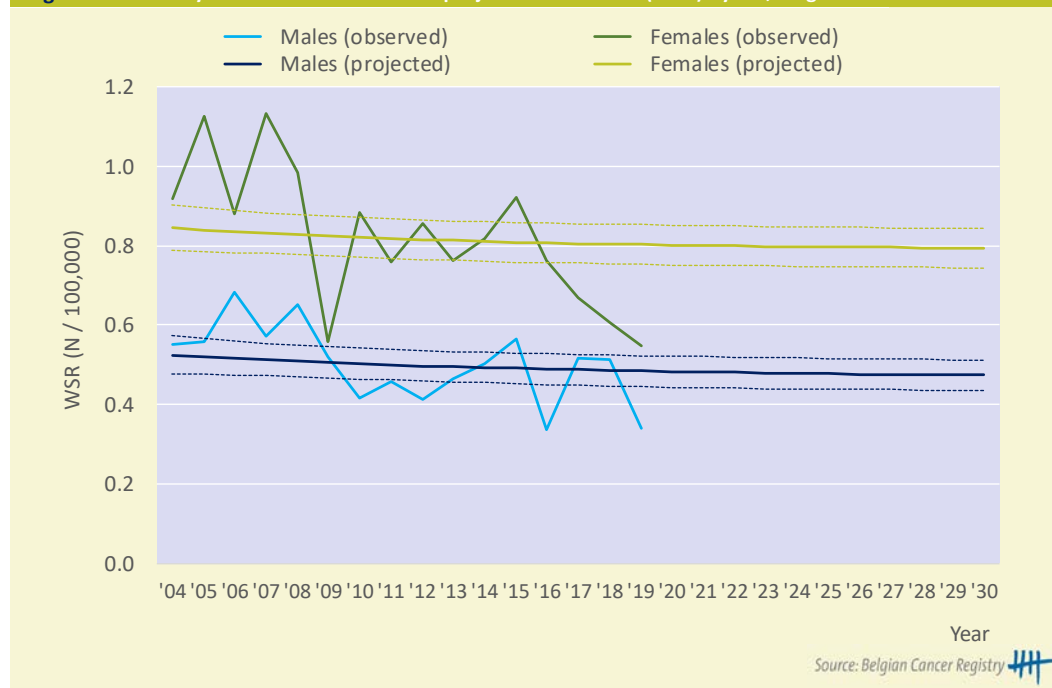
Source: Belgian Cancer Registry

AAPC: average annual percentage change

Period: When a joinpoint occurred, APC's are calculated for the period before and after the joinpoint. This column represents the corresponding time interval. AAPC's are always calculated over the entire study-period.

Incidence projections

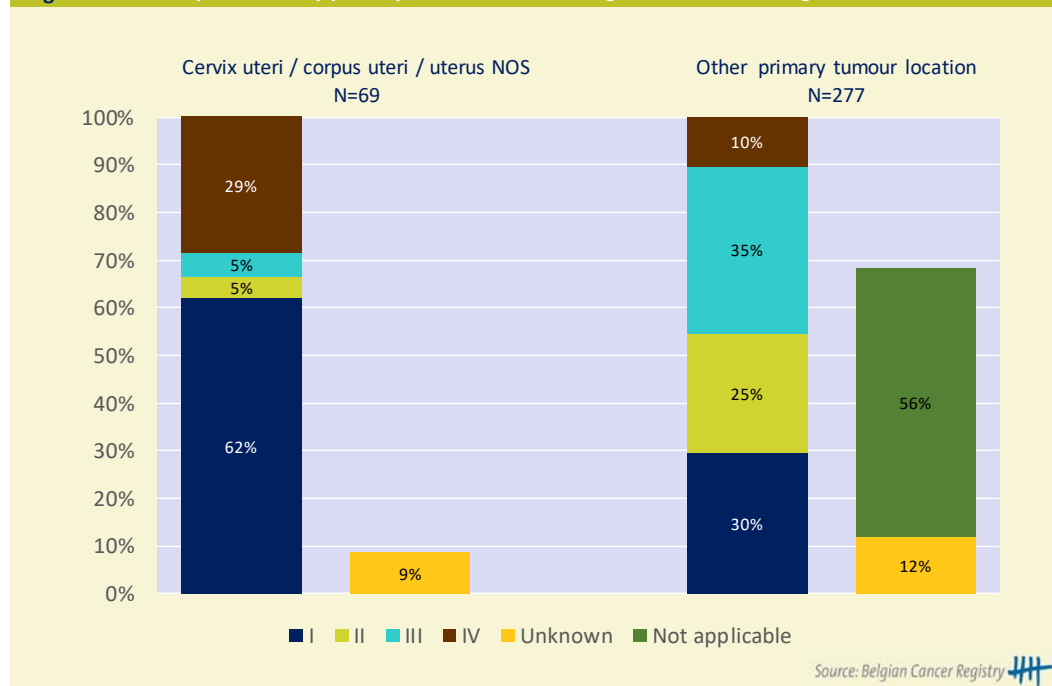
Figure 5 Leiomyosarcoma: Observed and projected* incidence (WSR) by sex, Belgium 2004-2030



WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

Stage distribution

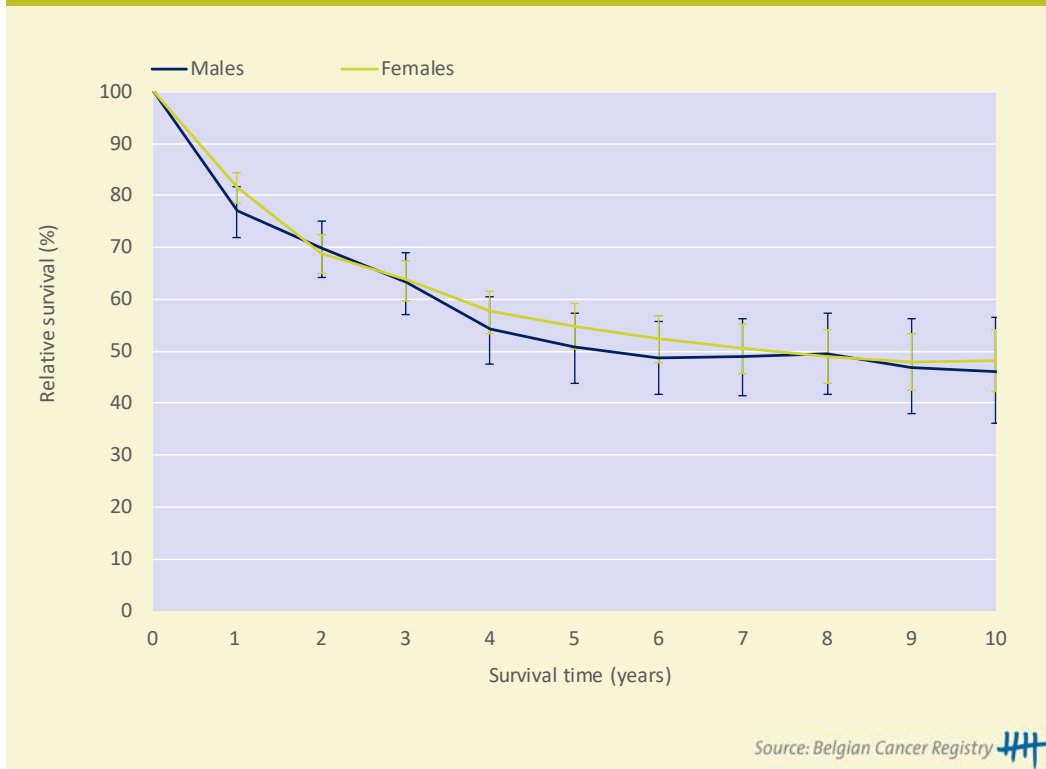
Figure 6 Leiomyosarcoma by primary tumour location: Stage distribution* , Belgium 2017-2019



* TNM Classification of Malignant Tumours, Eighth edition, UICC, 2017

Survival

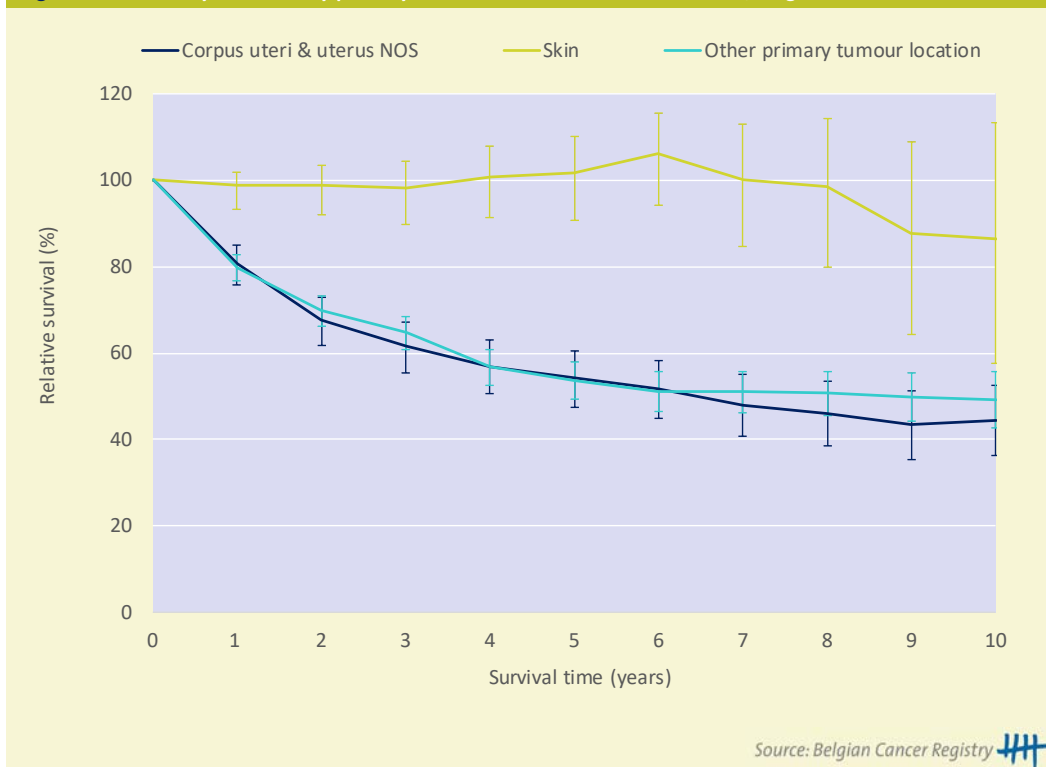
Figure 7 Leiomyosarcoma (excl. skin)[°]: Relative survival* by sex, Belgium 2010-2019



[°] Due to changes over time in the classification, leiomyosarcoma of the skin was excluded from the survival analysis. Before 2019 they were registered as malignant tumours. Currently they are mostly regarded as tumours with intermediate biologic potential.

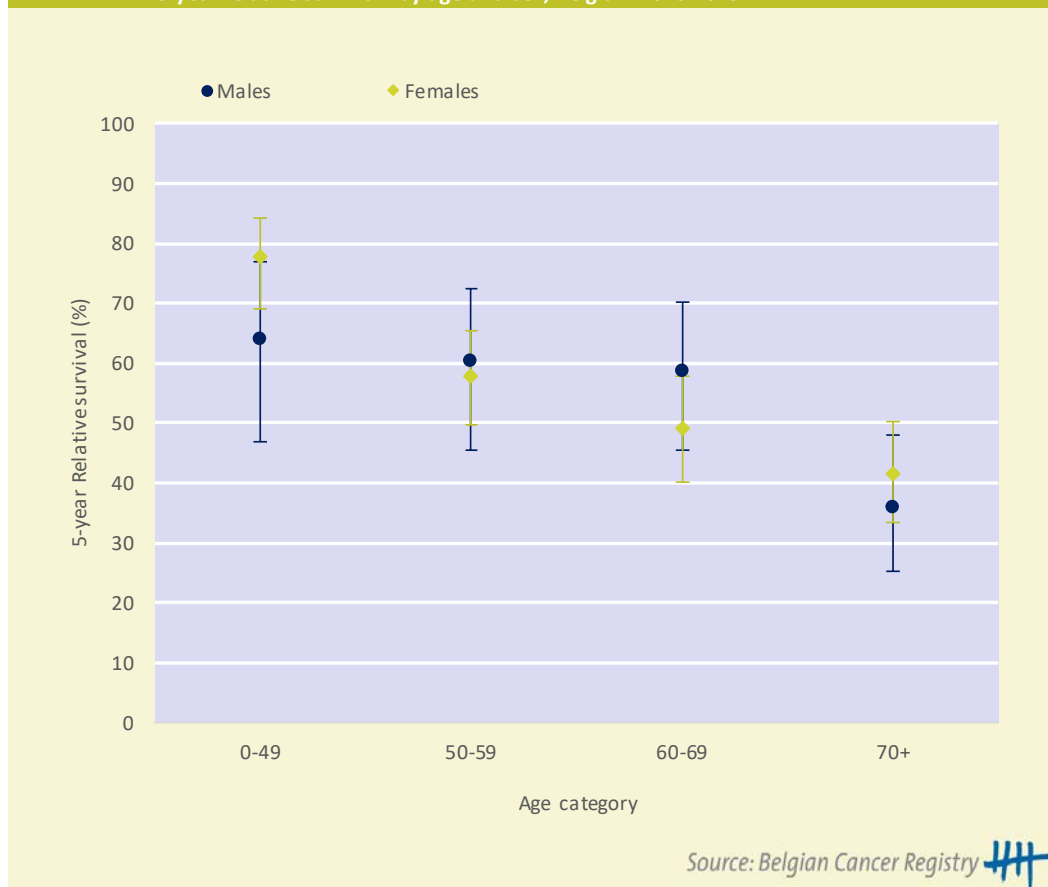
* The relative survival values are represented with 95% Confidence Intervals

Figure 8 Leiomyosarcoma by primary tumour location: Relative survival*, Belgium 2010-2019



* The relative survival values are represented with 95% Confidence Intervals

Figure 9 Leiomyosarcoma (excl. skin)[°]:
5-year relative survival* by age and sex, Belgium 2010-2019



[°] Due to changes over time in the classification, leiomyosarcoma of the skin was excluded from the survival analysis. Before 2019 they were registered as malignant tumours. Currently they are mostly regarded as tumours with intermediate biologic potential.

* The relative survival values are represented with 95% Confidence Intervals

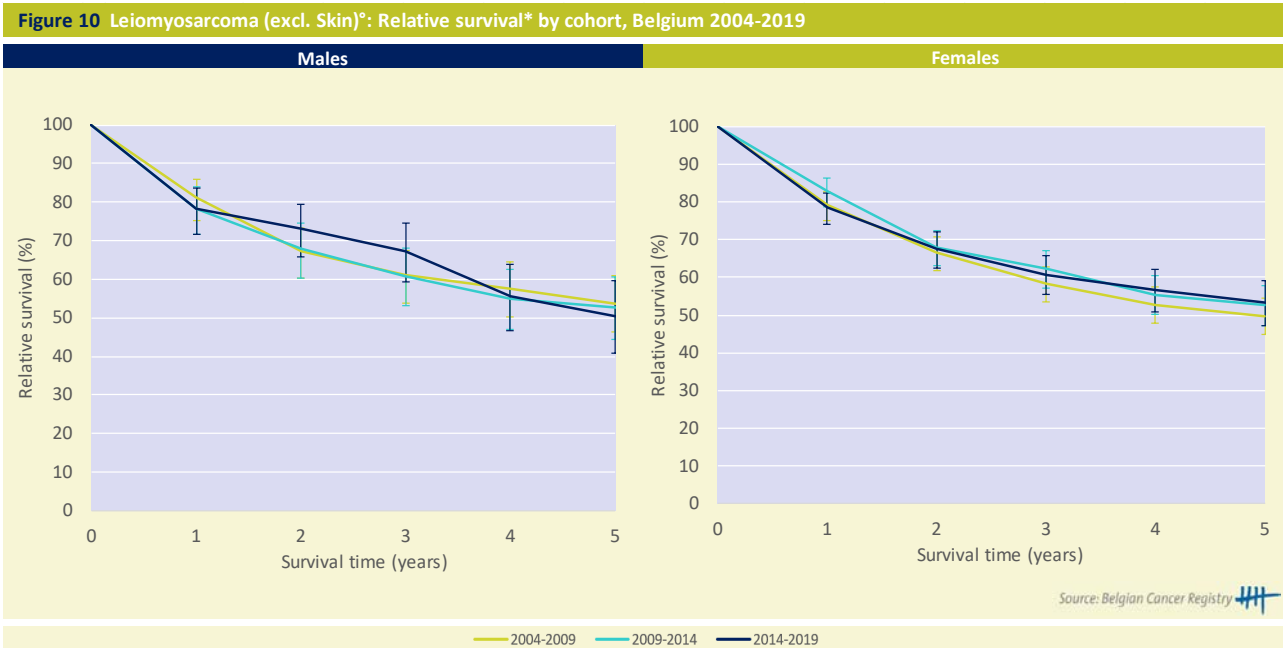
X years since diagnosis	N at risk	%
1 year	825	64.0
2 year	646	72.2
3 year	525	77.1

[°] Due to changes over time in the classification, leiomyosarcoma of the skin was excluded from the survival analysis. Before 2019 they were registered as malignant tumours. Currently they are mostly regarded as tumours with intermediate biologic potential.

* Unadjusted 5-yr relative survival probability conditional on surviving the first X years since diagnosis, %

* Interpretation in lay-man's terms: Given that a patient has already survived X years, what is the relative survival probability 5 years later.

Survival trends



[°] Due to changes over time in the classification, leiomyosarcoma of the skin was excluded from the survival analysis. Before 2019 they were registered as malignant tumours. Currently they are mostly regarded as tumours with intermediate biologic potential.

* The relative survival values are represented with 95% Confidence Intervals

3.2.5 RHABDOMYOSARCOMA

MAIN SUBTYPES:

- *Embryonal rhabdomyosarcoma (ERMS)*
- *Alveolar rhabdomyosarcoma (ARMS)*
- *Pleomorphic rhabdomyosarcoma*
- *Spindle cell / sclerosing rhabdomyosarcoma*
- *Rhabdomyosarcoma, NOS*

KEYNOTES

Incidence (table 1-2; figure 1-8)

- The age-standardised incidence of rhabdomyosarcoma is characterised by two peaks, corresponding to different rhabdomyosarcoma subtypes:
 - A first incidence peak in children and adolescents under the age of 20 years:
 - Rhabdomyosarcoma is the most frequent sarcoma type presenting in young children (<10 years) (See chapter 3.2 figure 4).
 - Typically the embryonal rhabdomyosarcoma subtype occurs more frequent, especially under the age of 10 years, followed by the alveolar rhabdomyosarcoma subtype.
 - A second -much smaller- incidence peak in the older population (60+ years):
 - In adults, subtypes other than embryonal and alveolar are more common, especially the pleomorphic adult-type and NOS rhabdomyosarcoma, and the spindle cell subtype to a lesser extent.
- Rhabdomyosarcoma is more common in males at both incidence peaks (male/female ratio of 1.4).
- The main primary tumour location of rhabdomyosarcomas is the trunk (28%) followed by head & neck. Especially in children and adolescents under the age of 20 years, these locations are predominant (See chapter 3.2 figure 7).

Survival (table 3; figure 9-12)

- The 5-year relative survival of patients with rhabdomyosarcoma:
 - strongly decreases with age (from 77% before 10 years to 27% after 20 years)
 - does not show consistent improvement over time (2004-2019)
- The 10-year relative survival rate is similar in males and females (~45%).

Table 1 Rhabdomyosarcoma: Overview of incidence, prevalence and survival by sex in Belgium				
Males				
Incidence	N	CR	WSR	
Incidence, 2010-2019	133	0.2	0.3	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	38	0.7	1.0	
10-year prevalence, 31.12.2019	62	1.1	1.5	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	133	45.9	[36.5;54.8]	
10-year relative survival, 2004-2019	209	43.7	[35.9;51.4]	
Females				
Incidence	N	CR	WSR	
Incidence, 2010-2019	105	0.2	0.2	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	40	0.7	0.7	
10-year prevalence, 31.12.2019	60	1.0	1.2	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	105	48.3	[36.7;59.1]	
10-year relative survival, 2004-2019	153	46.8	[37.8;55.5]	
Median age at diagnosis, 2010-2019 (y)	26 [Q1: 7; Q3: 65]			
M/F-ratio	1.4			

Source: Belgian Cancer Registry

N: number of new diagnoses

CR: crude (all ages) rate (N/100,000 person years)

WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

M/F-ratio: Male/Female ratio based on the age-standardised rates (2010-2019)

Incidence

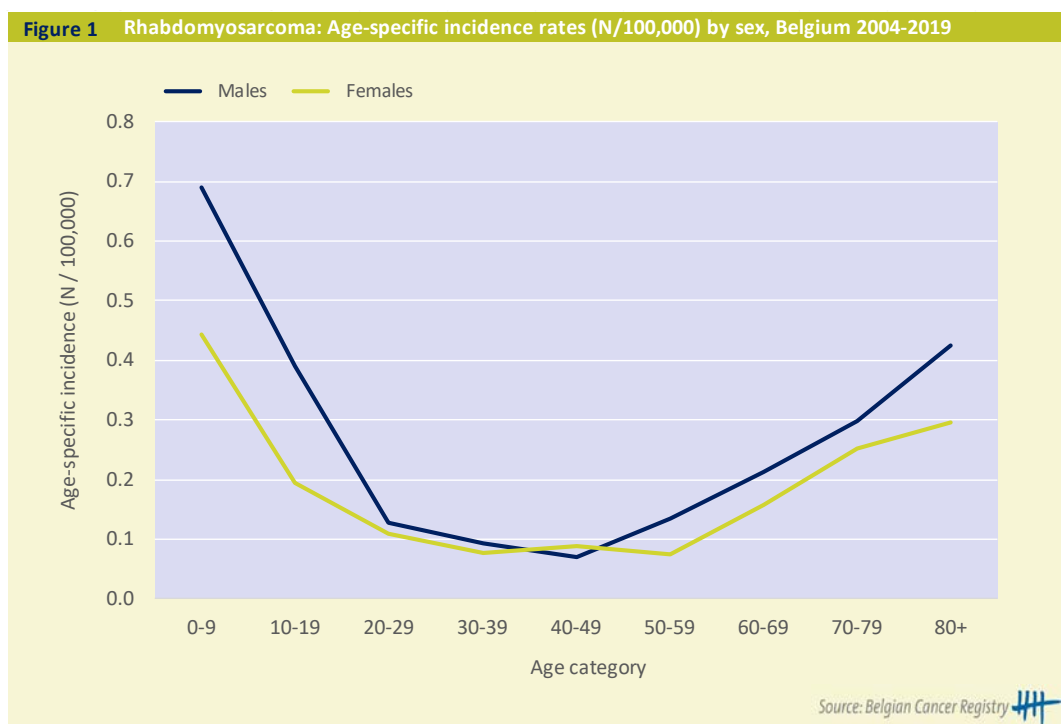
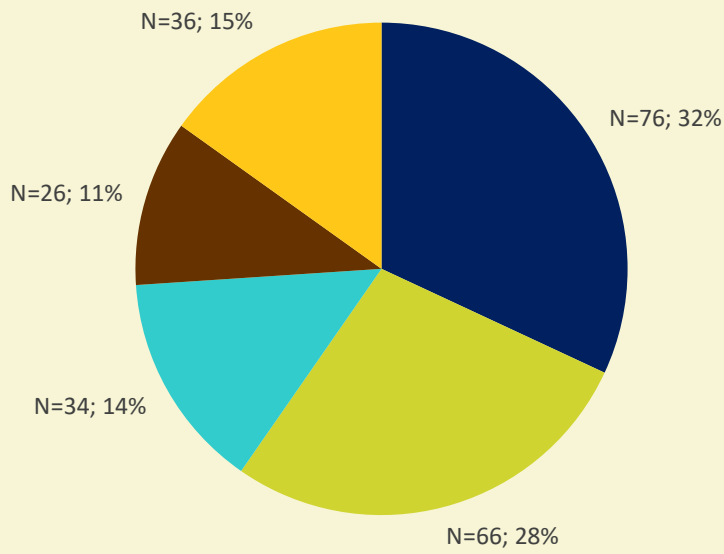


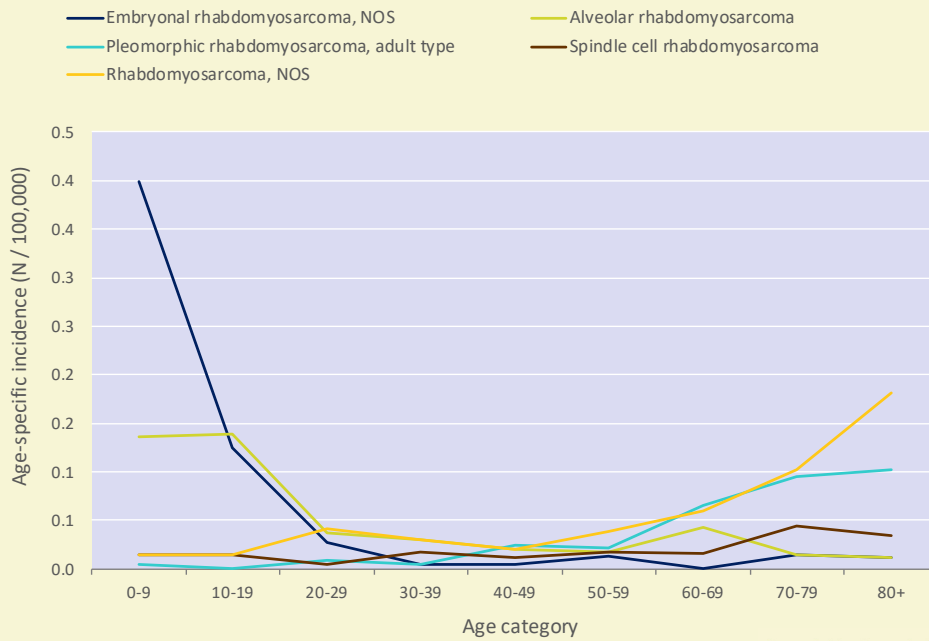
Figure 2 Rhabdomyosarcoma: Incidence distribution by subtype, Belgium 2010-2019



- Embryonal rhabdomyosarcoma, NOS
- Alveolar rhabdomyosarcoma
- Pleomorphic rhabdomyosarcoma, adult type
- Spindle cell rhabdomyosarcoma
- Rhabdomyosarcoma, NOS

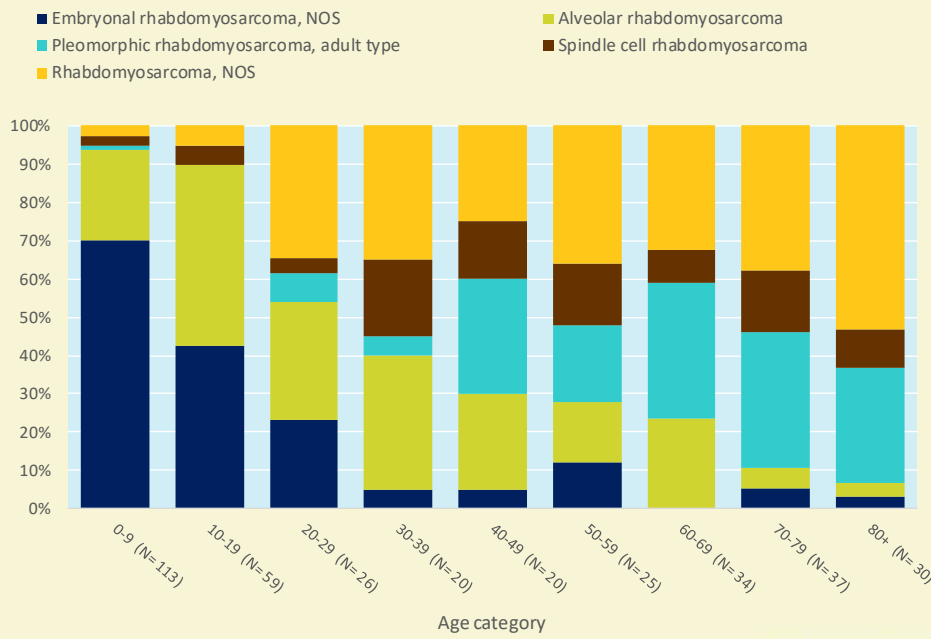
Source: Belgian Cancer Registry

Figure 3 Rhabdomyosarcoma: Age-specific incidence by subtype, Belgium 2004-2019



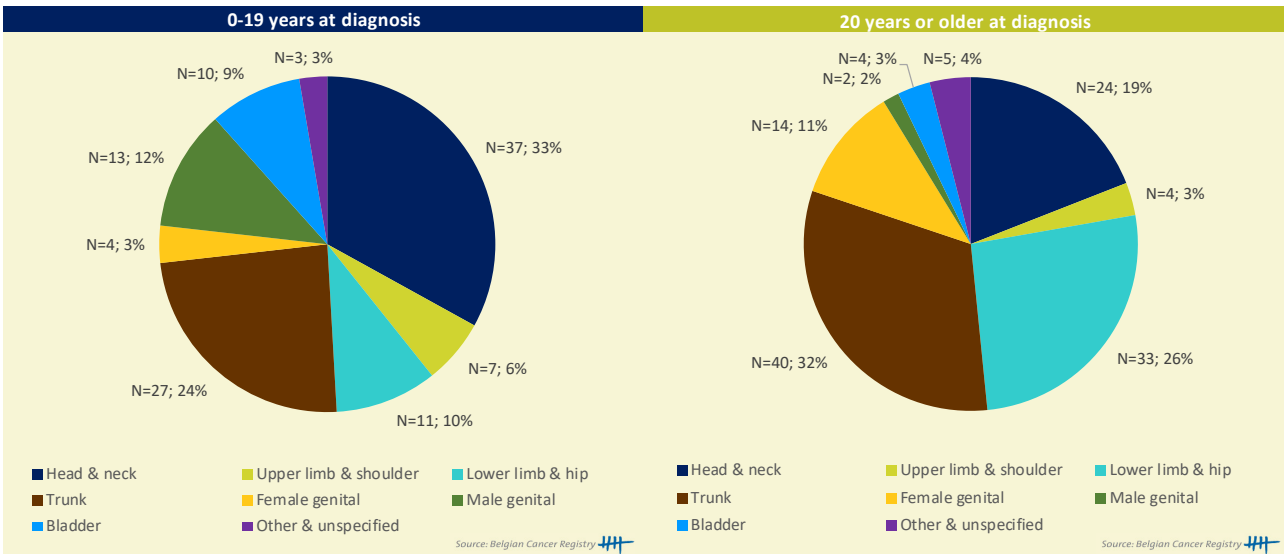
Source: Belgian Cancer Registry

Figure 4 Rhabdomyosarcoma: Subtype incidence distribution (%) by age category, Belgium 2004-2019



Source: Belgian Cancer Registry

Figure 5 Rhabdomyosarcoma: Incidence distribution by primary tumour location and age category, Belgium 2010-2019

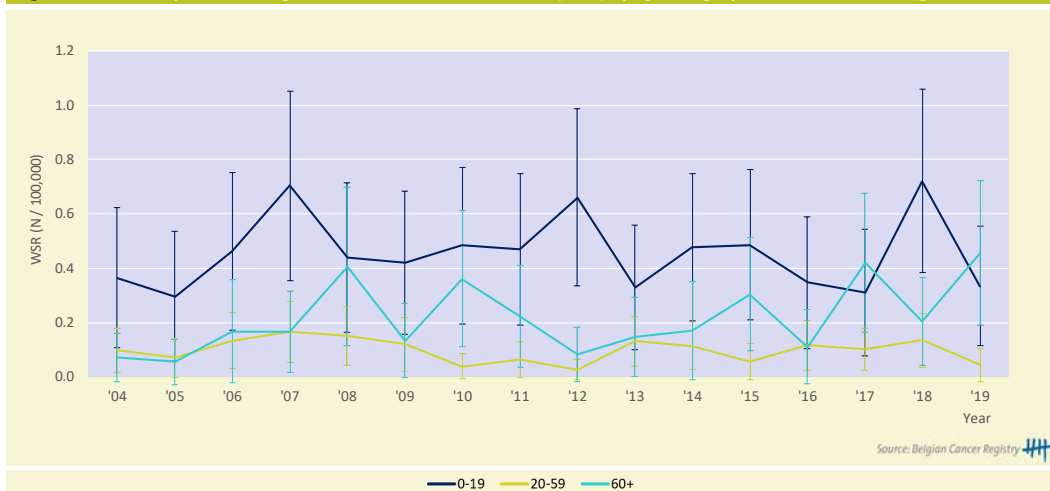


Source: Belgian Cancer Registry

Source: Belgian Cancer Registry

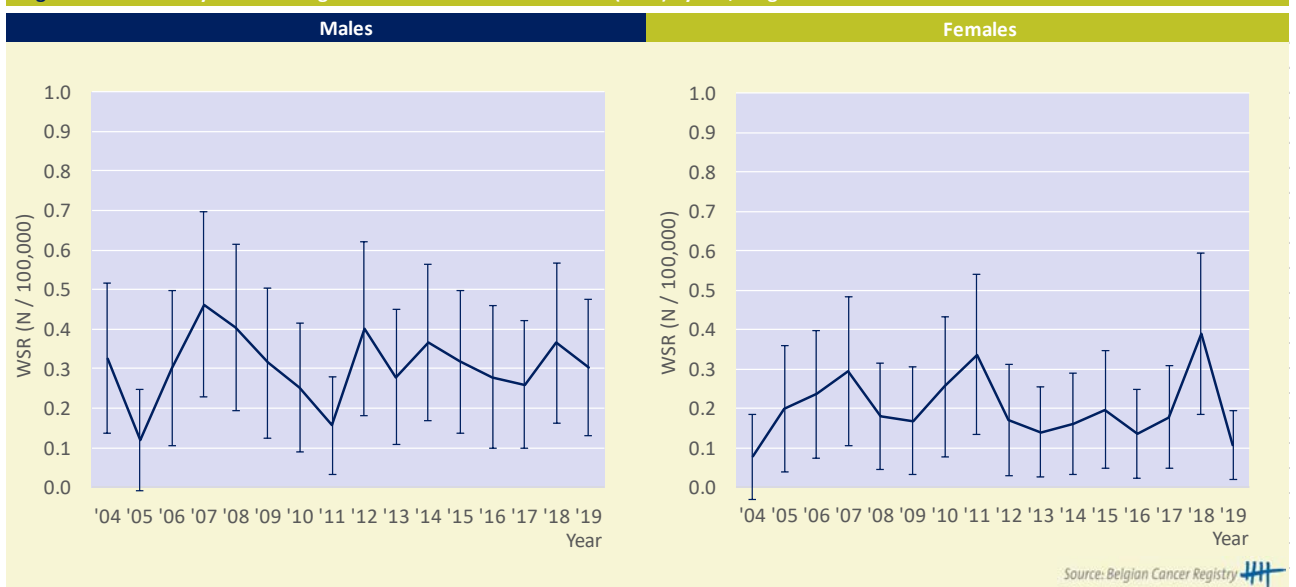
Incidence trends

Figure 6 Rhabdomyosarcoma: Age-standardised incidence rates* (WSR) by age category, males & females, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals.

Figure 7 Rhabdomyosarcoma: Age-standardised incidence rates* (WSR) by sex, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals.

Table 2 Rhabdomyosarcoma: Incidence trends by sex in Belgium, 2004-2019

Males			Females		
AAPC (%)	95%CI	Period	AAPC (%)	95%CI	Period
1.1	[-2.9; 5.3]	2004-2019	0.2	[-4.7; 5.3]	2004-2019

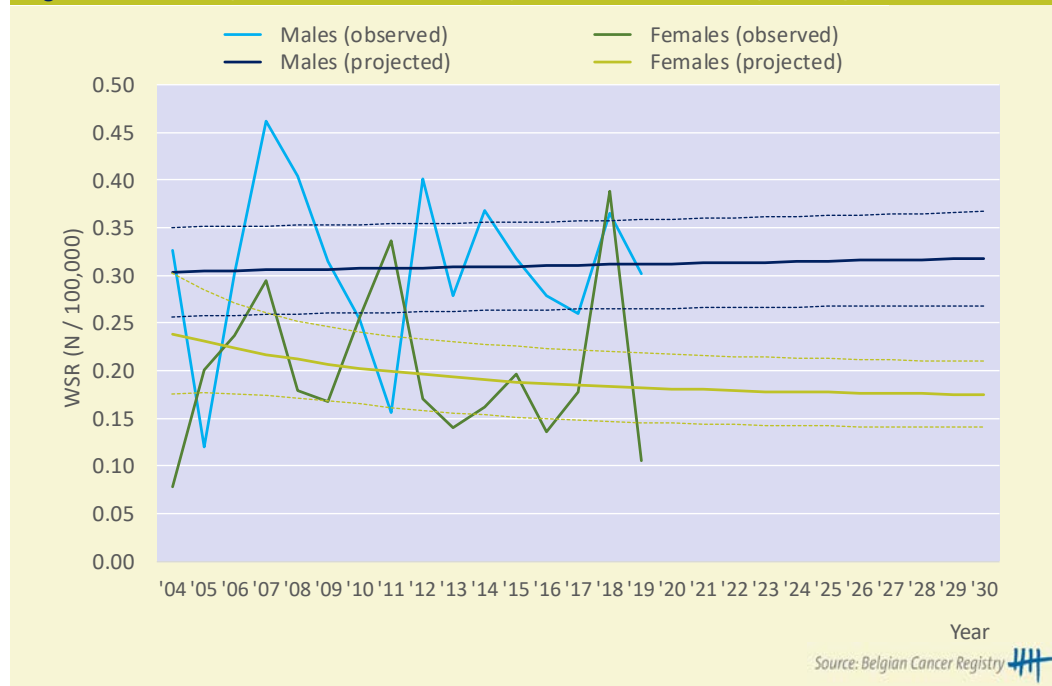
Source: Belgian Cancer Registry

AAPC: average annual percentage change

Period: when a joinpoint occurred, APC's are calculated for the period before and after the joinpoint. This column represents the corresponding time interval. AAPC's are always calculated over the entire study-period.

Incidence projections

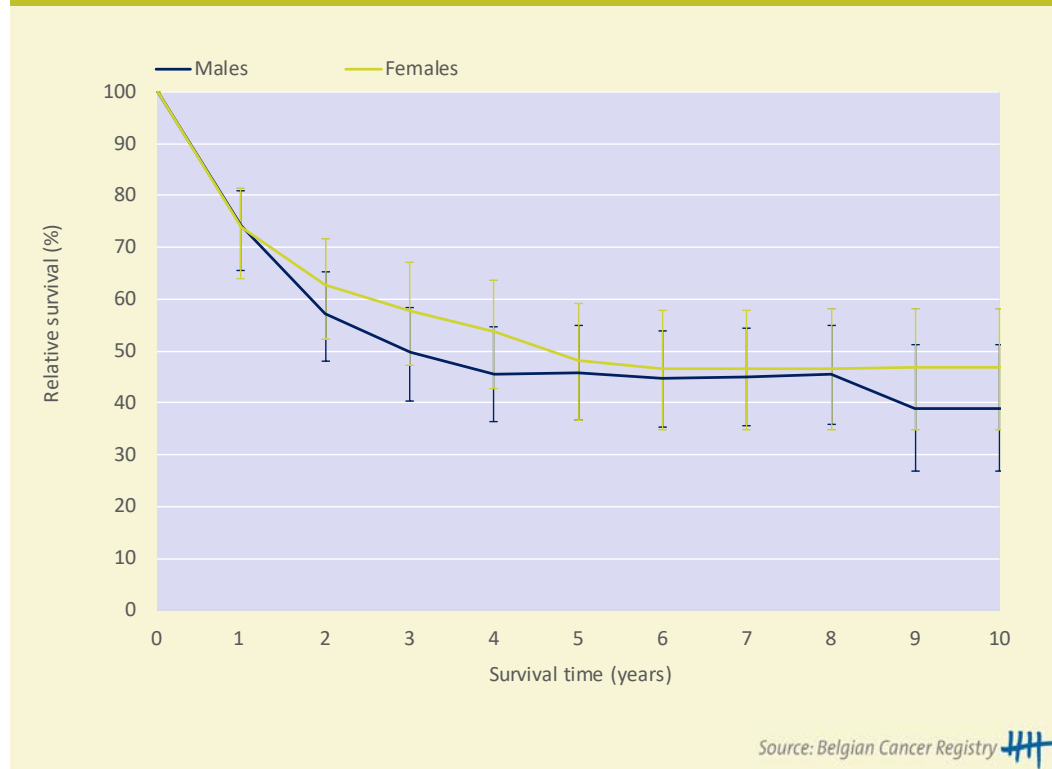
Figure 8 Rhabdomyosarcoma: Observed and projected* incidence (WSR) by sex, Belgium 2004-2030



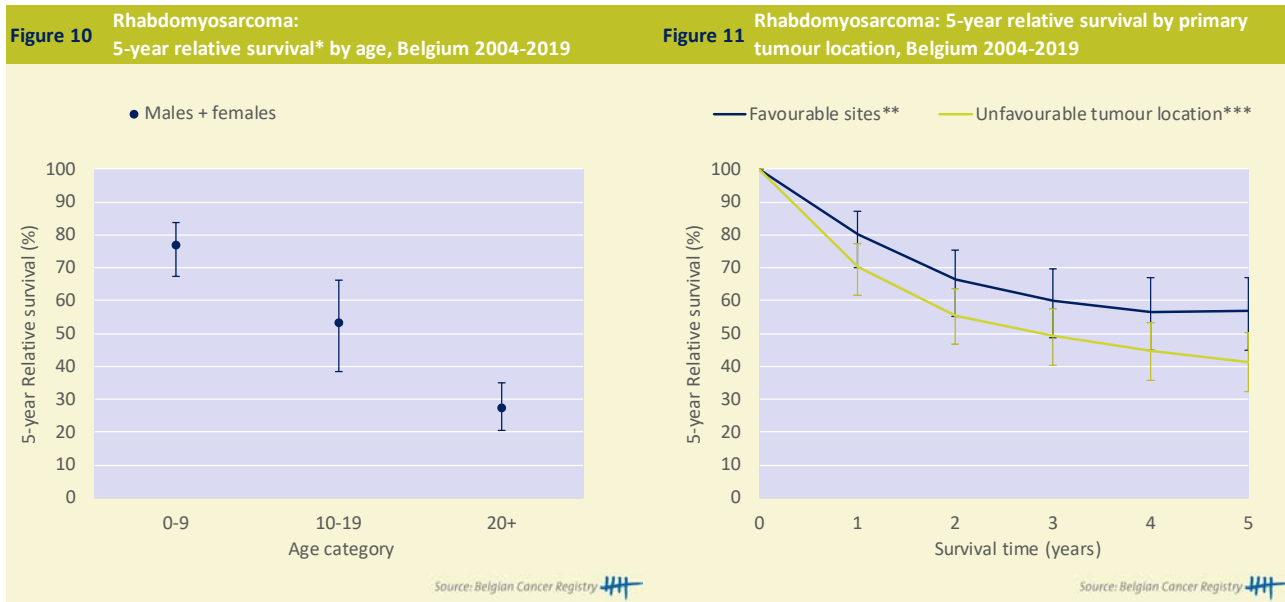
* WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

Survival

Figure 9 Rhabdomyosarcoma: Relative survival* by sex, Belgium 2010-2019



* The relative survival values are represented with 95% Confidence Intervals



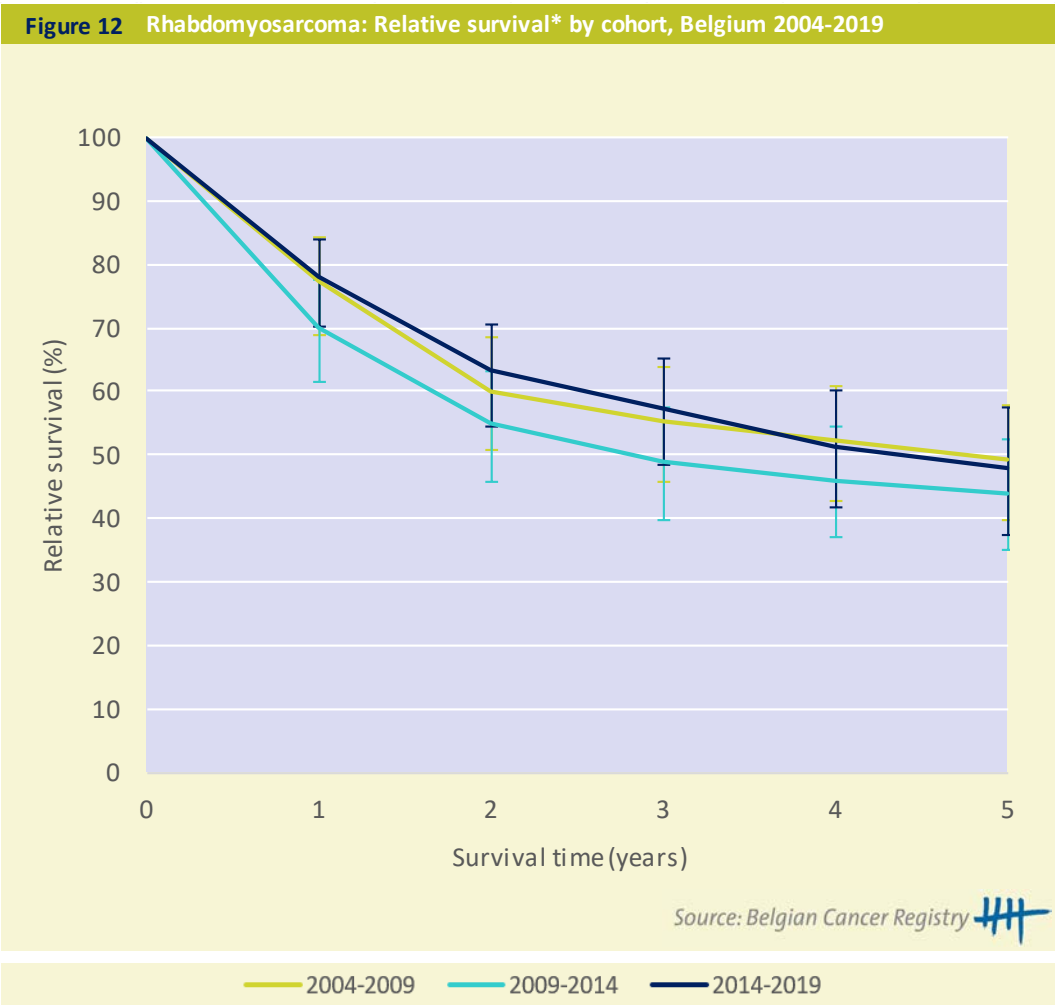
- * The relative survival values are represented with 95% Confidence Intervals
- ** Favourable tumour location: head- and neck, eye, genital and urinary except bladder and prostate
- *** Unfavourable tumour location: bladder, prostate, extremities, other or unknown sites

Table 3 Rhabdomyosarcoma: Conditional 5-year relative survival* in Belgium, 2010-2019

X years since diagnosis	N at risk	%
1 year	174	62.0
2 year	129	77.3
3 year	96	87.0

- * Unadjusted 5-yr relative survival probability conditional on surviving the first X years since diagnosis, %
- * Interpretation in lay-man's terms: Given that a patient has already survived X years, what is the relative survival probability 5 years later.

Survival trends



* The relative survival values are represented with 95% Confidence Intervals

3.2.6 PERIPHERAL NERVE SHEATH TUMOURS

KEYNOTES

Incidence (table 1-2; figure 1-5)

- Male/female ratio for peripheral nerve sheath tumours is 0.9
- The incidence of peripheral nerve sheath tumours increases with age in a linear way, similarly in males and females.
- One out of four diagnoses occurs in the lower limb and hip, followed by head & neck (17%) and thorax (14%).

Survival (table 3; figure 6-8)

- The 5-year relative survival of patients with peripheral nerve sheath tumours:
 - decreases with age for males (from 61% under the age of 50 years to 40% for 50+) while it remains stable for females.
 - does not show consistent improvement over time (2004-2019).
- The 10-year relative survival tends to be higher for females (62%) than males (46%).

Table 1 Peripheral nerve sheath tumours: Overview of incidence, prevalence and survival by sex in Belgium, 2010-2019				
Males				
Incidence	N	CR	WSR	
Incidence, 2010-2019	128	0.2	0.2	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	41	0.7	0.5	
10-year prevalence, 31.12.2019	66	1.2	0.8	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	128	48.8	[38.5;58.7]	
10-year relative survival, 2010-2019	128	46.2	[34.3;57.9]	
Females				
Incidence	N	CR	WSR	
Incidence, 2010-2019	143	0.3	0.2	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	45	0.8	0.5	
10-year prevalence, 31.12.2019	85	1.5	1.0	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	142	63.6	[53.6;72.4]	
10-year relative survival, 2010-2019	142	61.9	[49.5;73.1]	
Median age at diagnosis, 2010-2019 (y)	55 [Q1: 40; Q3: 70]			
M/F-ratio	0.9			

Source: Belgian Cancer Registry 

N: number of new diagnoses

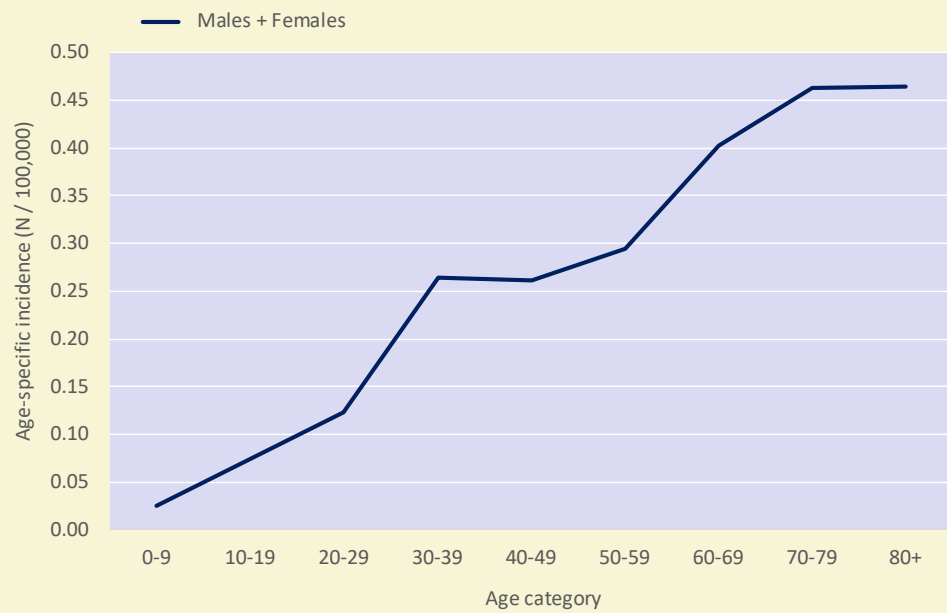
CR: crude (all ages) rate (N/100,000 person years)

WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

M/F-ratio: Male/Female ratio based on the age-standardised rates (2010-2019)

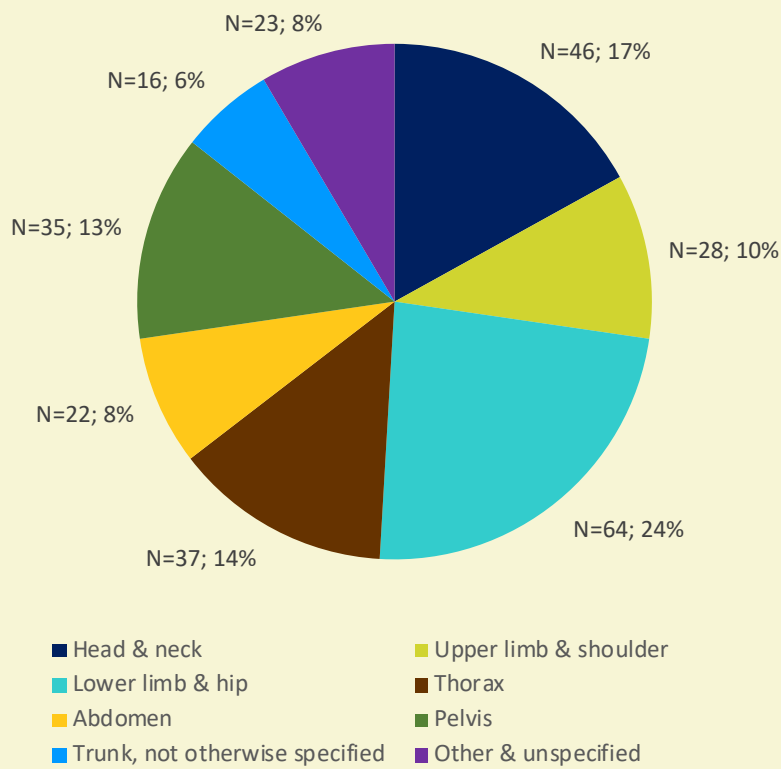
Incidence

Figure 1 Peripheral nerve sheath tumors: Age-specific incidence rates (N/100,000), Belgium 2004-2019



Source: Belgian Cancer Registry

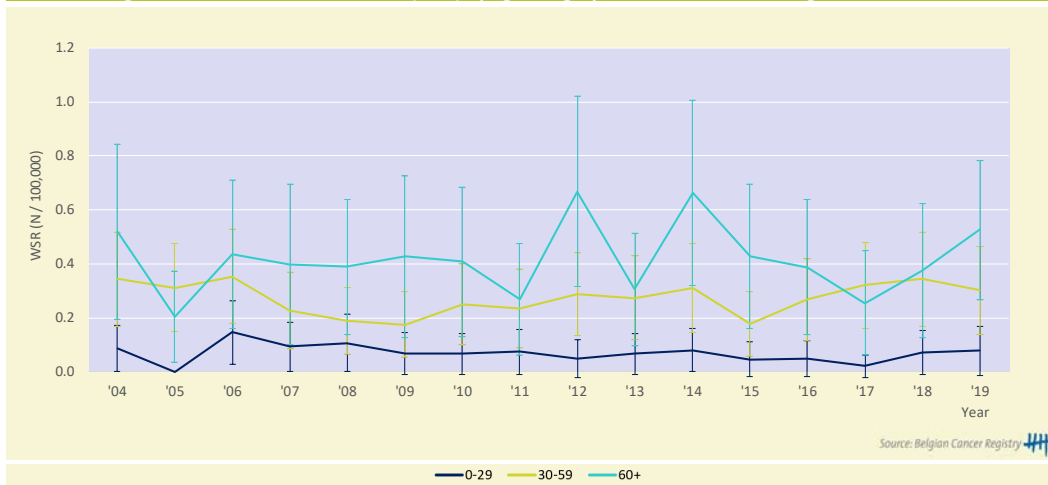
Figure 2 Peripheral nerve sheath tumours: Incidence distribution by primary tumour location, Belgium 2010-2019



Source: Belgian Cancer Registry

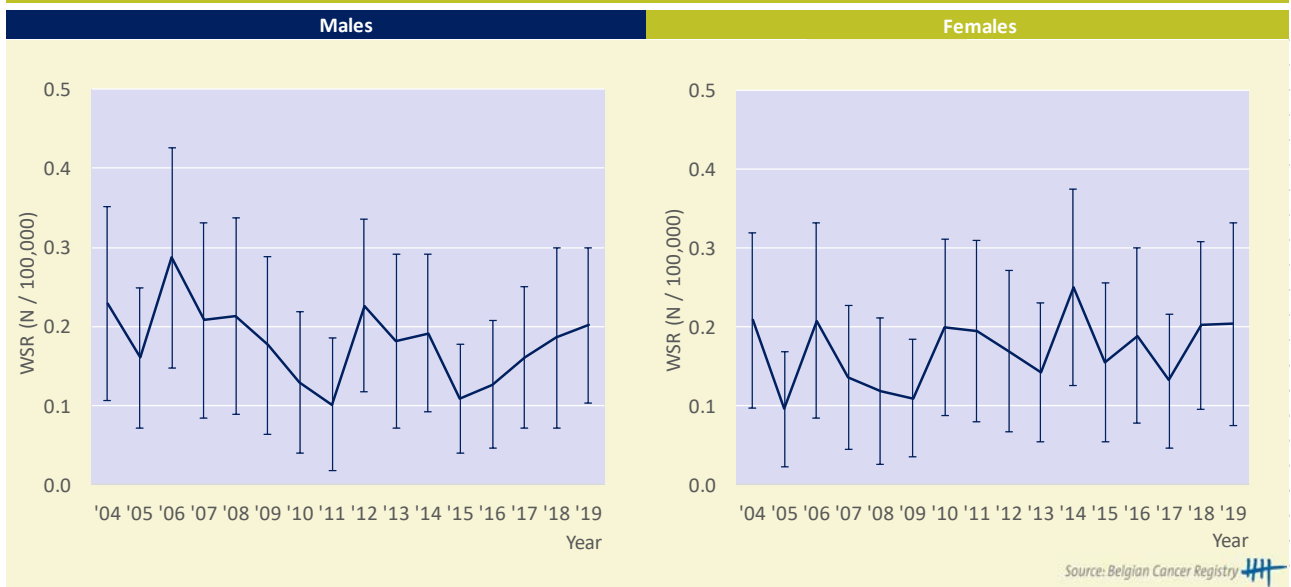
Incidence trends

Figure 3 Peripheral nerve sheath tumours: Age-standardised incidence rates* (WSR) by age category, males and females, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals.

Figure 4 Peripheral nerve sheath tumours: Age-standardised incidence rates* (WSR) by sex, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals.

Table 2 Peripheral nerve sheath tumours: Incidence trends by sex and age category in Belgium, 2004-2019

Age category	Males			Females		
	AAPC (%)	95%CI	Period	AAPC (%)	95%CI	Period
All ages	-1.9	[-5.0; 1.3]	2004-2019	1.8	[-1.3; 5.0]	2004-2019
0 - 29 y	-	-	-	-	-	-
30 - 59 y	0.5	[-3.8; 4.9]	2004-2019	0.4	[-3.8; 4.7]	2004-2019
60+ y	-0.7	[-5.8; 4.6]	2004-2019	3.8	[-4.5; 12.8]	2004-2019

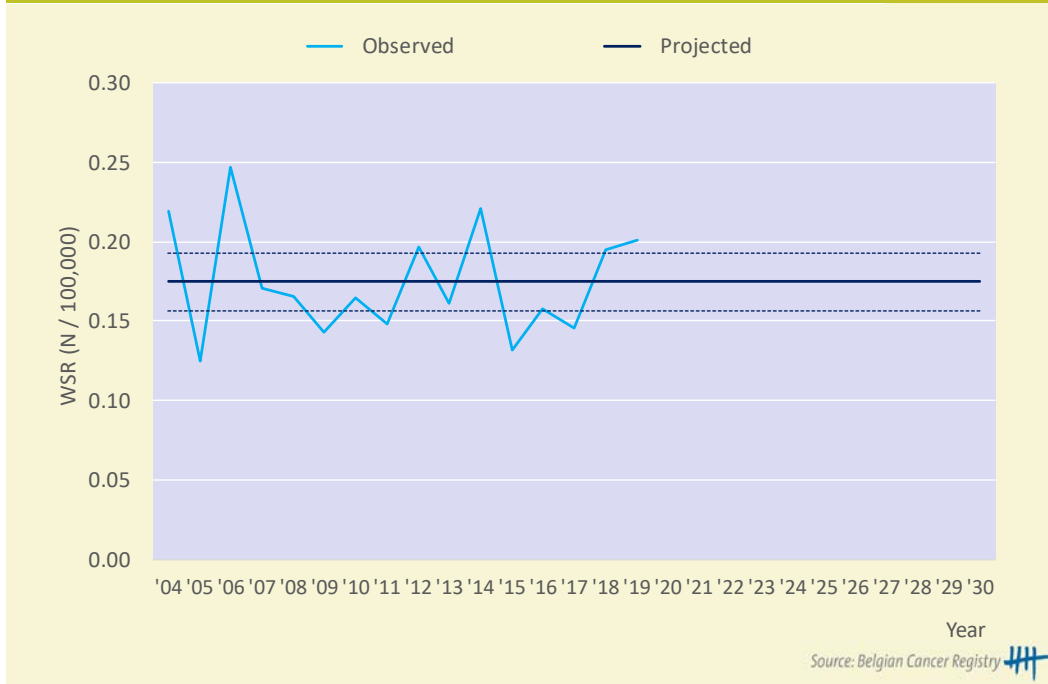
Source: Belgian Cancer Registry

AAPC: average annual percentage change

Period: when a joinpoint occurred, APC's are calculated for the period before and after the joinpoint. This column represents the corresponding time interval. AAPC's are always calculated over the entire study-period.

Incidence projections

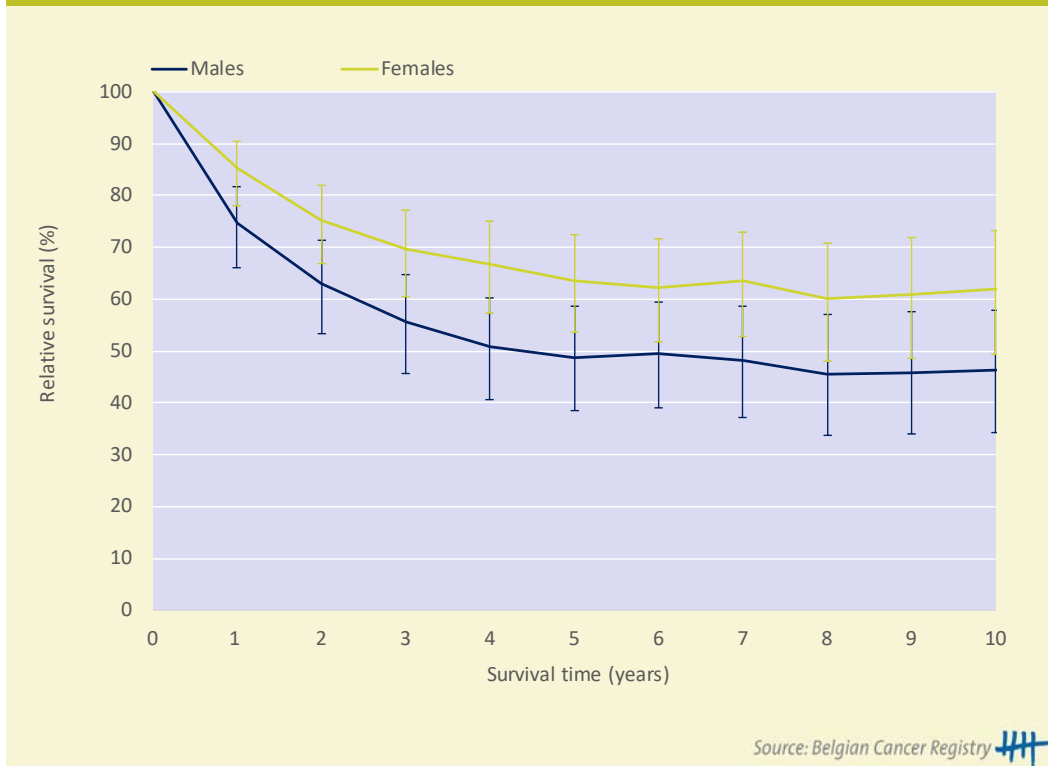
Figure 5 Peripheral nerve sheath tumours: Observed and projected* incidence (WSR), Belgium 2004-2030



WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

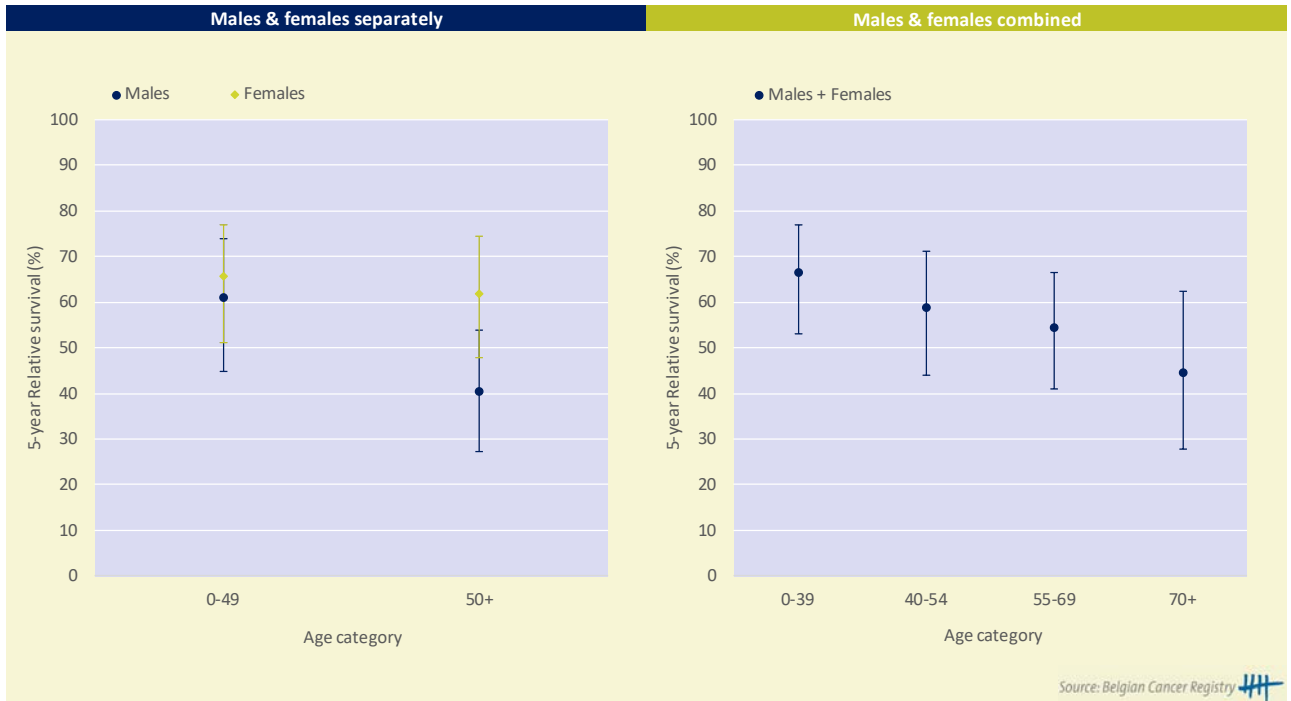
Survival

Figure 6 Peripheral nerve sheath tumours: Relative survival* by sex, Belgium 2010-2019



* The relative survival values are represented with 95% Confidence Intervals

Figure 7 Peripheral nerve sheath tumours: 5-year relative survival* by age and sex, Belgium 2010-2019



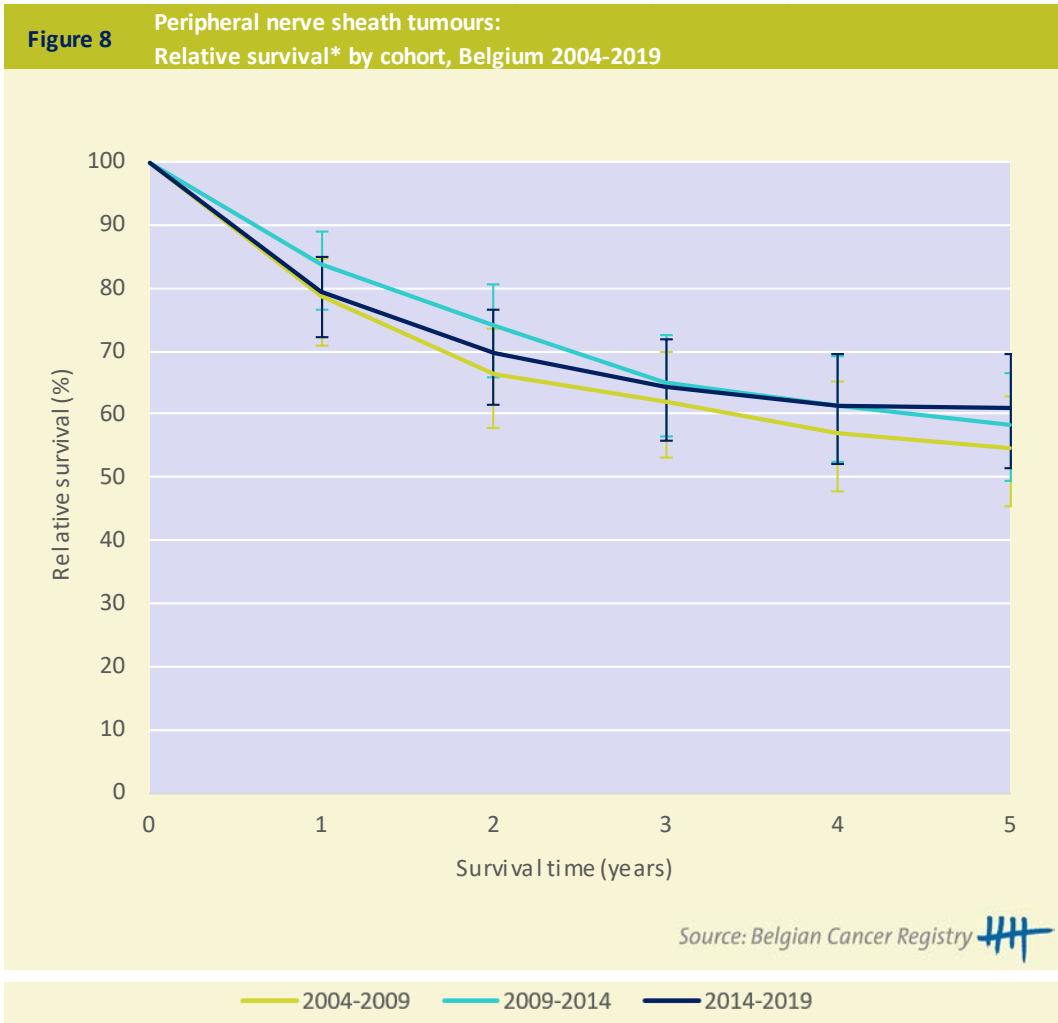
* The relative survival values are represented with 95% Confidence Intervals

Table 3 Peripheral nerve sheath tumours: Conditional 5-year relative survival* in Belgium, 2010-2019

X years since diagnosis	N at risk	%
1 year	212	70.0
2 year	162	81.0
3 year	131	84.4

* Unadjusted 5-yr relative survival probability conditional on surviving the first X years since diagnosis, %
 * Interpretation in lay-man's terms: Given that a patient has already survived X years, what is the relative survival probability 5 years later.

Survival trends



* The relative survival values are represented with 95% Confidence Intervals

3.2.7 OTHER TUMOURS OF UNCERTAIN DIFFERENTIATION

MAIN SUBTYPES:

- Synovial sarcoma
- Myoepithelioma
- Rhabdoid tumours
- Desmoplastic small round cell tumour
- Clear cell sarcoma
- Alveolar soft part sarcoma
- Epithelioid sarcoma, NOS

KEYNOTES

Incidence (table 1-2; figure 1-7)

- In young children (0-9 years) rhabdoid tumours are the most frequent subtype within the group other tumours of uncertain differentiation. Conversely, this subtype is extremely rare in older patients.
- From 10 years of age onwards, synovial sarcomas and desmoplastic small round cell tumours present at a relatively constant incidence rate, while the incidence of myoepithelioma (and of clear cell sarcoma to a lesser extent) increases with age.
- Desmoplastic small round cell tumours, clear cell sarcomas and alveolar soft part sarcomas are very rare tumours, with only 21, 42 and 9 cases respectively in 2010-2019.

Survival (table 3; figure 8-10)

- The 10-year relative survival of patients with these tumours of uncertain differentiation tends to be slightly better in females (59%) than males (49%). This could partly be explained by the relative survival being dependent on the subtype (see subchapters).

Other tumours of uncertain differentiation: Overview of incidence, prevalence and survival by sex in Belgium, 2010-2019				
Males				
Incidence	N	CR	WSR	
Incidence, 2010-2019	260	0.5	0.4	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	85	1.5	1.3	
10-year prevalence, 31.12.2019	144	2.5	2.2	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	257	57.6	[50.3;64.4]	
10-year relative survival, 2010-2019	257	49.4	[40.4;58.2]	
Females				
Incidence	N	CR	WSR	
Incidence, 2010-2019	255	0.4	0.4	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	90	1.5	1.3	
10-year prevalence, 31.12.2019	155	2.7	2.1	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	255	63.9	[56.5;70.6]	
10-year relative survival, 2010-2019	255	59.0	[49.0;68.5]	
Median age at diagnosis, 2010-2019 (y)	50 [Q1: 30; Q3: 66]			
M/F-ratio	1.2			

Source: Belgian Cancer Registry 

N: number of new diagnoses

CR: crude (all ages) rate (N/100,000 person years)

WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

M/F-ratio: Male/Female ratio based on the age-standardised rates (2010-2019)

Incidence

Figure 1 Other tumours of uncertain differentiation: Age-specific incidence rates (N/100,000) by sex, Belgium 2004-2019

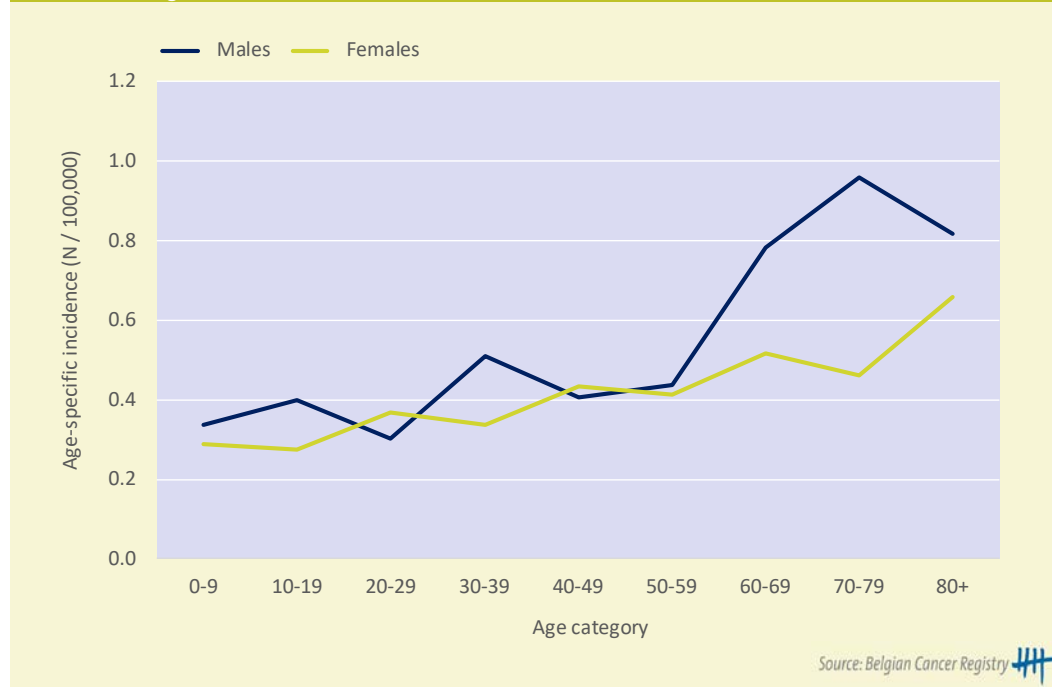
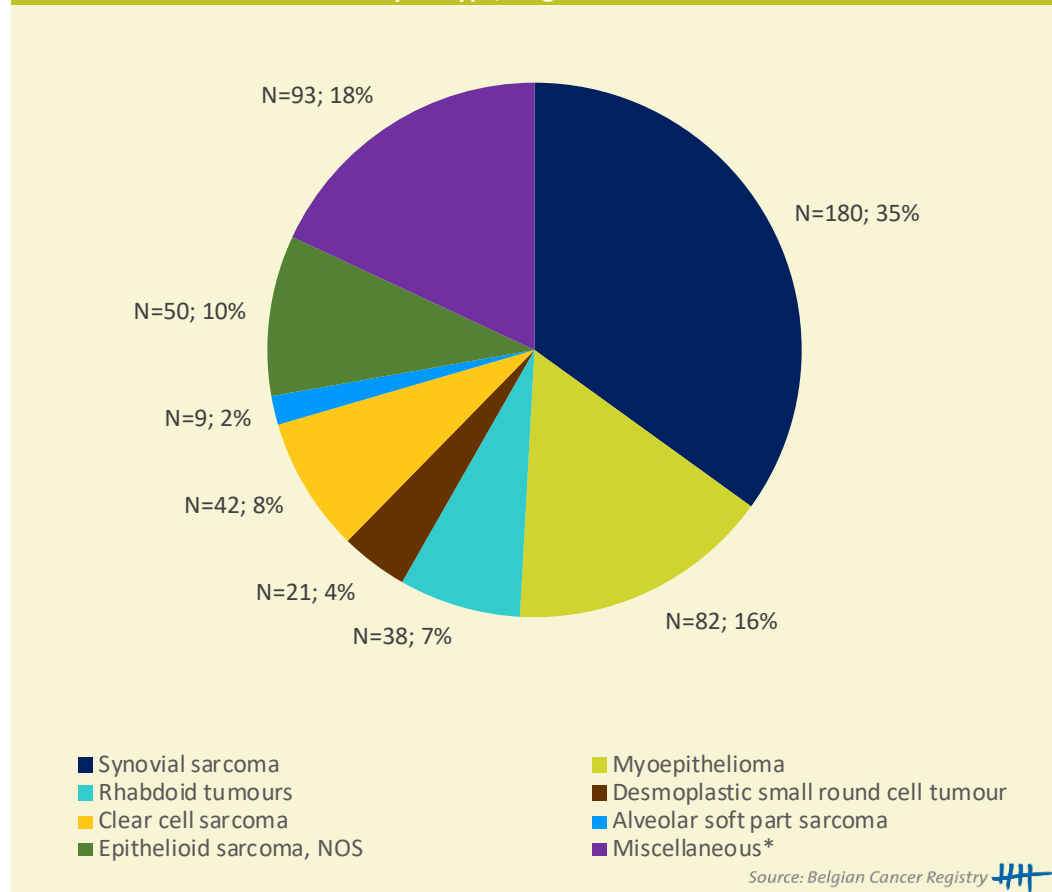
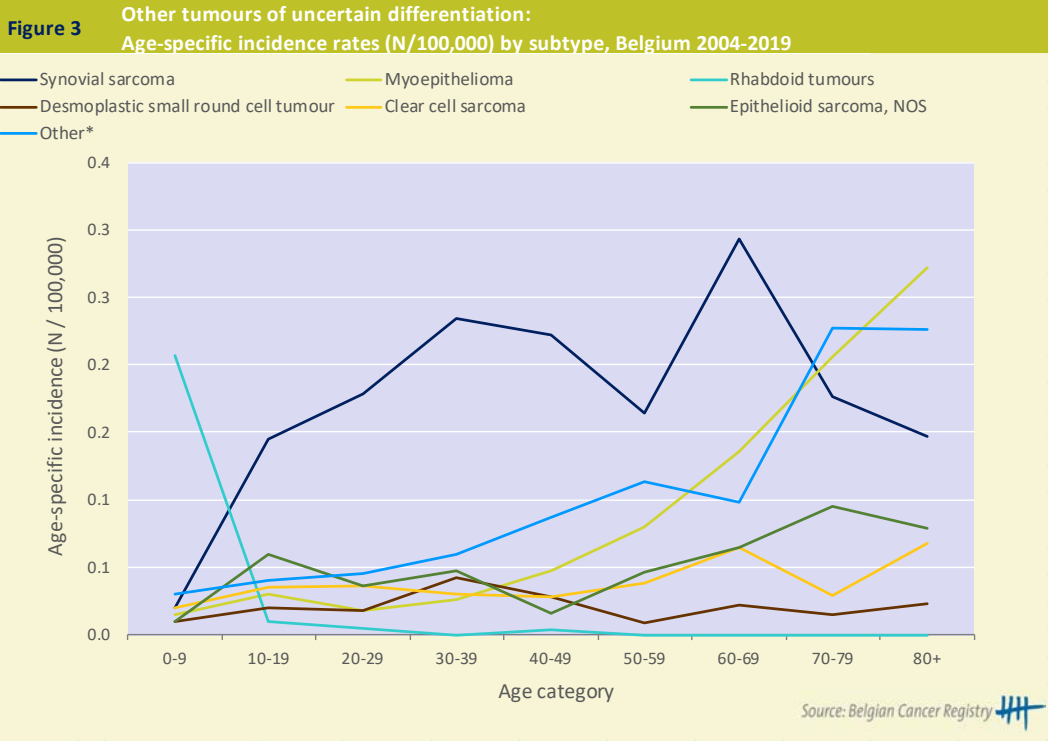


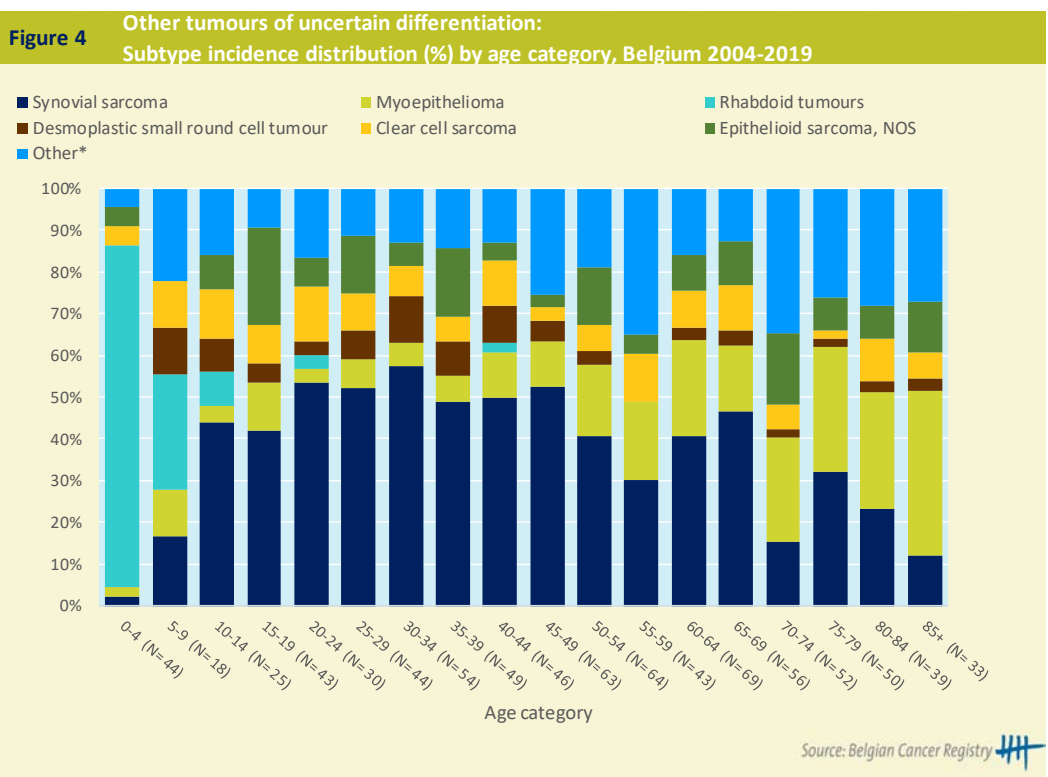
Figure 2 Other tumours of uncertain differentiation: Incidence distribution by subtype, Belgium 2010-2019



* This group includes malignant PEComa (N=15), malignant mesenchymoma (N=3), embryonal sarcoma (N=3), biphenotypic sinonasal sarcoma (N=5), intimal sarcoma (N=17), mixed tumour, malignant, NOS (N=43) and malignant ossifying fibromyxoid tumour (N=7).



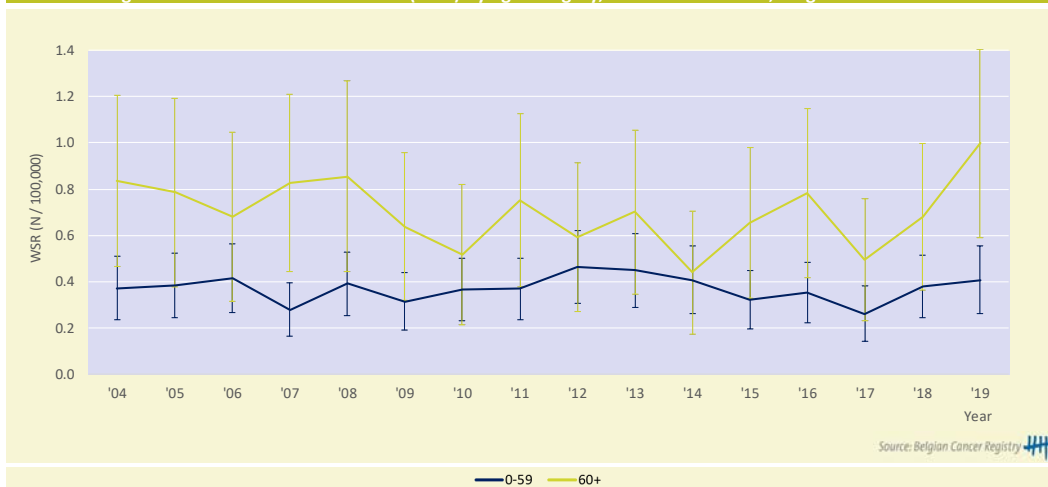
* The group with other tumours contains alveolar soft part sarcoma, malignant PEComa, malignant mesenchymoma, embryonal sarcoma, biphenotypic sinonasal sarcoma, intimal sarcoma, mixed tumour malignant NOS and malignant ossifying fibromyxoid tumour.



* The group with other tumours contains alveolar soft part sarcoma, malignant PEComa, malignant mesenchymoma, embryonal sarcoma, biphenotypic sinonasal sarcoma, intimal sarcoma, mixed tumour malignant NOS and malignant ossifying fibromyxoid tumour.

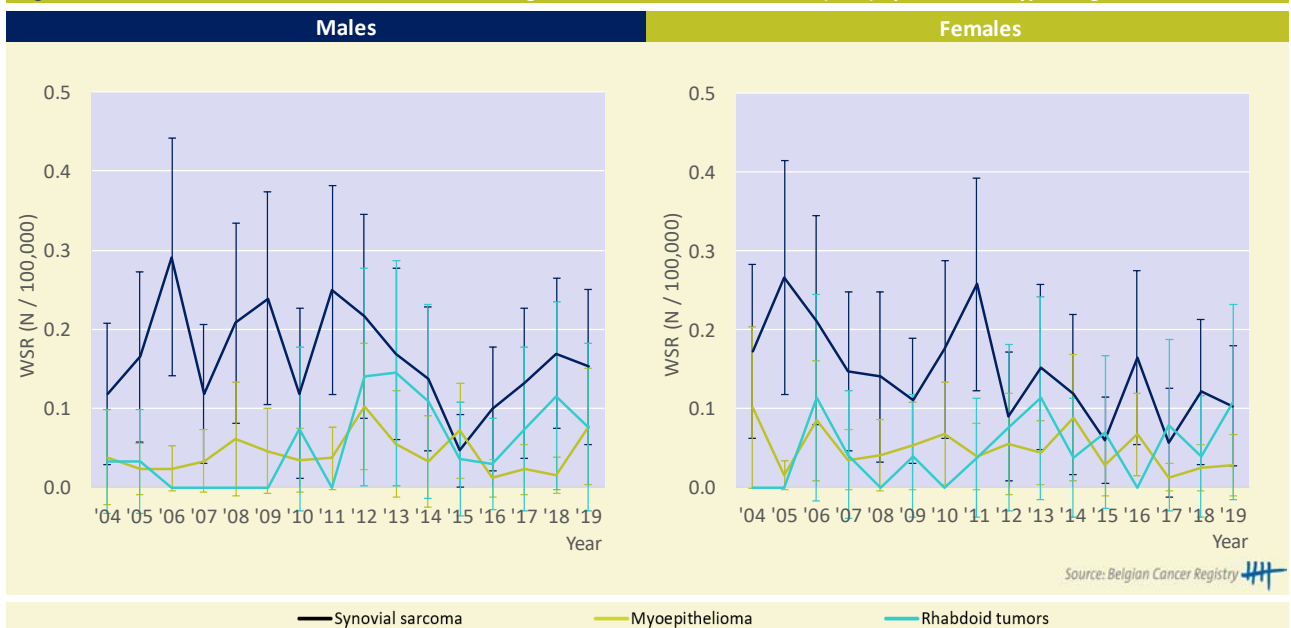
Incidence trends

Figure 5 Other tumours of uncertain differentiation: Age-standardised incidence rates* (WSR) by age category, males and females, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals.

Figure 6 Other tumours of uncertain differentiation: Age-standardised incidence rates* (WSR) by sex and subtype, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals.

Table 2 Other tumours of uncertain differentiation: Incidence trends by sex and age category in Belgium, 2004-2019

Age category	Males			Females		
	AAPC (%)	95%CI	Period	AAPC (%)	95%CI	Period
All ages	0.3	[-1.9; 2.5]	2004-2019	-1.1	[-2.9; 0.8]	2004-2019
0 - 59 y	1.4	[-1.1; 4.0]	2004-2019	-1.8	[-3.9; 0.4]	2004-2019
60+ y	-4.1	[-8.2; 0.2]	2004-2019	2.3	[-1.2; 6.1]	2004-2019

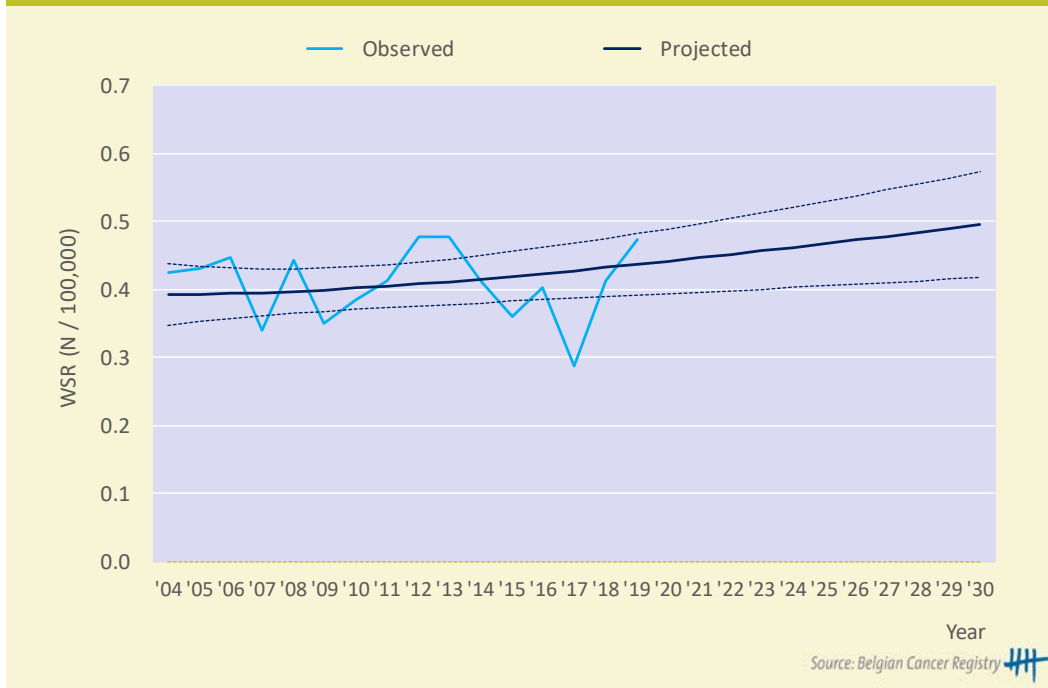
Source: Belgian Cancer Registry

AAPC: average annual percentage change

Period: when a joinpoint occurred, APC's are calculated for the period before and after the joinpoint. This column represents the corresponding time interval. AAPC's are always calculated over the entire study-period.

Incidence projections

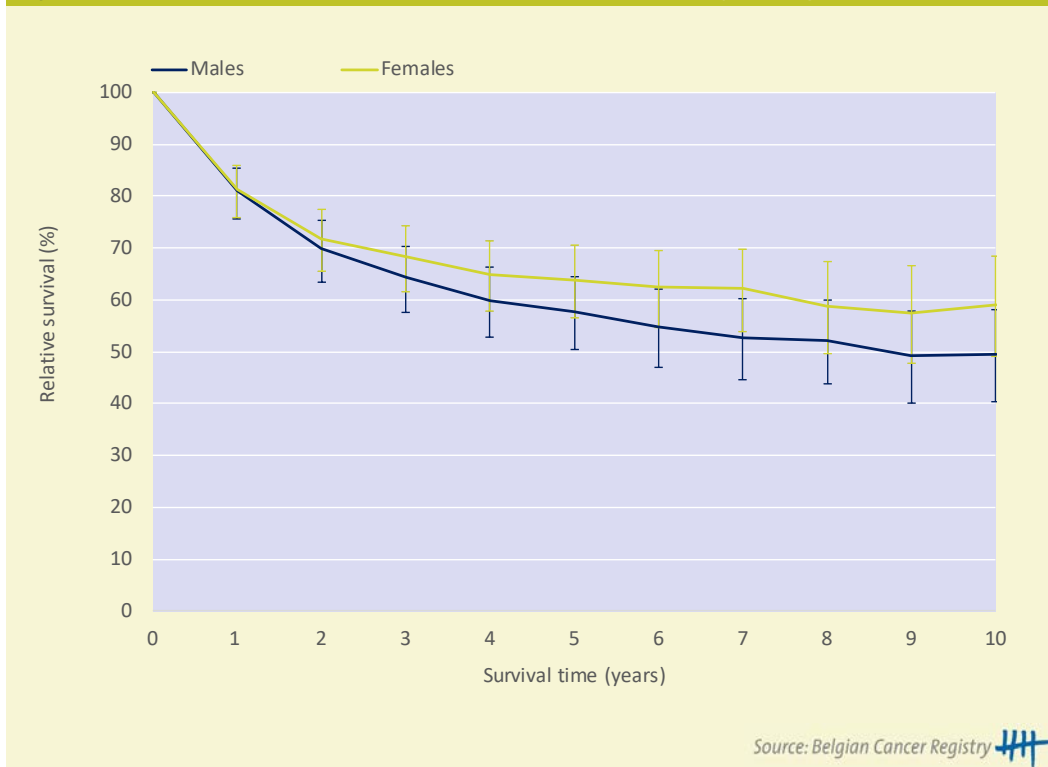
Figure 7 Other tumours of uncertain differentiation: Observed and projected* incidence (WSR), Belgium 2004-2030



WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

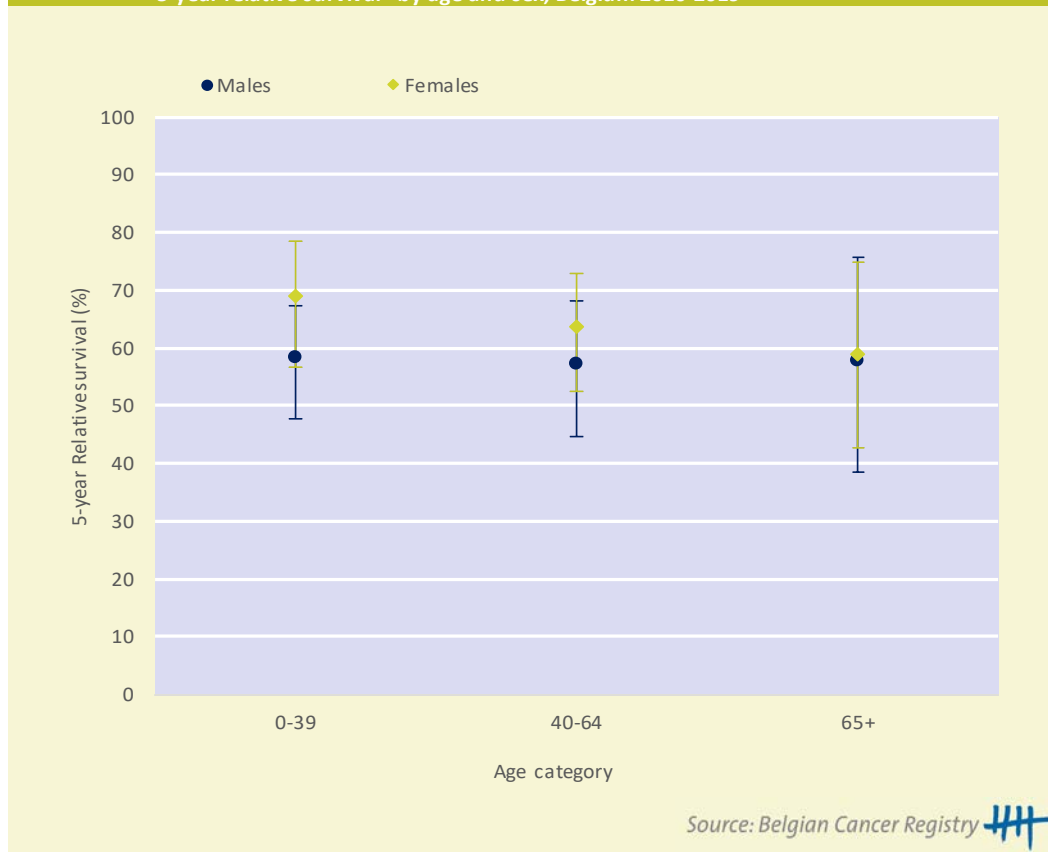
Survival

Figure 8 Other tumours of uncertain differentiation: Relative survival* by sex, Belgium 2010-2019



* The relative survival values are represented with 95% Confidence Intervals

Figure 9 Other tumours of uncertain differentiation:
5-year relative survival* by age and sex, Belgium 2010-2019



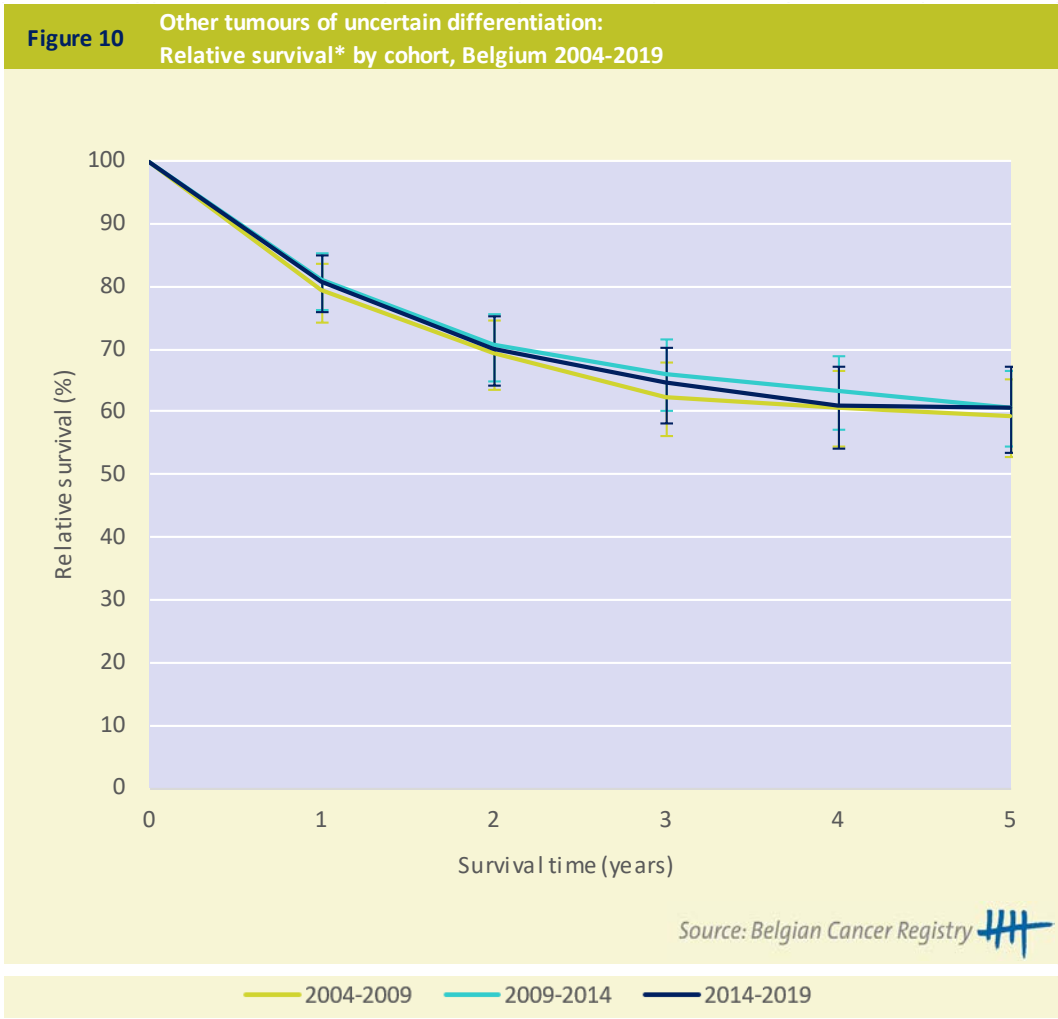
* The relative survival values are represented with 95% Confidence Intervals

Table 3 Other tumours of uncertain differentiation: Conditional
5-year relative survival* in Belgium, 2010-2019

X years since diagnosis	N at risk	%
1 year	408	72.1
2 year	322	81.0
3 year	249	83.7

* Unadjusted 5-yr relative survival probability conditional on surviving the first X years since diagnosis, %
* Interpretation in lay-man's terms: Given that a patient has already survived X years, what is the relative survival probability 5 years later.

Survival trends



* The relative survival values are represented with 95% Confidence Intervals

3.2.7.1 SYNOVIAL SARCOMA

KEYNOTES

Incidence (table 1-2; figure 1-4)

- The age-specific incidence of synovial sarcomas is relatively stable in adults from the age of 20 years onwards.
- More than one third (39%) of the synovial sarcomas occur in the lower limbs and hip, followed by the thorax (27%).
- The decreasing incidence trend, especially in females (average annual percentage change: -5.5%), may be partly explained by more accurate diagnosis and registration. Based on the incidence projections, the incidence rates (WSR) are expected to remain stable in the future.

Survival (table 3; figure 5-7)

- The relative survival of patients with synovial sarcoma does not show a consistent difference between males and females.
- The 5-year relative survival rate:
 - decreases with age from 79% under the age of 40 to 52% in patients older than 60 years.
 - tends to increase over time from 61% in 2004-2009 to 68% in 2014-2019 (attention for small absolute number of cases and large confidence intervals).

Table 1 Synovial sarcoma: Overview of incidence, prevalence and survival by sex, Belgium, 2010-2019

Males				
Incidence	N	CR	WSR	
Incidence, 2010-2019	96	0.2	0.1	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	31	0.5	0.4	
10-year prevalence, 31.12.2019	58	1.0	0.8	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	94	64.1	[51.7;74.4]	
10-year relative survival, 2010-2019	94	55.4	[40.3;68.7]	
Females				
Incidence	N	CR	WSR	
Incidence, 2010-2019	84	0.1	0.1	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	29	0.5	0.4	
10-year prevalence, 31.12.2019	59	1.0	0.9	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	84	65.1	[52.7;75.3]	
10-year relative survival, 2010-2019	84	61.0	[47.2;72.6]	
Median age at diagnosis, 2010-2019 (y)	47 [Q1: 31; Q3: 62]			
M/F-ratio	1.1			

Source: Belgian Cancer Registry 

N: number of new diagnoses

CR: crude (all ages) rate (N/100,000 person years)

WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

M/F-ratio: Male/Female ratio based on the age-standardised rates (2010-2019)

Incidence

Figure 1 Synovial sarcoma: Age-specific incidence rates (N/100,000), Belgium 2004-2019

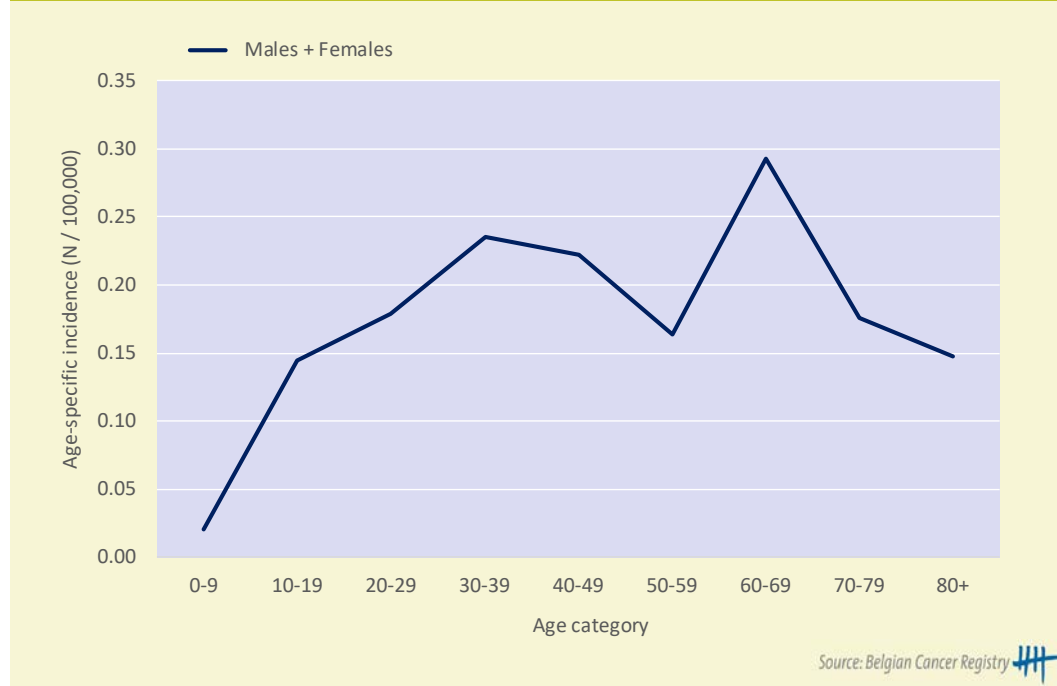
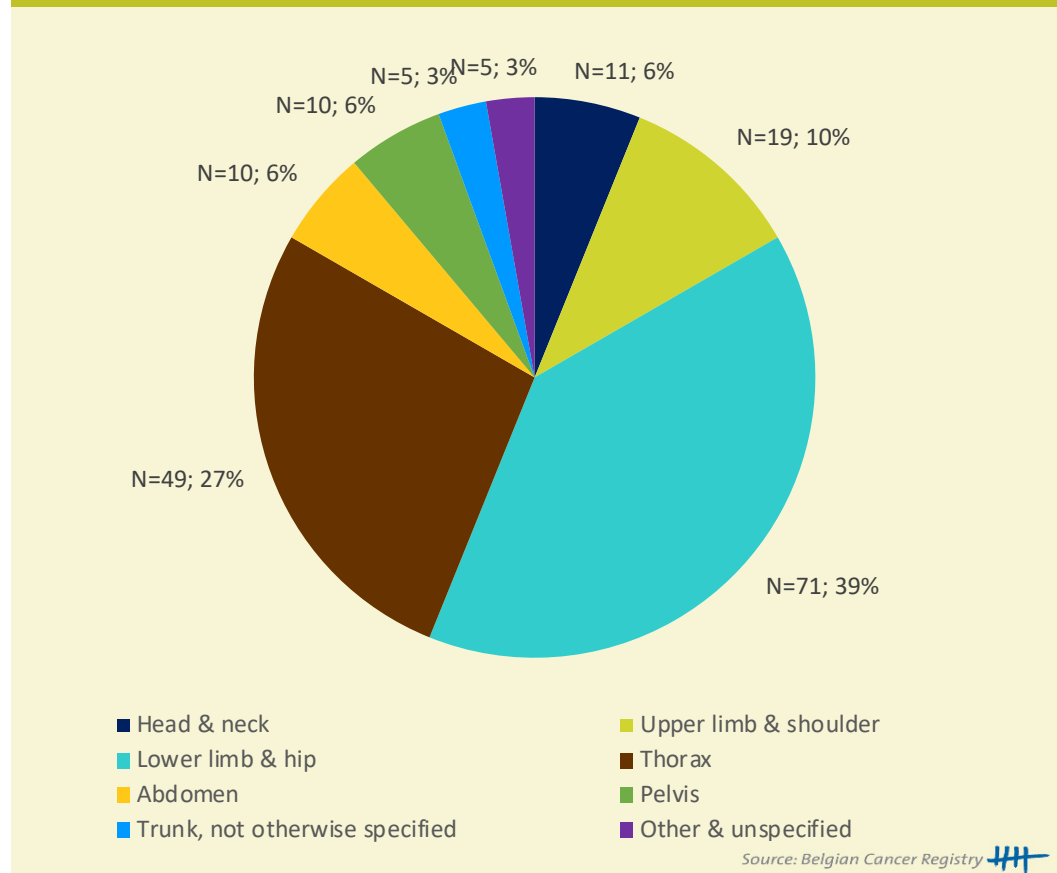
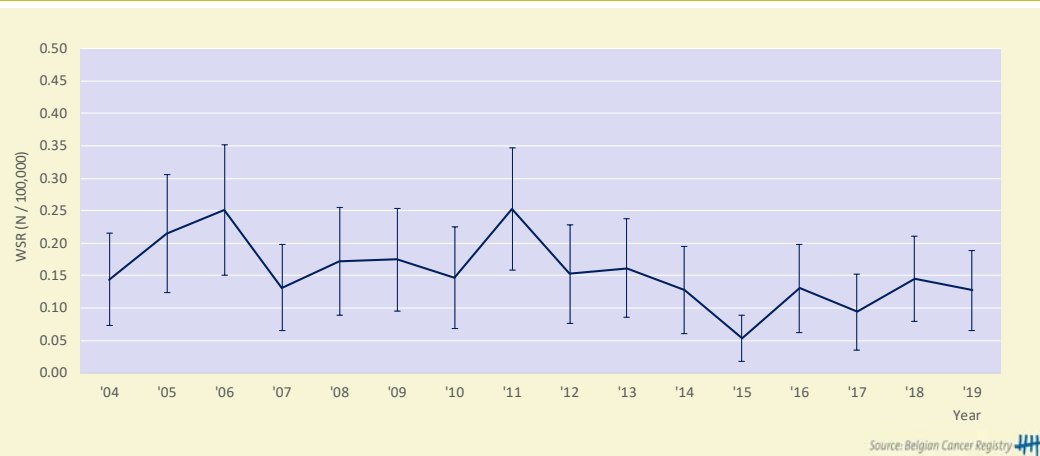


Figure 2 Synovial sarcoma: Incidence distribution by primary tumour location, Belgium 2010-2019



Incidence trends

Figure 3 Synovial sarcoma: Age-standardised incidence rates* (WSR), males and females, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals

Table 2 Synovial sarcoma: Incidence trends by sex in Belgium, 2004-2019

Age category	Males			Females		
	AAPC (%)	95%CI	Period	AAPC (%)	95%CI	Period
All ages	-2.7	[-7.4; 2.4]	2004-2019	-5.5	[-9.5; -1.4]	2004-2019

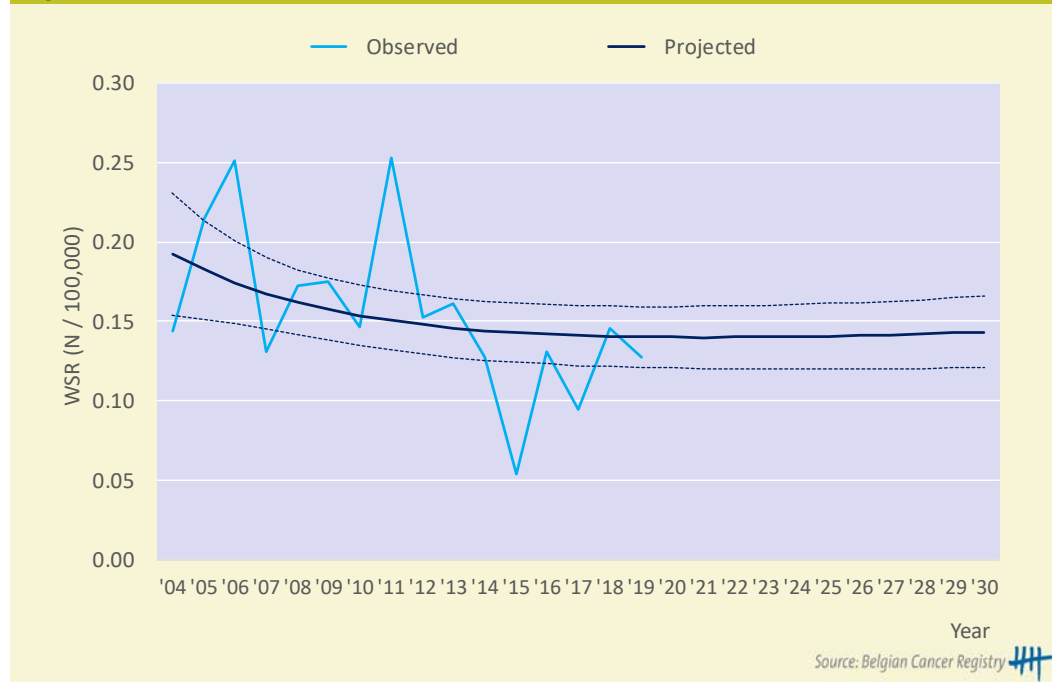
Source: Belgian Cancer Registry

AAPC: average annual percentage change

Period: when a joinpoint occurred, APC's are calculated for the period before and after the joinpoint. This column represents the corresponding time interval. AAPC's are always calculated over the entire study-period.

Incidence projections

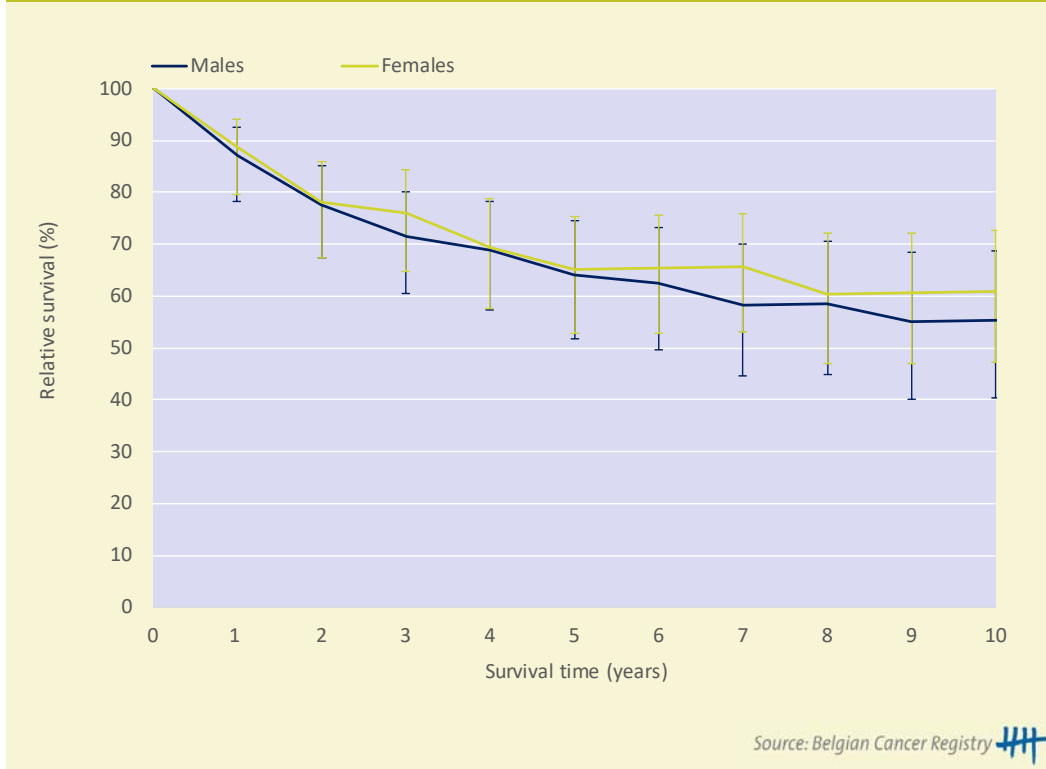
Figure 4 Synovial sarcoma: Observed and projected* incidence (WSR), Belgium 2004-2030



WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

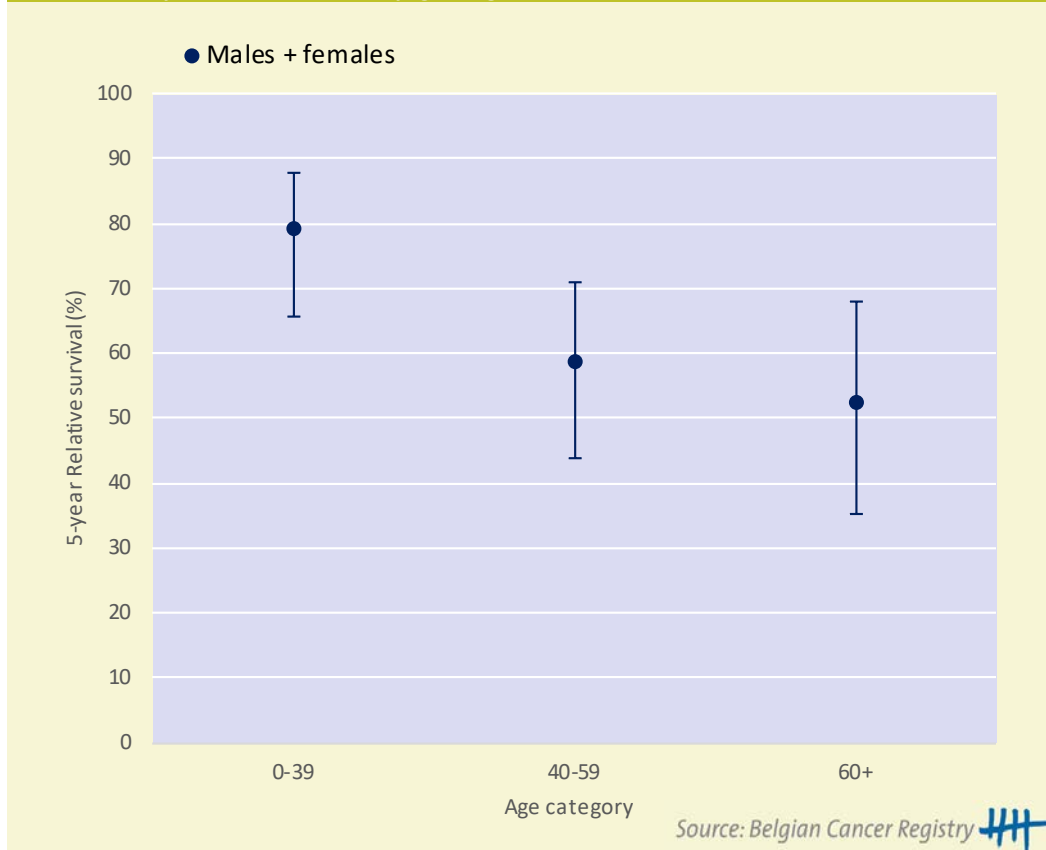
Survival

Figure 5 Synovial sarcoma: Relative survival* by sex, Belgium 2010-2019



* The relative survival values are represented with 95% Confidence Intervals

Figure 6 Synovial sarcoma: 5-year relative survival* by age, Belgium 2010-2019



* The relative survival values are represented with 95% Confidence Intervals

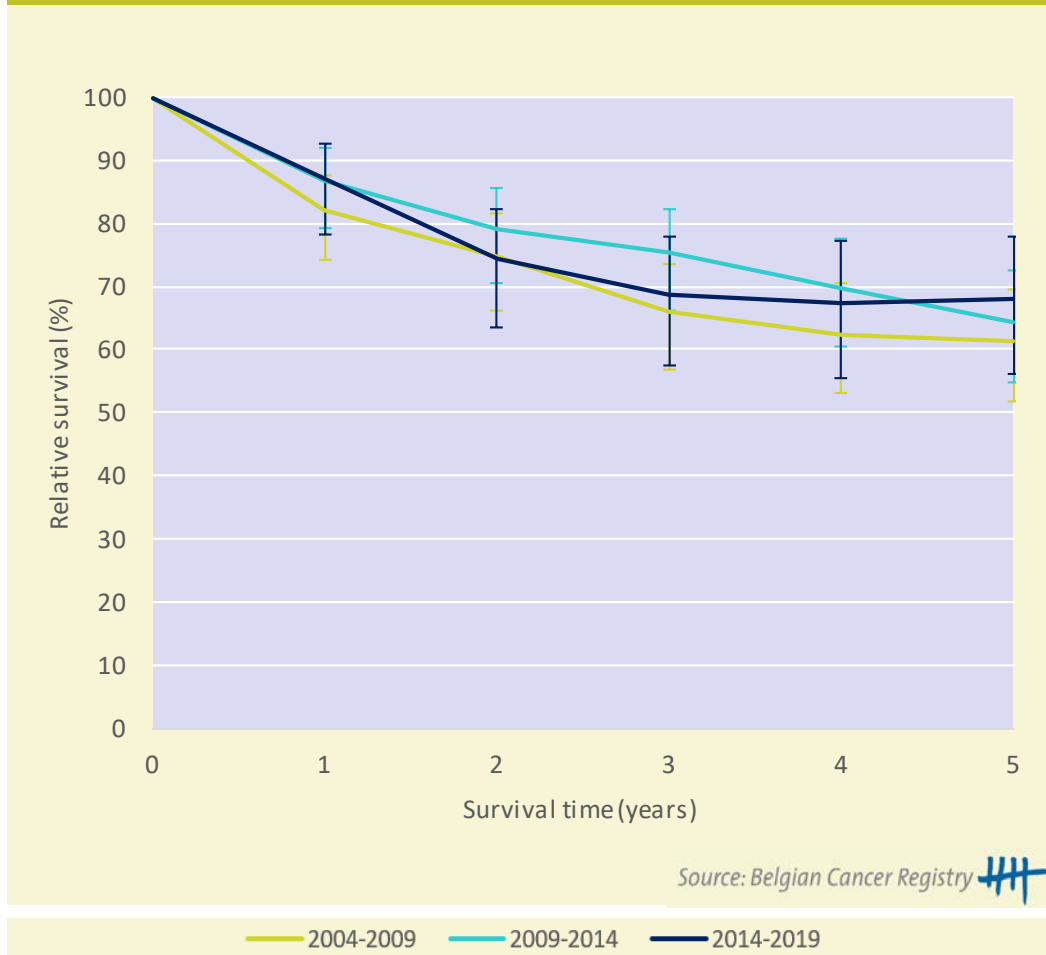
Table 3 Synovial sarcoma: Conditional 5-year relative survival* in Belgium, 2010-2019

X years since diagnosis	N at risk	%
1 year	154	72.6
2 year	126	79.5
3 year	100	80.7

* Unadjusted 5-yr relative survival probability conditional on surviving the first X years since diagnosis, %
 * Interpretation in lay-man's terms: Given that a patient has already survived X years, what is the relative survival probability 5 years later.

Survival trends

Figure 7 Synovial sarcoma: Relative survival* by cohort, Belgium 2004-2019



* The relative survival values are represented with 95% Confidence Intervals

3.2.7.2 MYOEPITHELIOMA

Notes:

- Most myoepithelial tumours are currently classified as benign tumours (WHO classification of 2020) and the registration might not be exhaustive.
- Myoepithelioma shares morphological, immunophenotypic, and genetic features with their counterparts in salivary glands, which are not considered 'soft tissue' tumours, but are reported together in this chapter⁽¹⁾.

KEYNOTES

Incidence (table 1-2; figure 1-4)

- The incidence of myoepithelioma increases with age in a similar way in males and females, showing an incidence peak in patients aged older than 60.
- About half of myoepitheliomas arise in the head & neck region (in which the majority (2/3) in the parotid gland), followed by the breast (16%).

Survival (table 3; figure 5-7)

- Given that a patient with myoepithelioma survives the first three years after diagnosis, the conditional relative survival 5 years later is 97%.
- The 5-year relative survival rate does not show a consistent difference according to age group (under 65 years of age versus above), but seems to improve over time from 64% in 2004-2011 to 73% in 2012-2019 (attention: subject to small absolute number of cases and large confidence intervals).
- The 10-year relative survival rate seems better in females (70%) than in males (57%).

Table 1 Myoepithelioma: Overview of incidence, prevalence and survival by sex in Belgium

Males				
Incidence	N	CR	WSR	
Incidence, 2010-2019	37	0.07	0.05	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	11	0.19	0.13	
10-year prevalence, 31.12.2019	20	0.35	0.26	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2004-2019	55	59.4	[41.9;75.0]	
10-year relative survival, 2004-2019	55	57.3	[36.4;77.5]	
Females				
Incidence	N	CR	WSR	
Incidence, 2010-2019	45	0.08	0.05	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	11	0.19	0.10	
10-year prevalence, 31.12.2019	28	0.48	0.24	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2004-2019	71	74.1	[59.6;85.7]	
10-year relative survival, 2004-2019	71	69.9	[50.8;87.2]	
Median age at diagnosis, 2010-2019 (y)	62 [Q1: 51; Q3: 78]			
M/F-ratio	1.0			

Source: Belgian Cancer Registry 

N: number of new diagnoses

CR: crude (all ages) rate (N/100,000 person years)

WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

M/F-ratio: Male/Female ratio based on the age-standardised rates (2010-2019)

Incidence

Figure 1 Myoepithelioma: Age-specific incidence rates (N/100,000) by sex, Belgium 2004-2019

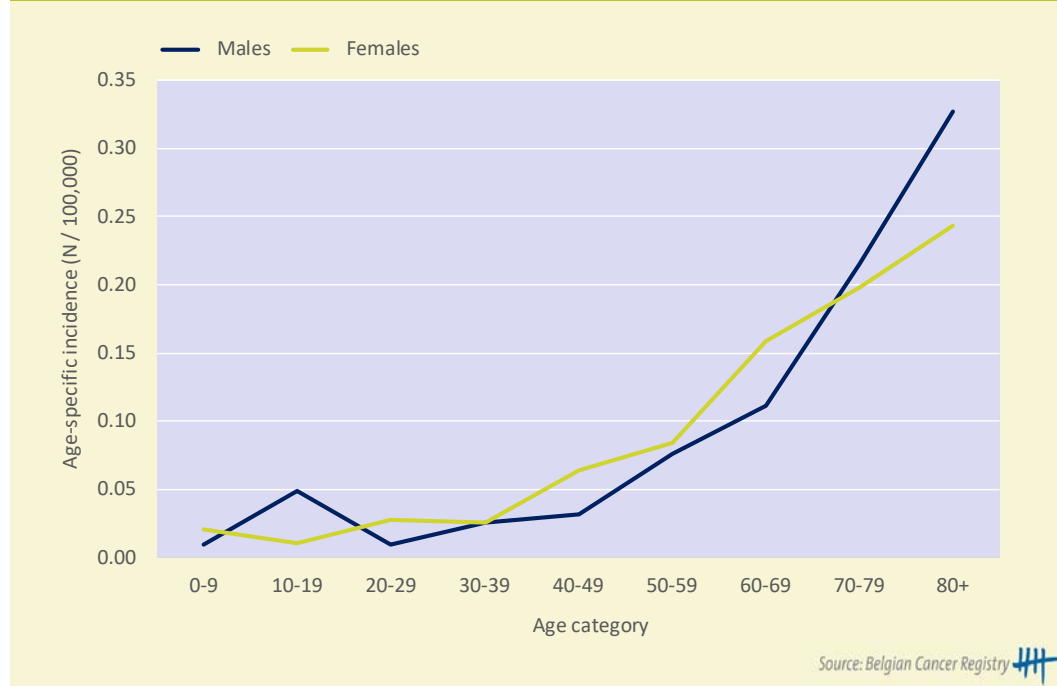
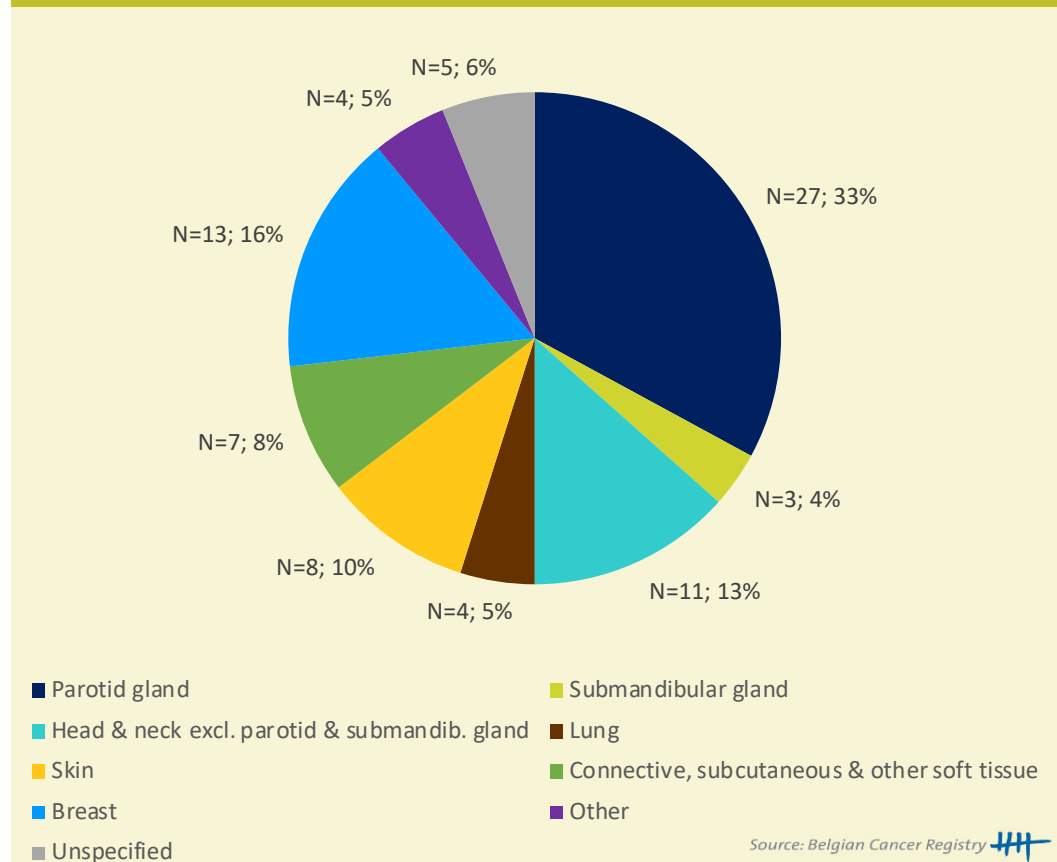
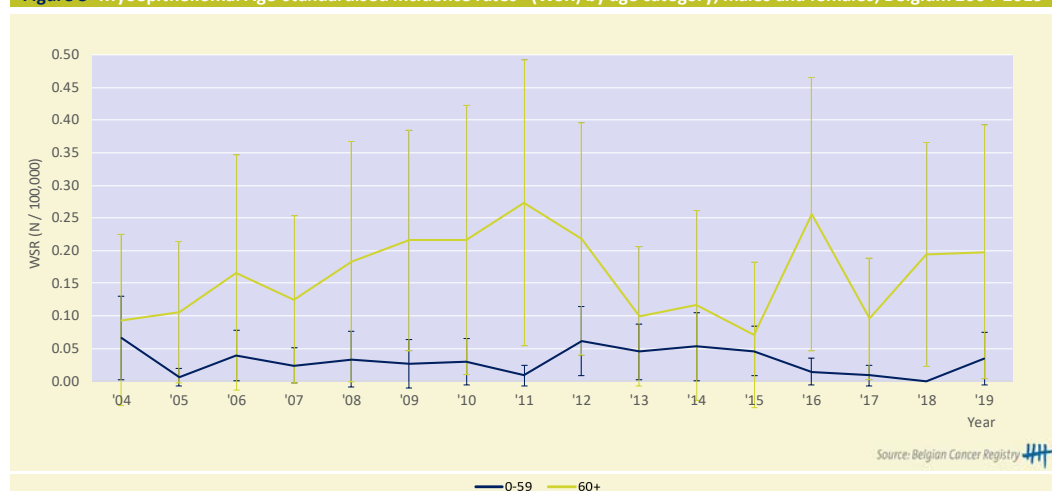


Figure 2 Myoepithelioma: Incidence distribution by primary tumour location, Belgium 2010-2019



Incidence trends

Figure 3 Myoepithelioma: Age-standardised incidence rates* (WSR) by age category, males and females, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals

Table 2 Myoepithelioma: Incidence trends by sex in Belgium, 2004-2019

Age category	Males			Females		
	AAPC (%)	95%CI	Period	AAPC (%)	95%CI	Period
All ages	-0.3	[-7.0; 6.8]	2004-2019	-4.1	[-10.4; 2.7]	2004-2019

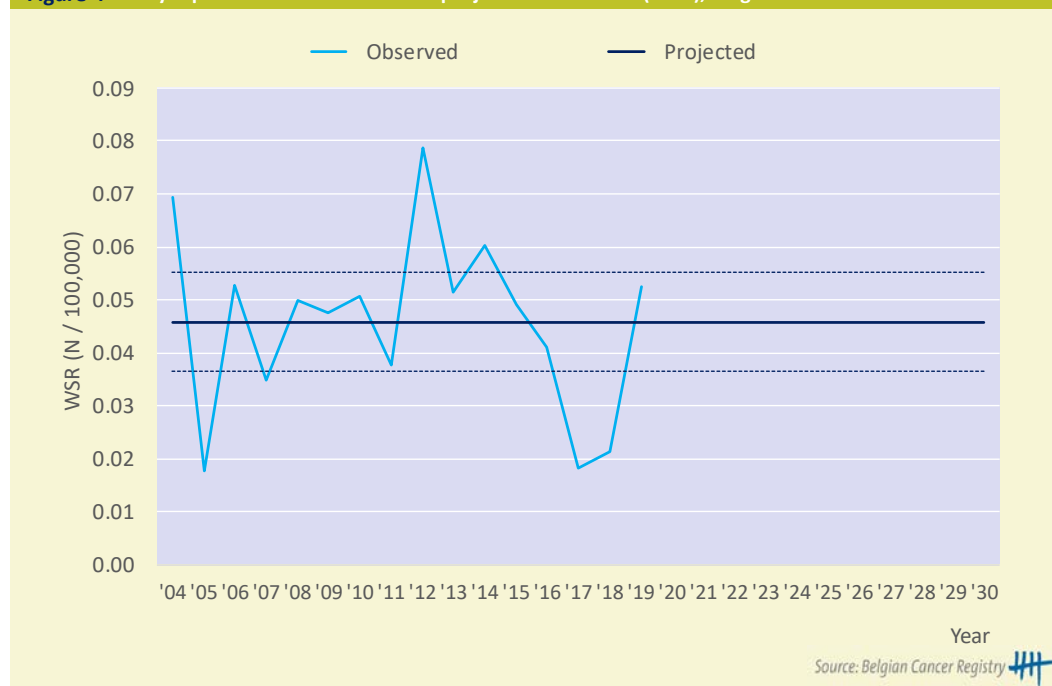
Source: Belgian Cancer Registry

AAPC: average annual percentage change

Period: when a joinpoint occurred, APC's are calculated for the period before and after the joinpoint. This column represents the corresponding time interval. AAPC's are always calculated over the entire study-period.

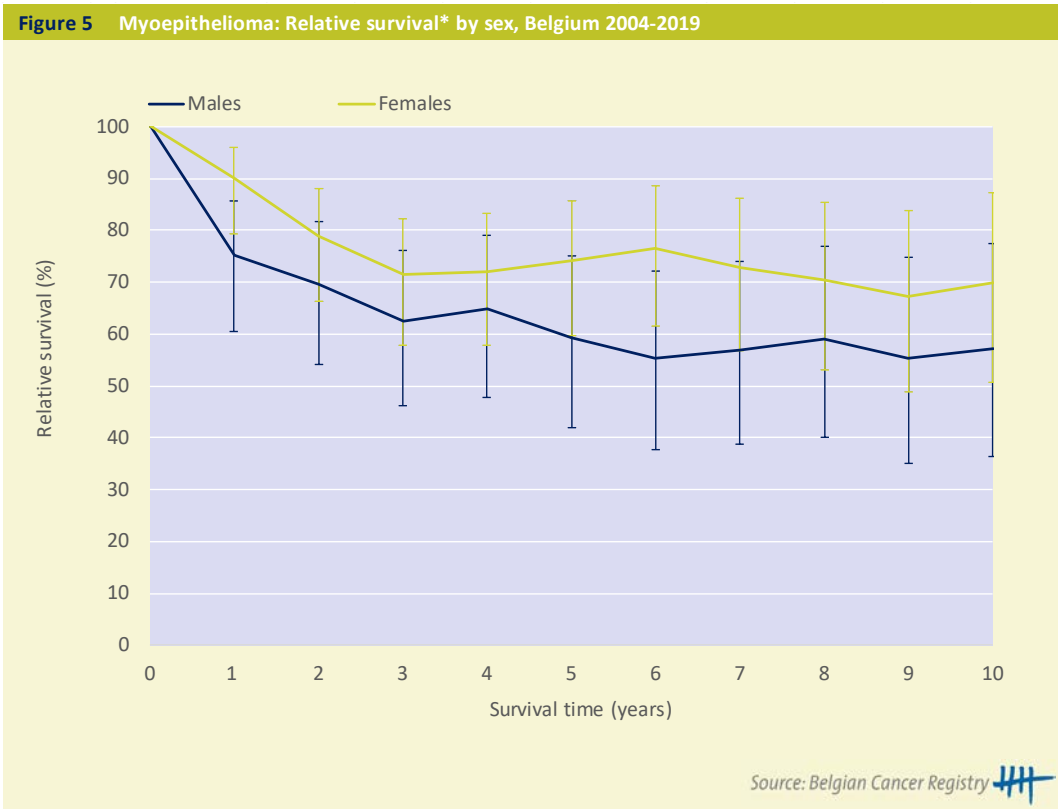
Incidence projections

Figure 4 Myoepithelioma: Observed and projected* incidence (WSR), Belgium 2004-2030

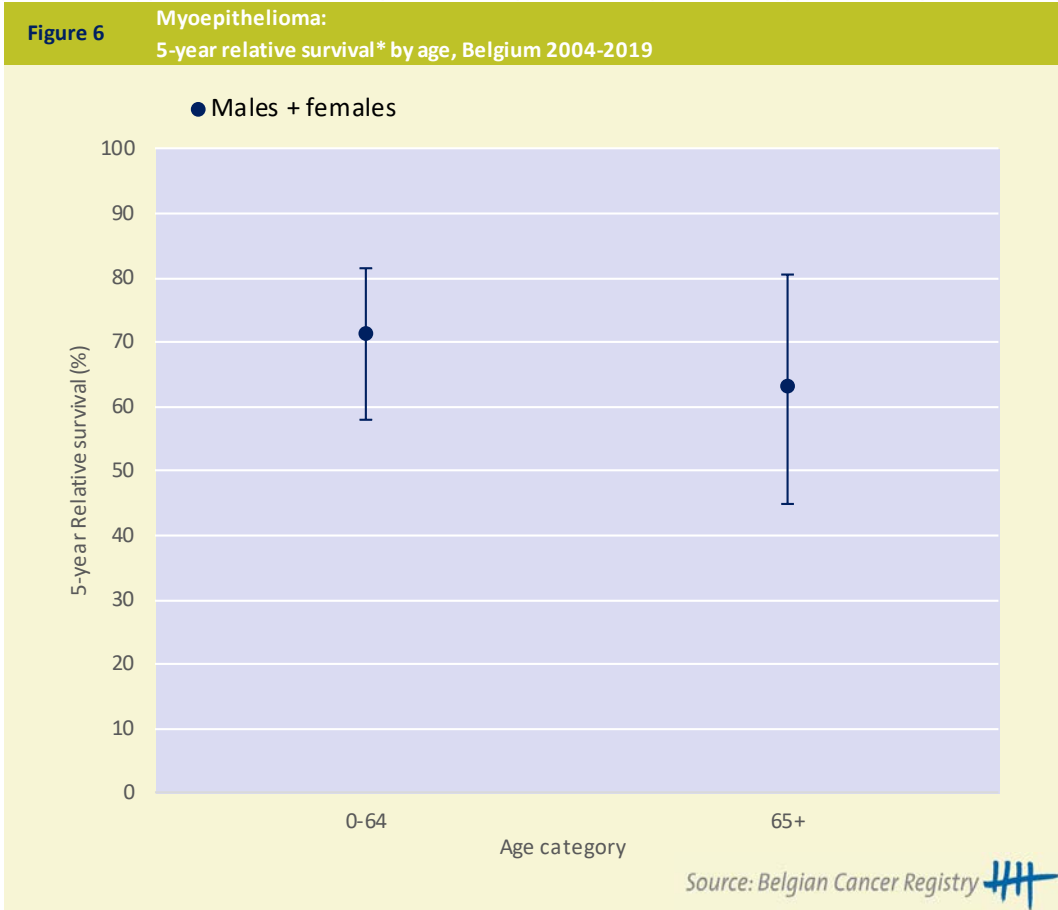


WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

Survival



* The relative survival values are represented with 95% Confidence Intervals



* The relative survival values are represented with 95% Confidence Intervals

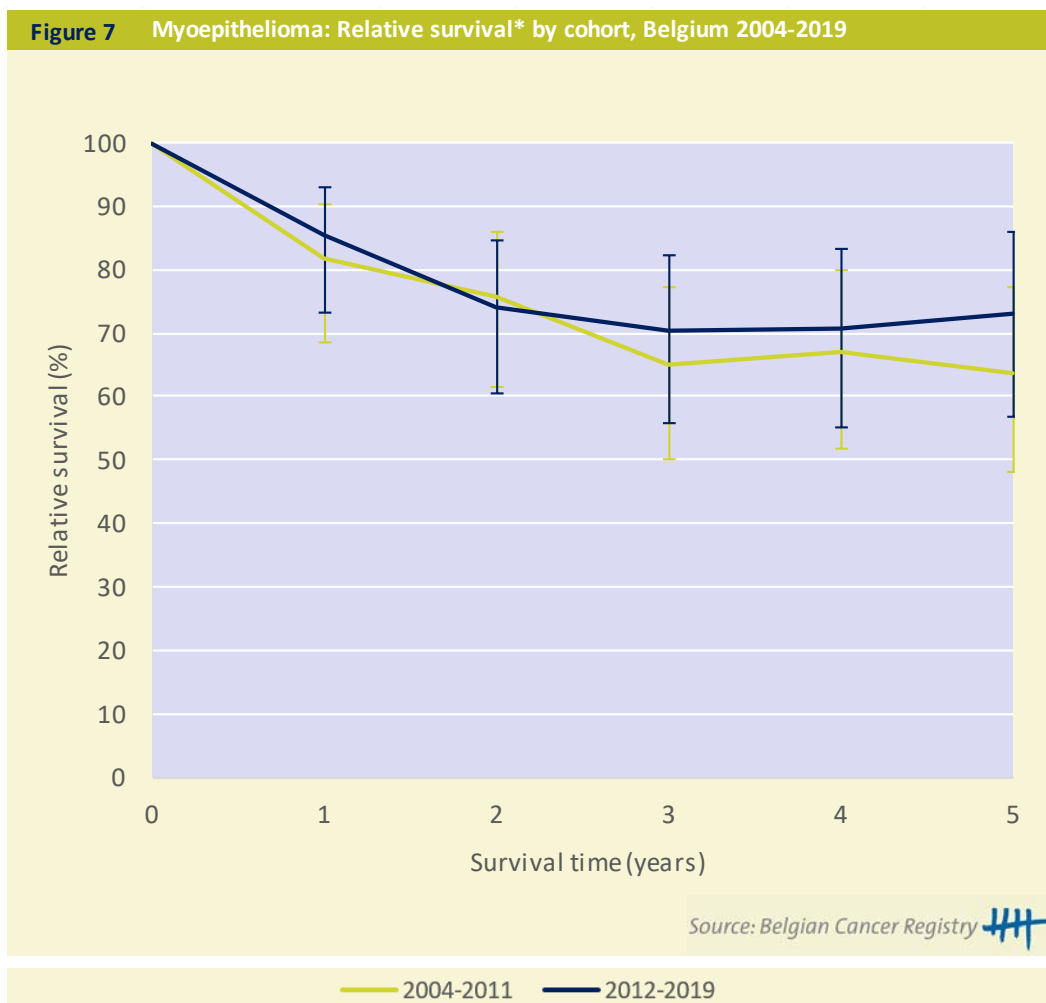
Table 3 Myoepithelioma: Conditional 5-year relative survival* in Belgium, 2004-2019

X years since diagnosis	N at risk	%
1 year	101	81.0
2 year	85	88.1
3 year	67	96.8

* Unadjusted 5-yr relative survival probability conditional on surviving the first X years since diagnosis, %
 * Interpretation in lay-man's terms: Given that a patient has already survived X years, what is the relative survival probability 5 years later.

Survival trends

Figure 7 Myoepithelioma: Relative survival* by cohort, Belgium 2004-2019



* The relative survival values are represented with 95% Confidence Intervals

3.2.7.3 RHABDOID TUMOURS

KEYNOTES

Incidence (table 1; figure 1-4)

- Rhabdoid tumours are very rare. They present more frequently in males than in females (M/F ratio of 1.4) and are almost exclusively diagnosed in children under the age of 10 years.
- The predominant localization of rhabdoid tumours is the central nervous system (71%) but they can occur anywhere throughout the soft tissues and visceral organs.
- The apparent increasing incidence over time of rhabdoid tumours is mostly due to improved diagnosis, better classification and registration.

Survival (table 2; figure 5)

- Rhabdoid tumours are very aggressive with a 10-year relative survival rate around 30% (plateau reached after 6 years), hereby they represent the second worst prognosis of all sarcomas, after angiosarcoma (See chapter 3.2, figure 8).

Table 1 Rhabdoid tumours: Overview of incidence, prevalence and survival by sex in Belgium, 2010-2019				
Males				
Incidence	N	CR	WSR	
Incidence, 2010-2019	23	0.04	0.08	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	4	0.07	0.12	
10-year prevalence, 31.12.2019	8	0.14	0.23	
Relative survival	Not enough patients for representative survival analysis			
Females				
Incidence	N	CR	WSR	
Incidence, 2010-2019	15	0.03	0.06	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	5	0.09	0.18	
10-year prevalence, 31.12.2019	7	0.12	0.24	
Relative survival	Not enough patients for representative survival analysis			
Median age at diagnosis, 2010-2019 (y)	1 [Q1: 0; Q3: 3]			
M/F-ratio	1.4			

Source: Belgian Cancer Registry 

N: number of new diagnoses

CR: crude (all ages) rate (N/100,000 person years)

WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

M/F-ratio: Male/Female ratio based on the age-standardised rates (2015-2019)

Incidence

Figure 1 Rhabdoid tumours: Age-specific incidence rates (N/100,000), Belgium 2004-2019

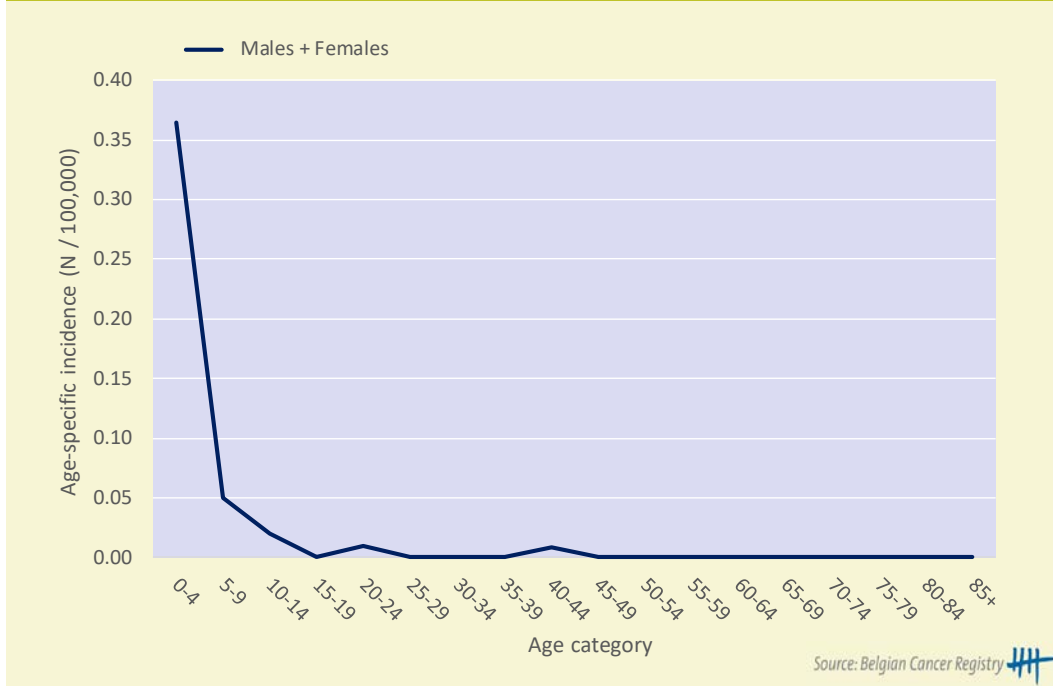
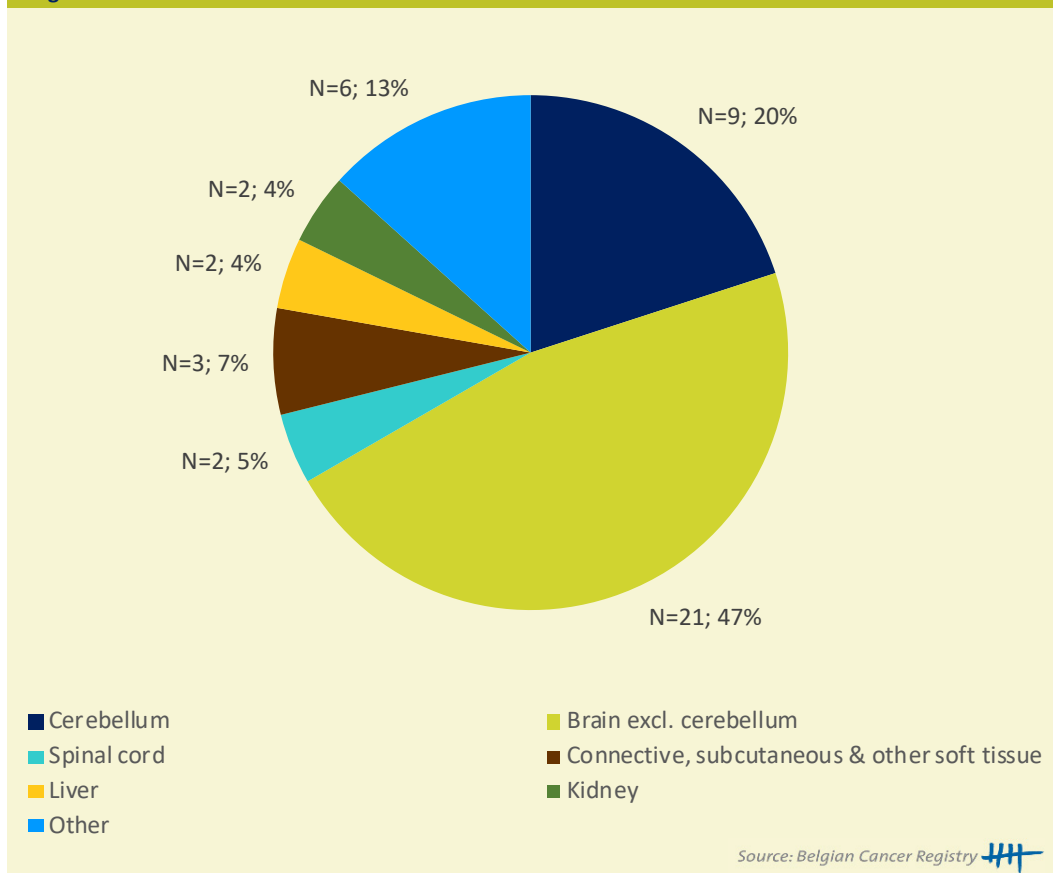
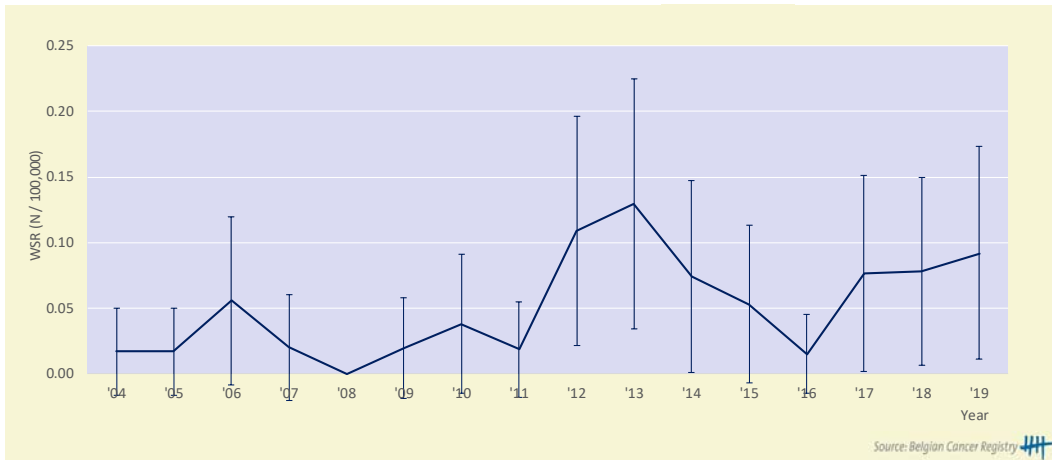


Figure 2 Rhabdoid tumours: Incidence distribution by primary tumour location, Belgium 2004-2019



Incidence trends

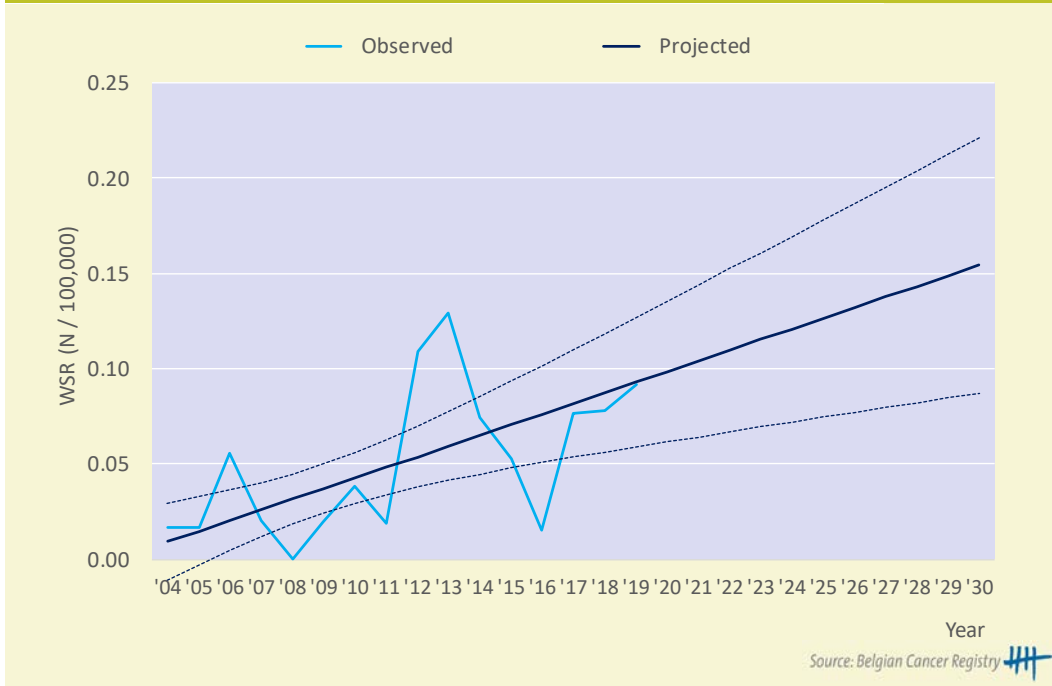
Figure 3 Rhabdoid tumours: Age-standardised incidence rates* (WSR), males and females, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals

Incidence projections

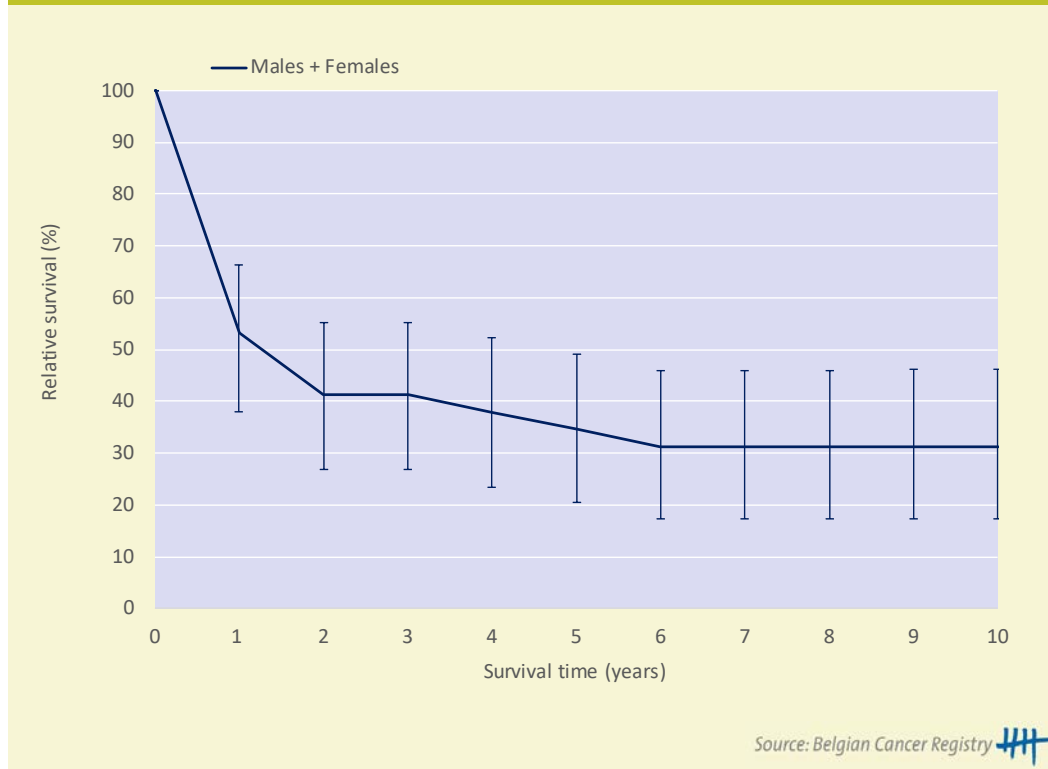
Figure 4 Rhabdoid tumours: Observed and projected* incidence (WSR), Belgium 2004-2030



WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

Survival

Figure 5 Rhabdoid tumours: Relative survival*, Belgium 2004-2019



* The relative survival values are represented with 95% Confidence Intervals.

Note: survival analysis is based on 45 patients only.

X years since diagnosis	N at risk	%
1 year	24	58.7
2 year	17	75.5
3 year	14	75.5

* Unadjusted 5-yr relative survival probability conditional on surviving the first X years since diagnosis, %

* Interpretation in lay-man's terms: Given that a patient has already survived X years, what is the relative survival probability 5 years later.

3.2.8 GASTROINTESTINAL STROMAL TUMOUR (GIST)

KEYNOTES

Incidence (table 1-2; figure 1-6)

- GIST is the most frequent type of soft tissue tumour (see chapter 3.2, figure 5), the incidence increases with age (slightly more pronounced in males than in females) with a peak in the older population (50+ years).
- In the digestive system GIST is the most often diagnosed soft tissue tumour subtype (see chapter 3.2, figure 7). The most frequent primary location of GIST is the stomach (68%) followed by the small intestine (23%).
- The increasing incidence rates observed between 2004 and 2019 (AAPC of 4.4% for males and 6.1% for females) can be mostly explained by modifications of the classification and more accurate registration over time. Data for GIST also include so-called “micro-GIST” (tumours smaller than 1 cm in diameter) sometimes discovered as incidental findings. There is an increased awareness among pathologists and increased use of biomarkers (KIT, DOG1, mutational analysis) to identify GIST.
- As for stage at presentation (TNM classification), in 2017-2019, the majority (70%) of GIST with a known size were diagnosed with a size ≤ 5 cm. Positive regional lymph nodes are only seen in 2%, and distant metastasis in 5% of the diagnoses.

Survival (table 3; figure 7-9)

- Patients with GIST have the second-best prognosis (after dermatofibrosarcoma protuberans, see chapter 3.2 Figure 8).
- The 10-year relative survival rate tends to be slightly better in females (90%) than in males (81%).
- The 5-year relative survival rate does not seem to change according to age (always above 80%) nor to improve over time (2004-2019).

Table 1 GIST: Overview of incidence, prevalence and survival by sex in Belgium, 2010-2019

Males				
	N	CR	WSR	
Incidence				
Incidence, 2010-2019	1,464	2.7	1.5	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	695	12.3	6.3	
10-year prevalence, 31.12.2019	1,095	19.3	9.7	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	1,452	89.7	[86.7;92.5]	
10-year relative survival, 2010-2019	1,452	81.4	[74.4;88.1]	
Females				
	N	CR	WSR	
Incidence				
Incidence, 2010-2019	1,362	2.4	1.2	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	662	11.4	5.5	
10-year prevalence, 31.12.2019	1,112	19.1	8.8	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	1,358	93.0	[90.3;95.5]	
10-year relative survival, 2010-2019	1,358	90.1	[83.3;96.4]	
Median age at diagnosis, 2010-2019 (y)	67 [Q1: 58; Q3: 76]			
M/F-ratio	1.2			

Source: Belgian Cancer Registry 

N: number of new diagnoses

CR: crude (all ages) rate (N/100,000 person years)

WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

M/F-ratio: Male/Female ratio based on the age-standardised rates (2010-2019)

Incidence

Figure 1 GIST: Age-specific incidence rates (N/100,000) by sex, Belgium 2004-2019

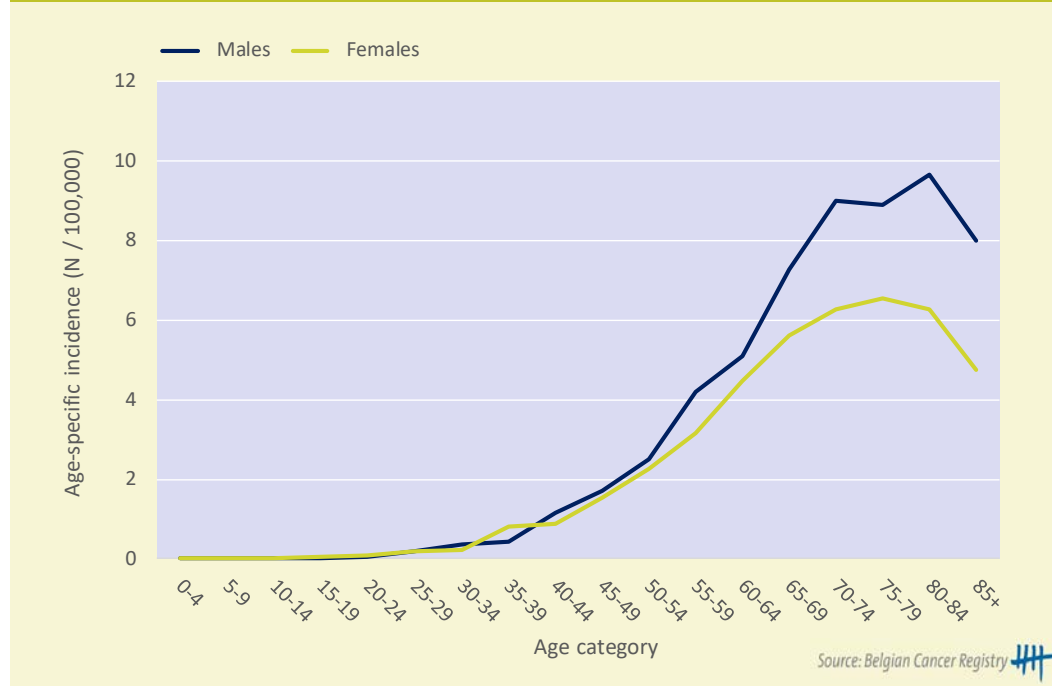
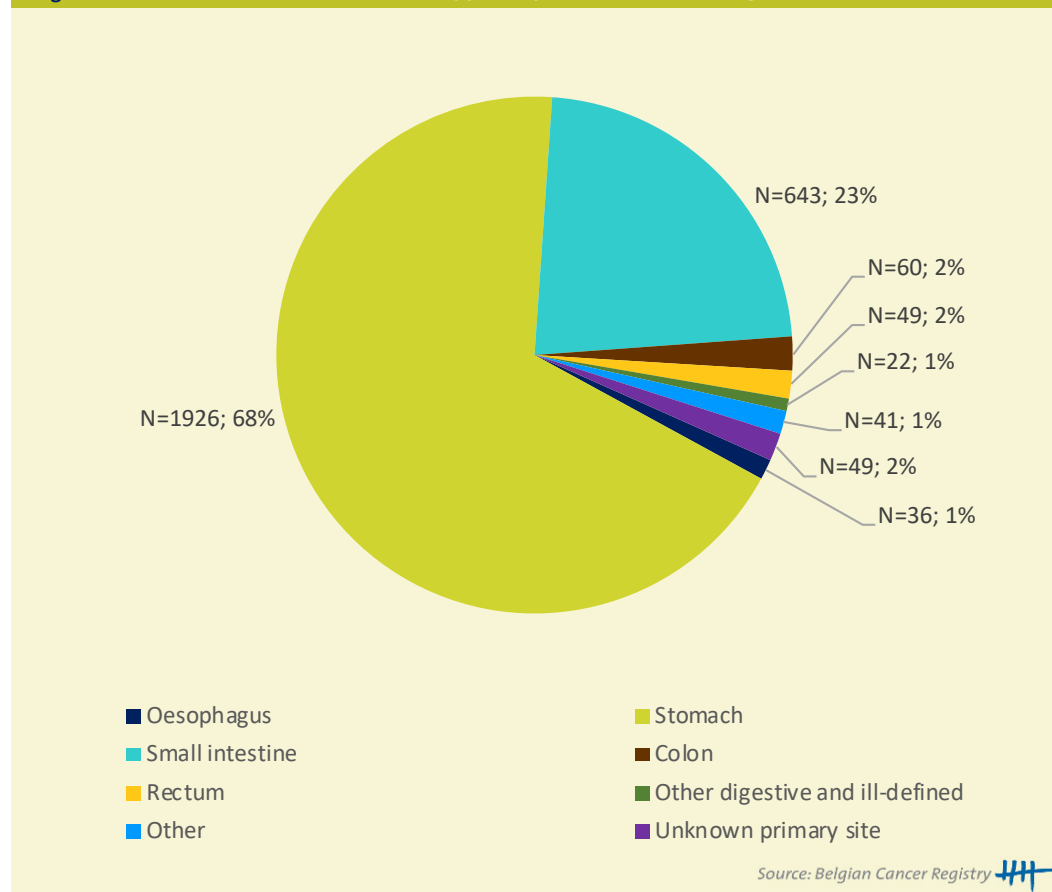
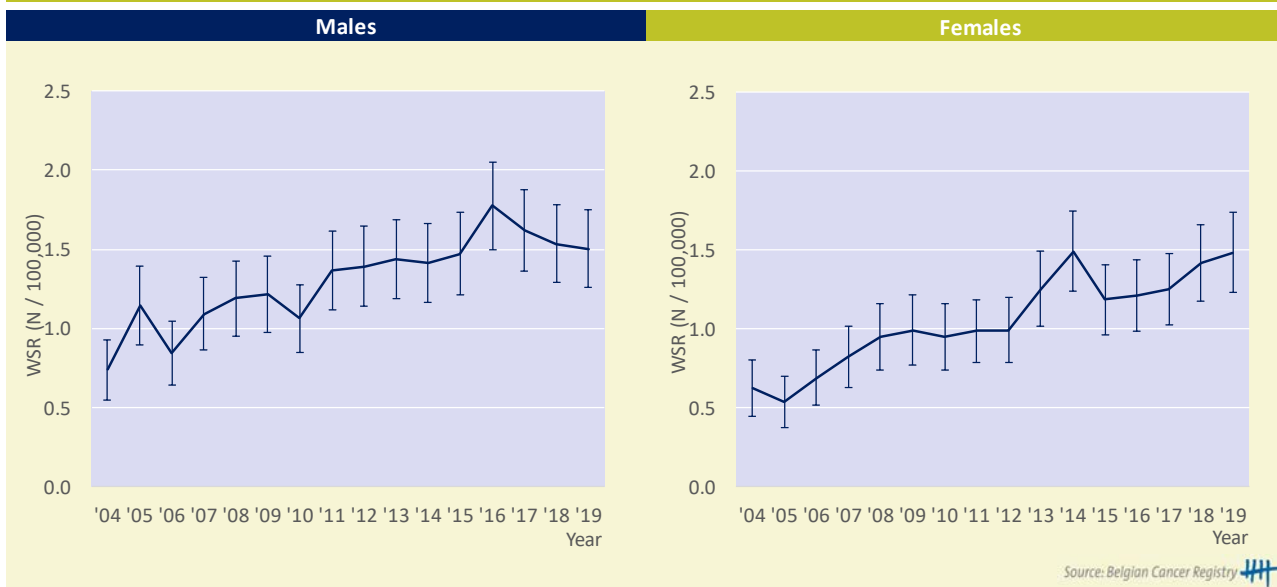


Figure 2 GIST: Incidence distribution by primary tumour location, Belgium 2010-2019



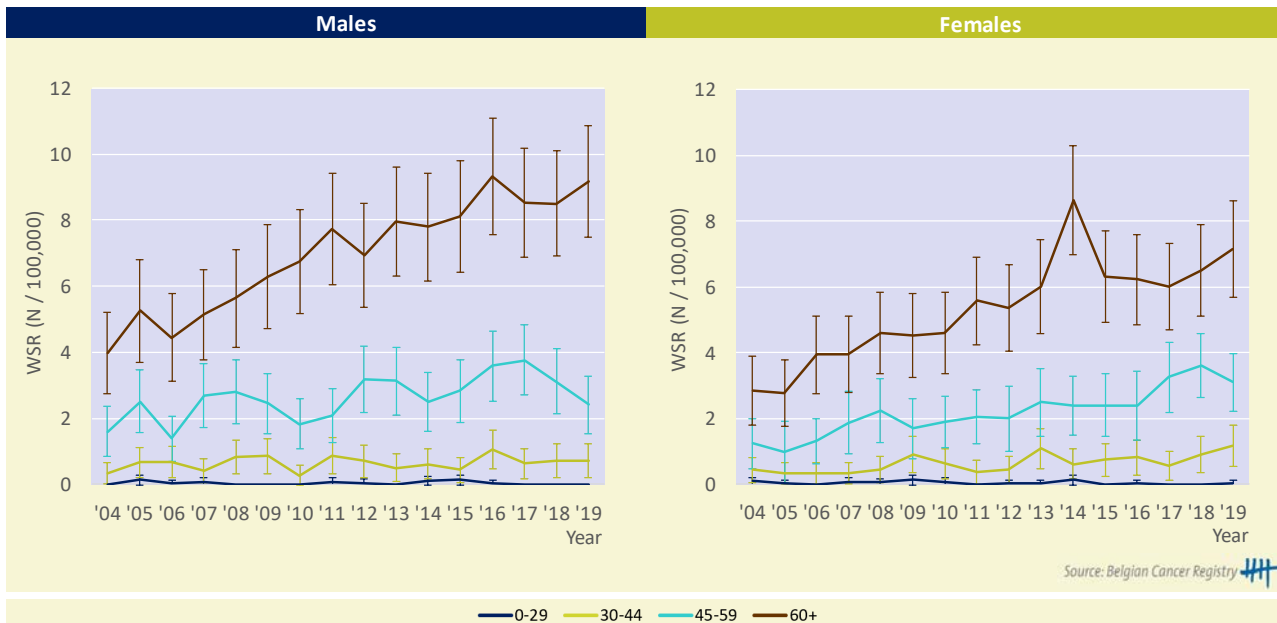
Incidence trends

Figure 3 GIST: Age-standardised incidence rates* (WSR) by sex, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals.

Figure 4 GIST: Age-standardised incidence rates* (WSR) by sex and age category, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals.

Table 2 GIST: Incidence trends by sex and age category in Belgium, 2004-2019

Age category	Males			Females		
	AAPC (%)	95%CI	Period	AAPC (%)	95%CI	Period
All ages	4.4	[2.9; 5.8]	2004-2019	6.1	[4.8; 7.5]	2004-2019
				9.1	[5.8; 12.5]	2004-2011
				3.6	[0.9; 6.4]	2011-2019
0 - 29 y	-	-	-	-	-	-
30 - 44 y	2.4	[-1.9; 6.9]	2004-2019	6.8	[3.0; 10.7]	2004-2019
45 - 59 y	3.7	[1.0; 6.4]	2004-2019	7.0	[5.1; 8.8]	2004-2019
60+ y	5.3	[4.4; 6.1]	2004-2019	5.4	[4.0; 6.7]	2004-2019
	7.7	[5.9; 9.6]	2004-2012	9.0	[6.9; 11.2]	2004-2014
	2.5	[0.5; 4.6]	2012-2019	-1.6	[-5.8; 2.8]	2014-2019

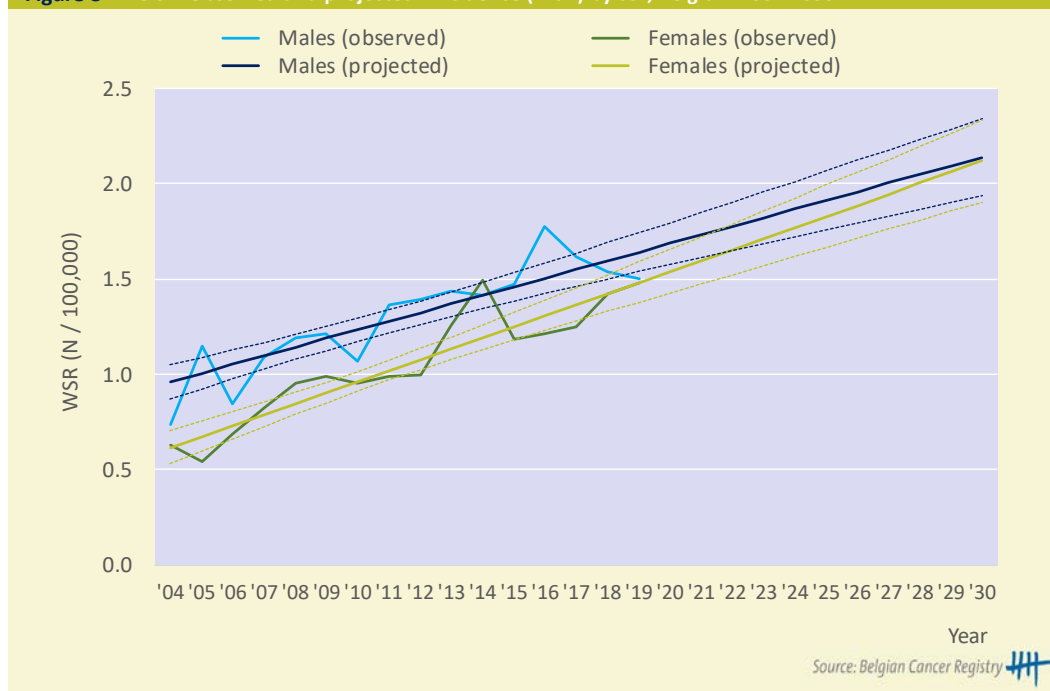
Source: Belgian Cancer Registry 

AAPC: average annual percentage change

Period: when a joinpoint occurred, APC's are calculated for the period before and after the joinpoint. This column represents the corresponding time interval. AAPC's are always calculated over the entire study-period.

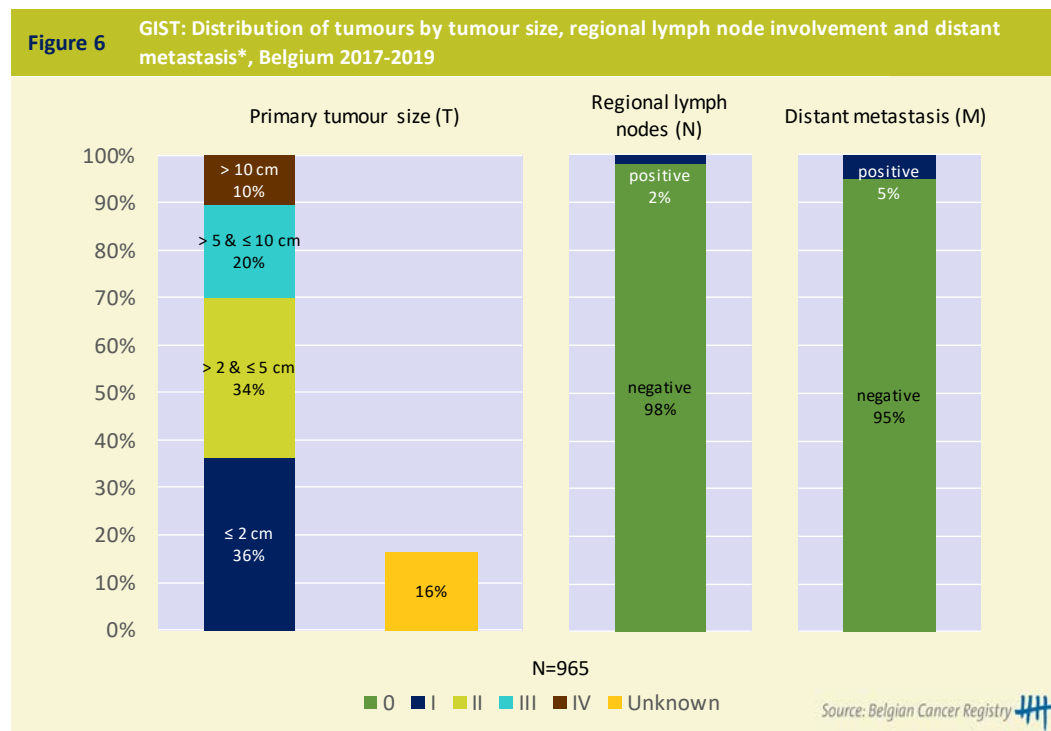
Incidence projections

Figure 5 GIST: Observed and projected* incidence (WSR) by sex, Belgium 2004-2030



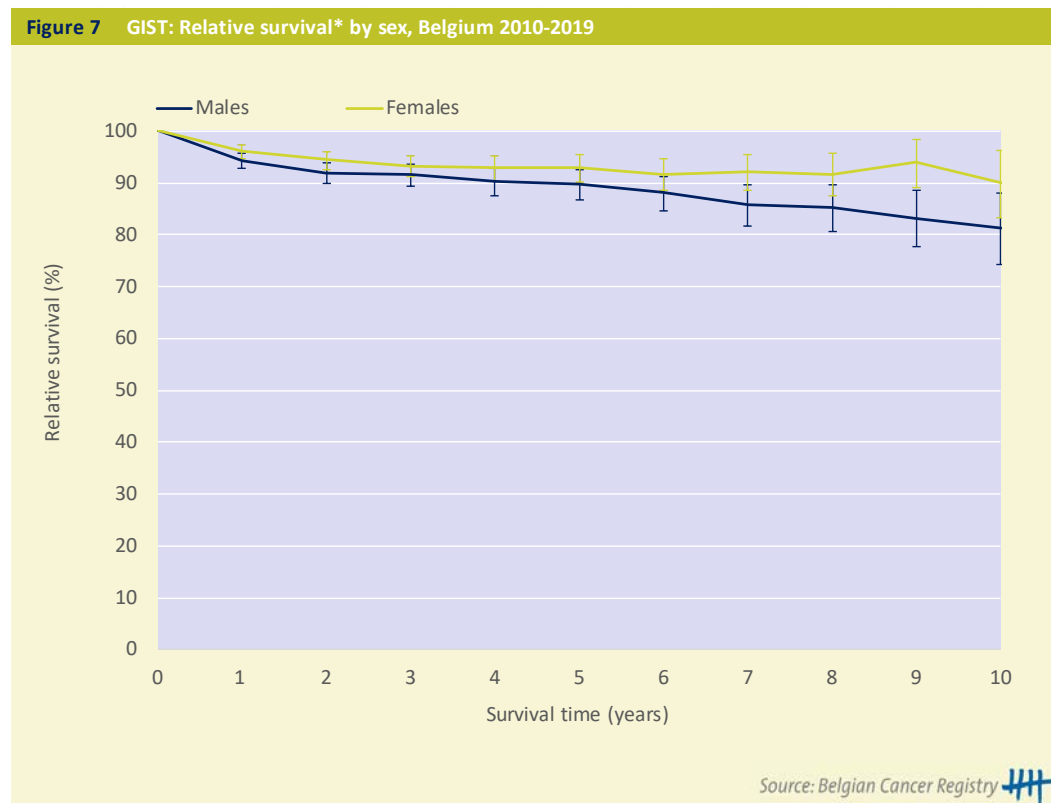
WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

TNM Classification



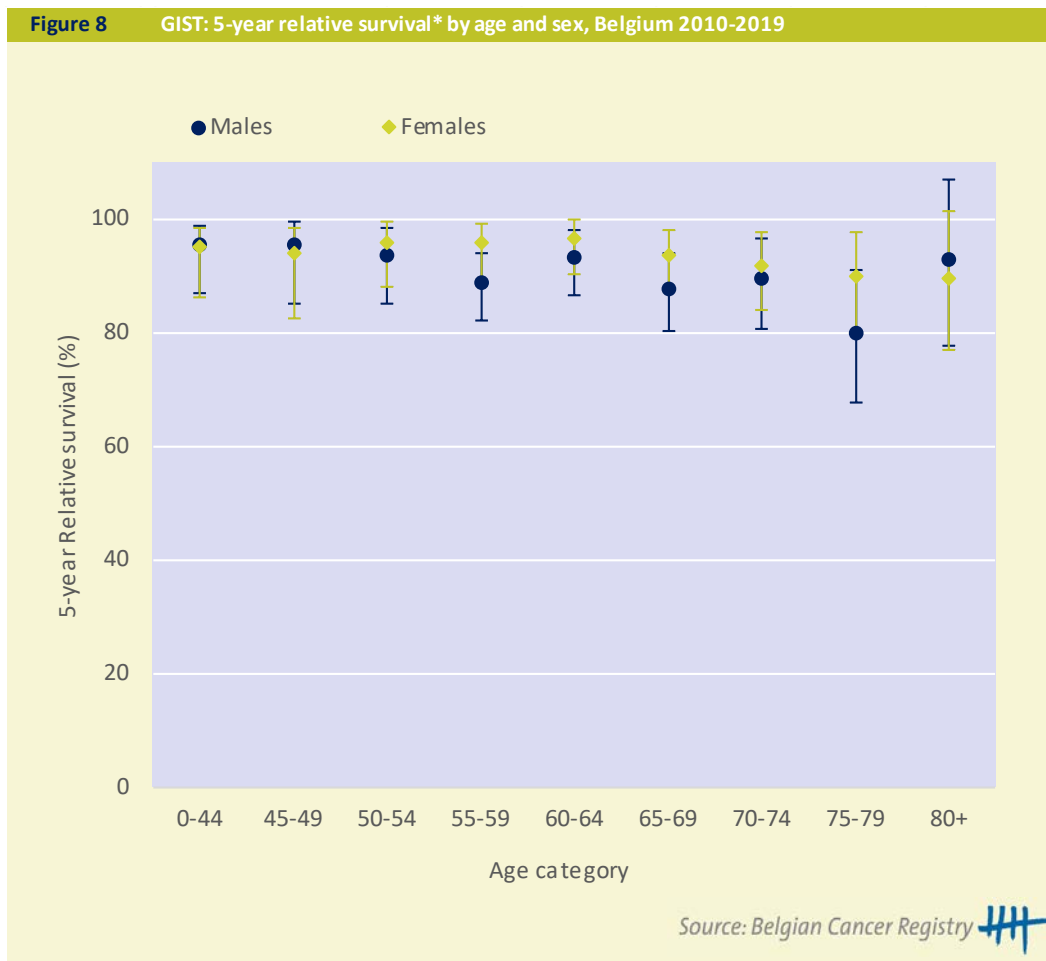
* TNM Classification of Malignant Tumours, Eighth edition, UICC, 2017. Based on the pathological T, N and M categories. If no pathological information was available, the clinical TNM was used.

Survival



* The relative survival values are represented with 95% Confidence Intervals

Figure 8 GIST: 5-year relative survival* by age and sex, Belgium 2010-2019



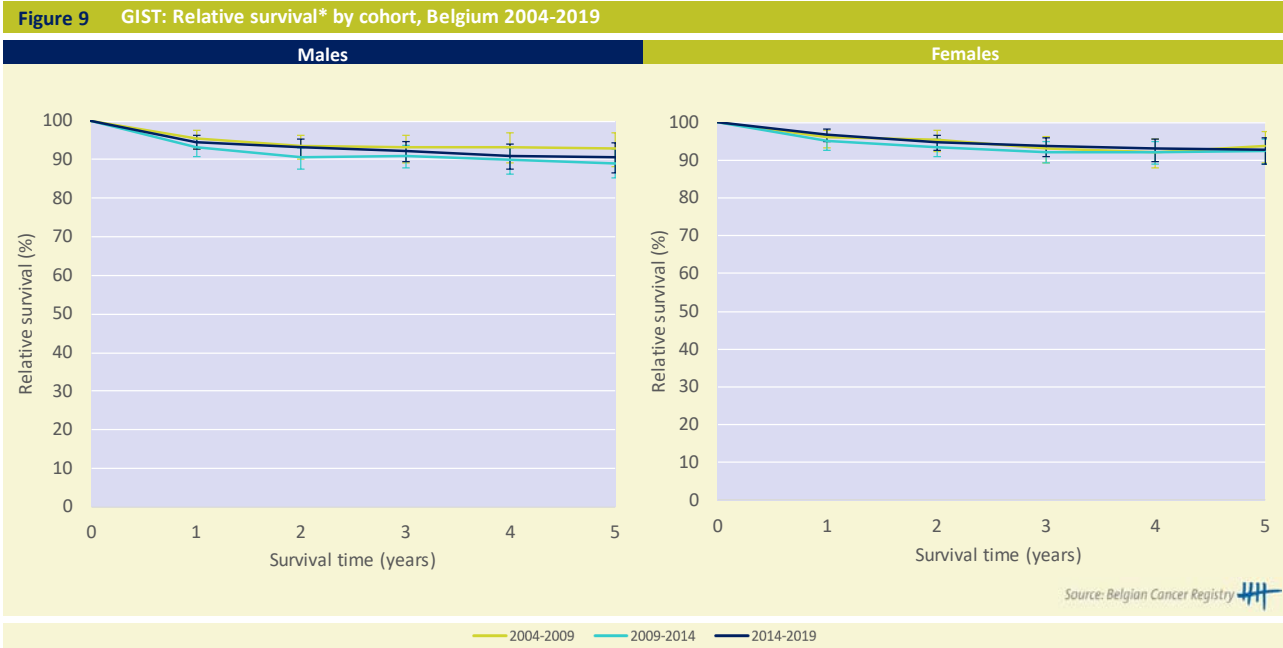
* The relative survival values are represented with 95% Confidence Intervals

X years since diagnosis	N at risk	%
1 year	2,599	94.4
2 year	2,255	95.5
3 year	1,873	95.7

* Unadjusted 5-yr relative survival probability conditional on surviving the first X years since diagnosis, %

* Interpretation in lay-man's terms: Given that a patient has already survived X years, what is the relative survival probability 5 years later.

Survival trends



* The relative survival values are represented with 95% Confidence Intervals

3.2.9 ENDOMETRIAL STROMAL SARCOMA

KEYNOTES

Incidence (table 1-2; figure 1-5)

- Endometrial stromal sarcoma is mostly diagnosed in female patients aged older than 40 years. It is never diagnosed under 20 years of age.
- The large majority (82%) of endometrial stromal sarcomas diagnosed in 2017-2019 are diagnosed in an early stage (stage I and II).

Survival (table 3; figure 6-8)

- The relative survival of patients with endometrial stromal sarcoma seems to reach a plateau after surviving 6 years, with a 10-year relative survival of 68%.
- Given that a patient survives the first five years, the relative survival probability 5 years later is 93%.
- The 5-year relative survival rate of endometrial stromal sarcoma:
 - strongly decreases with the age of patients, from 92% for patients under the age of 50 years to 48% for patients older than 70 years.
 - improved over time, from 64% in 2004-2009 to 76% in 2014-2019.

Endometrial stromal sarcoma:				
Table 1 Overview of incidence, prevalence and survival by sex in Belgium, 2010-2019				
Incidence		N	CR	WSR
Incidence, 2010-2019		238	0.4	0.3
Prevalence		N	CR	WSR
5-year prevalence, 31.12.2019		97	1.7	1.1
10-year prevalence, 31.12.2019		167	2.9	1.8
Relative survival		N at risk	%	95%CI
5-year relative survival, 2010-2019		238	73.2	[66.1;79.4]
10-year relative survival, 2010-2019		238	68.2	[58.1;77.1]
Median age at diagnosis, 2010-2019 (y)		56 [Q1: 47; Q3: 68]		

Source: Belgian Cancer Registry 

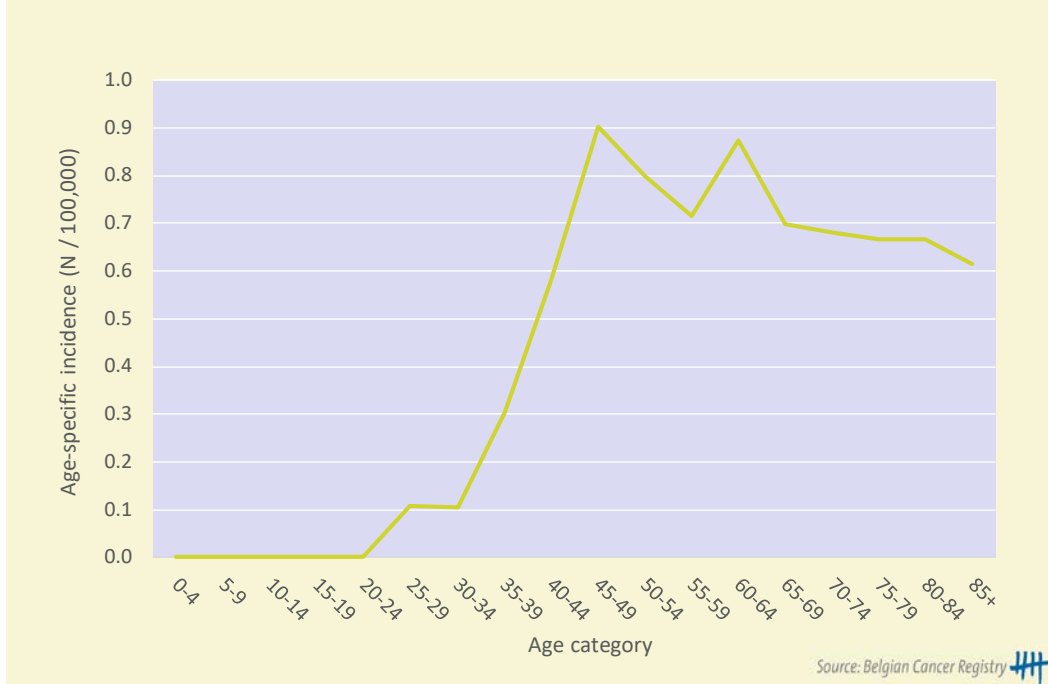
N: number of new diagnoses

CR: crude (all ages) rate (N/100,000 person years)

WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

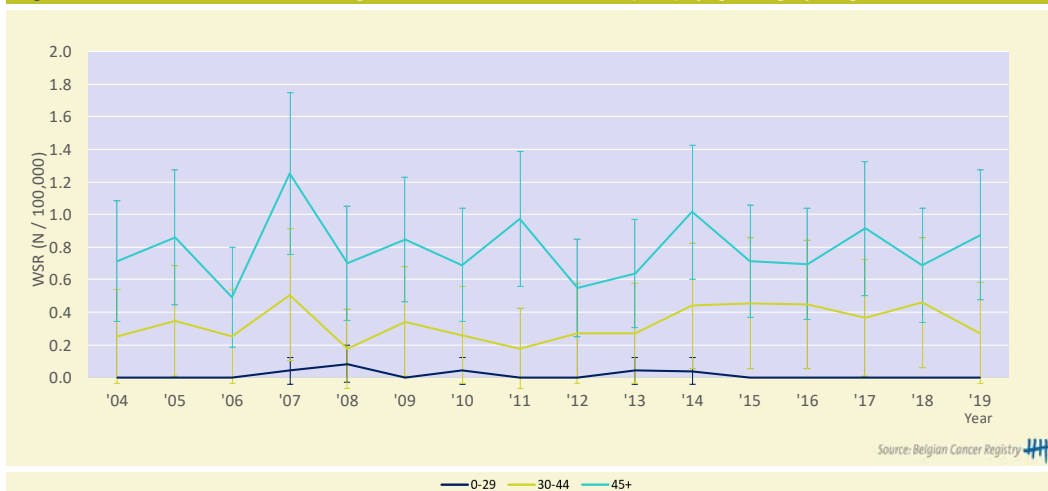
Incidence

Figure 1 Endometrial stromal sarcoma: Age-specific incidence rates (N/100,000) by sex, Belgium 2004-2019



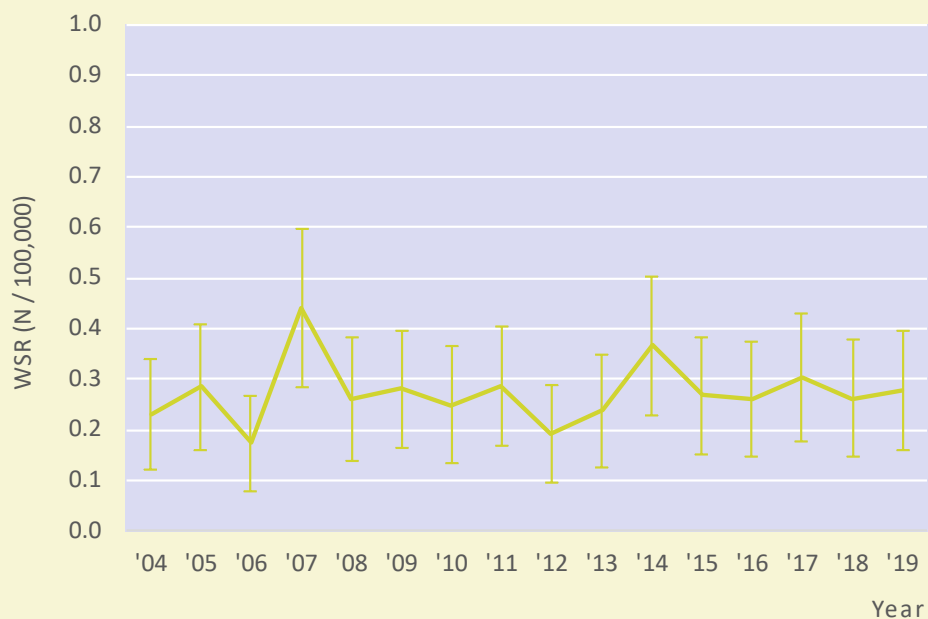
Incidence trends

Figure 2 Endometrial stromal sarcoma: Age-standardised incidence rates* (WSR) by age category, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals.

Figure 3 Endometrial stromal sarcoma:
Age-standardised incidence rates* (WSR), Belgium 2004-2019



Source: Belgian Cancer Registry 

* The age-standardised incidence rates are represented with 95% Confidence Intervals.

Table 2 Endometrial stromal sarcoma:
Incidence trends by age category in Belgium, 2004-2019

Age category	AAPC (%)	95%CI	Period
All ages	0.6	[-2.0; 3.2]	2004-2019
0 - 29 yrs	-	-	-
30 - 44 yrs	2.4	[-1.3; 6.3]	2004-2019
45+ yrs	0.3	[-2.5; 3.1]	2004-2019

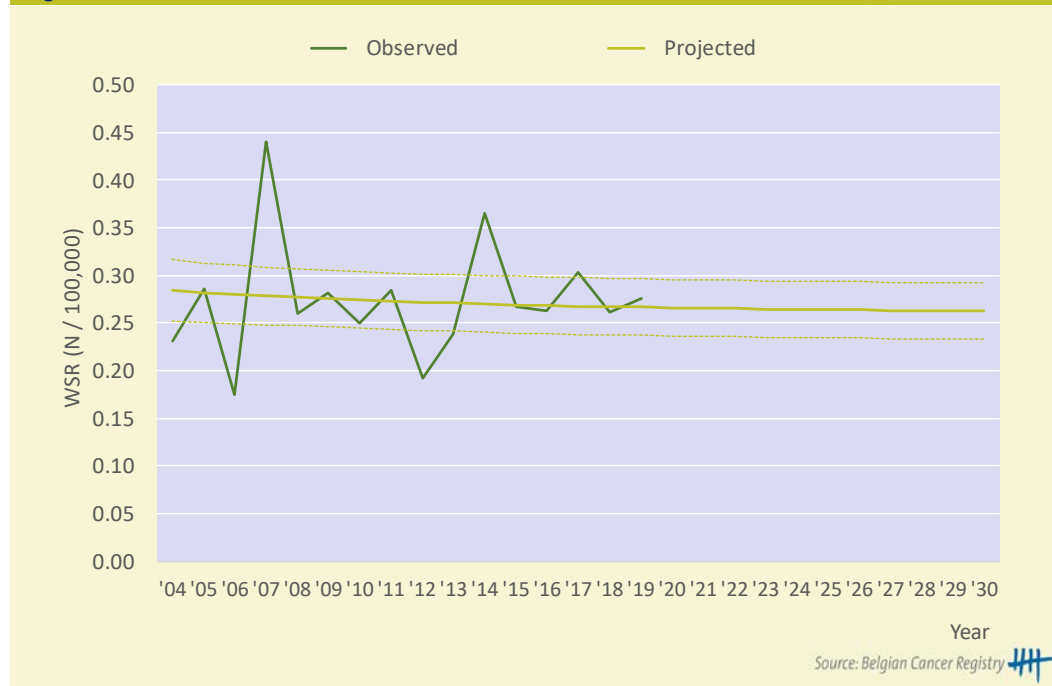
Source: Belgian Cancer Registry 

AAPC: average annual percentage change

Period: when a joinpoint occurred, APC's are calculated for the period before and after the joinpoint. This column represents the corresponding time interval. AAPC's are always calculated over the entire study-period.

Incidence projections

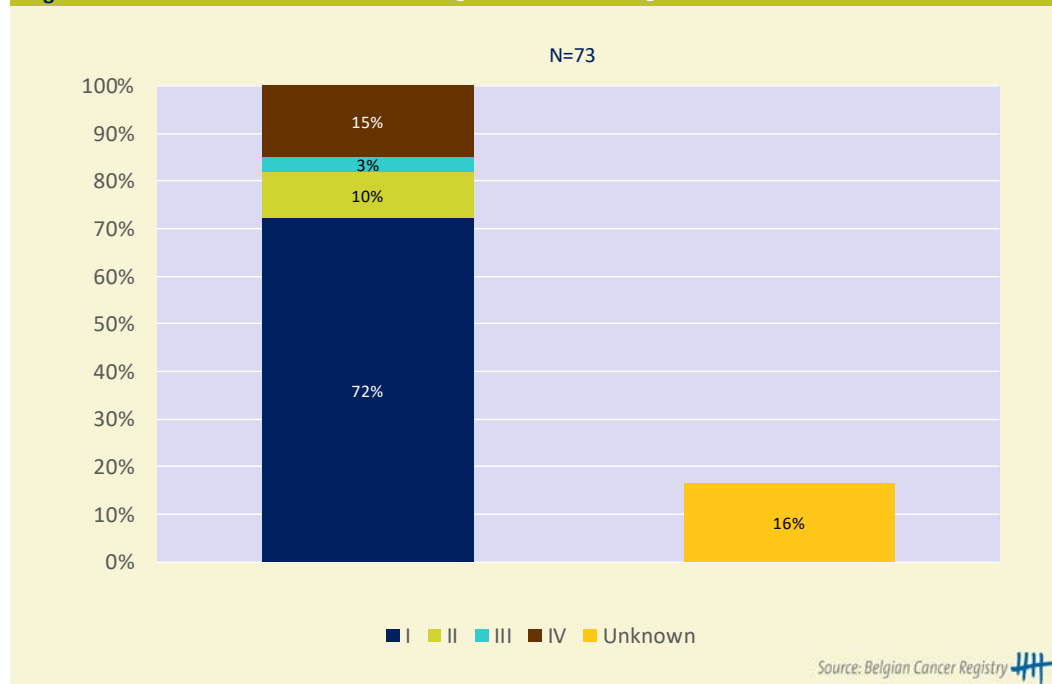
Figure 4 Endometrial stromal sarcoma: Observed and projected* incidence (WSR), Belgium 2004-2030



WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

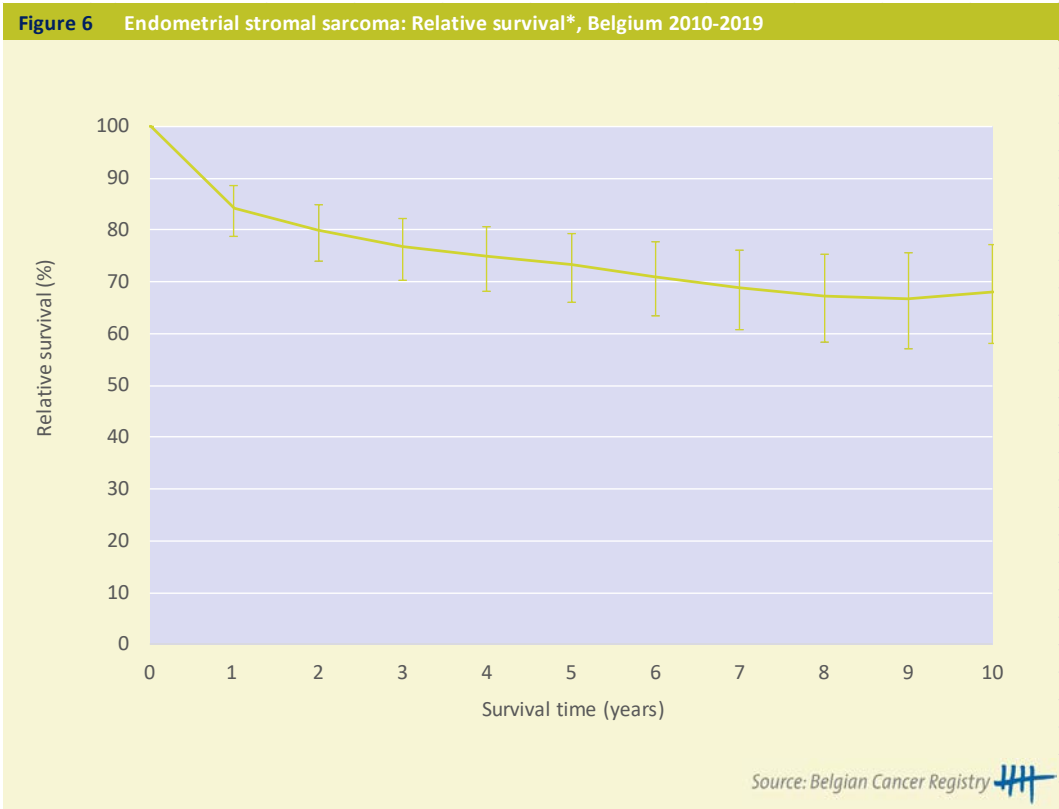
Stage distribution

Figure 5 Endometrial stromal sarcoma: Stage distribution*, Belgium 2017-2019

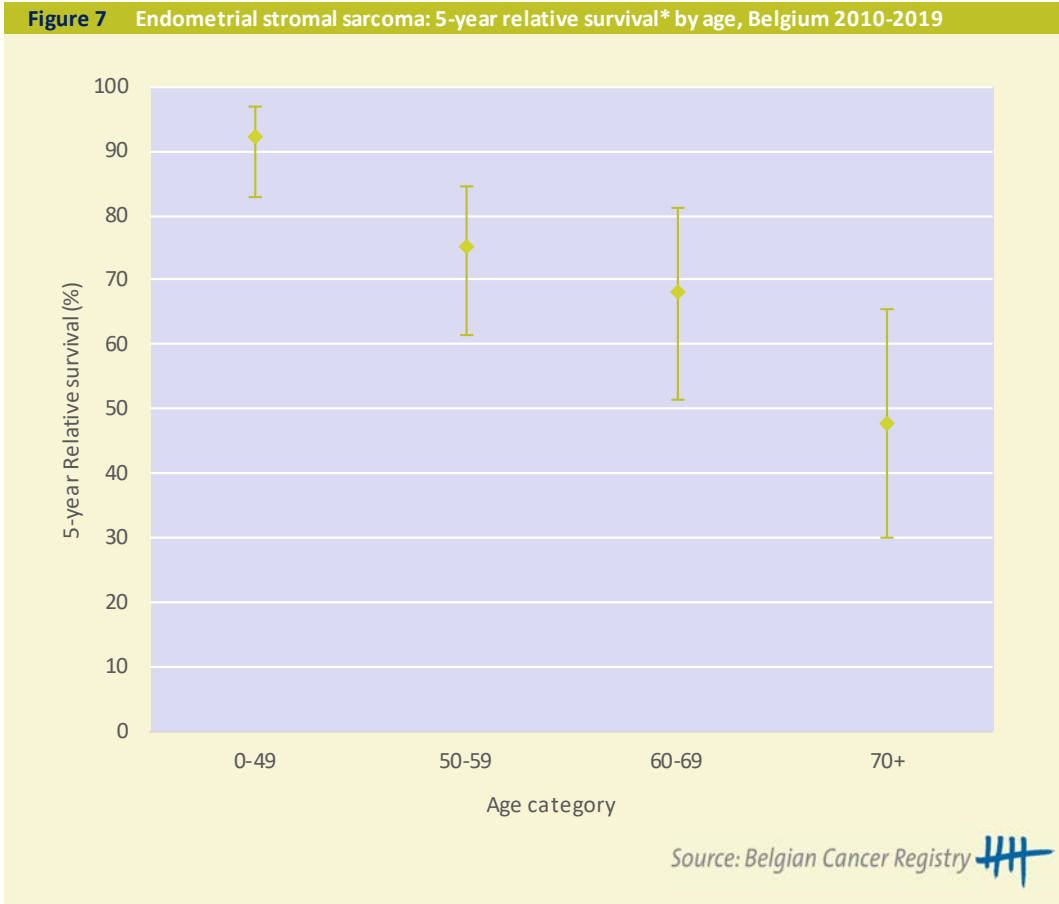


* TNM Classification of Malignant Tumours, Eighth edition, UICC, 2017. Stage combines the pathological (pTNM) and clinical (cTNM). The pTNM prevails over the cTNM but clinical evidence of metastasis is always taken into account.

Survival



* The relative survival values are represented with 95% Confidence Intervals

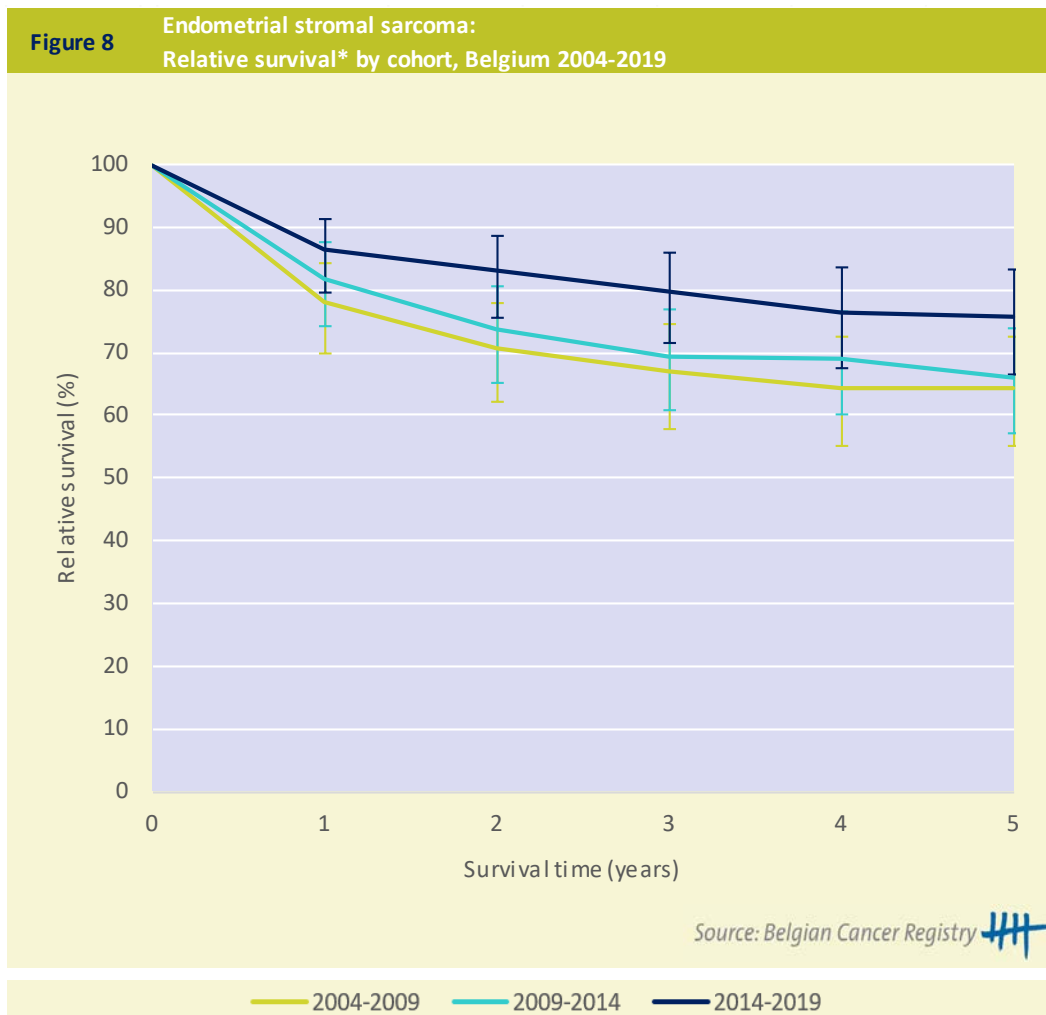


* The relative survival values are represented with 95% Confidence Intervals

Endometrial stromal sarcoma: Conditional 5-year relative survival* in Belgium, 2010-2019		
X years since diagnosis	N at risk	%
1 year	198	84.3
2 year	177	86.2
3 year	144	87.6
4 year	125	89.2
5 year	97	93.1

* Unadjusted 5-yr relative survival probability conditional on surviving the first X years since diagnosis, %
 * Interpretation in lay-man's terms: Given that a patient has already survived X years, what is the relative survival probability 5 years later.

Survival trends



* The relative survival values are represented with 95% Confidence Intervals

3.2.10 EWING SARCOMA

KEYNOTES

Incidence (table 1-2; figure 1-6)

- Ewing sarcomas are mainly diagnosed in children, adolescents and young adults under the age of 25, with a higher proportion of males (male/female ratio of 1.3).
- Although predominantly localised in bones (66%), extra-skeletal locations are diagnosed in 34%, i.e. mostly connective, subcutaneous and other soft tissues of the upper limb.

Survival (table 3; figure 7-10)

- The 5-year relative survival rate of patients with Ewing sarcoma:
 - decreases according to age in females (from 79% in patients younger than 20 to 54% in patients older than 20 years), no relation with age is seen in males with a constant 5-year relative survival rate of ~58%.
 - seems to increase over time, from 57% in 2004-2009 to 64% in 2014-2019 (the improved survival is only attained 3 years after diagnosis).
- The 10-year relative survival rate tends to be better in female patients (62%) than in male patients (49%).

Table 1 Ewing sarcoma: Overview of incidence, prevalence and survival by sex in Belgium, 2010-2019

Males				
Incidence	N	CR	WSR	
Incidence, 2010-2019	139	0.3	0.3	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	48	0.8	1.0	
10-year prevalence, 31.12.2019	82	1.4	1.7	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	139	58.1	[48.7;66.5]	
10-year relative survival, 2010-2019	139	48.9	[38.0;59.1]	
Females				
Incidence	N	CR	WSR	
Incidence, 2010-2019	106	0.2	0.2	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	35	0.6	0.9	
10-year prevalence, 31.12.2019	70	1.2	1.6	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	104	66.2	[55.3;75.1]	
10-year relative survival, 2010-2019	104	62.3	[48.3;73.8]	
Median age at diagnosis, 2010-2019 (y)	22 [Q1: 14; Q3: 40]			
M/F-ratio	1.3			

Source: Belgian Cancer Registry 

N: number of new diagnoses

CR: crude (all ages) rate (N/100,000 person years)

WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

M/F-ratio: Male/Female ratio based on the age-standardised rates (2010-2019)

Incidence

Figure 1 Ewing sarcoma: Age-specific incidence rates (N/100,000) by sex, Belgium 2004-2019

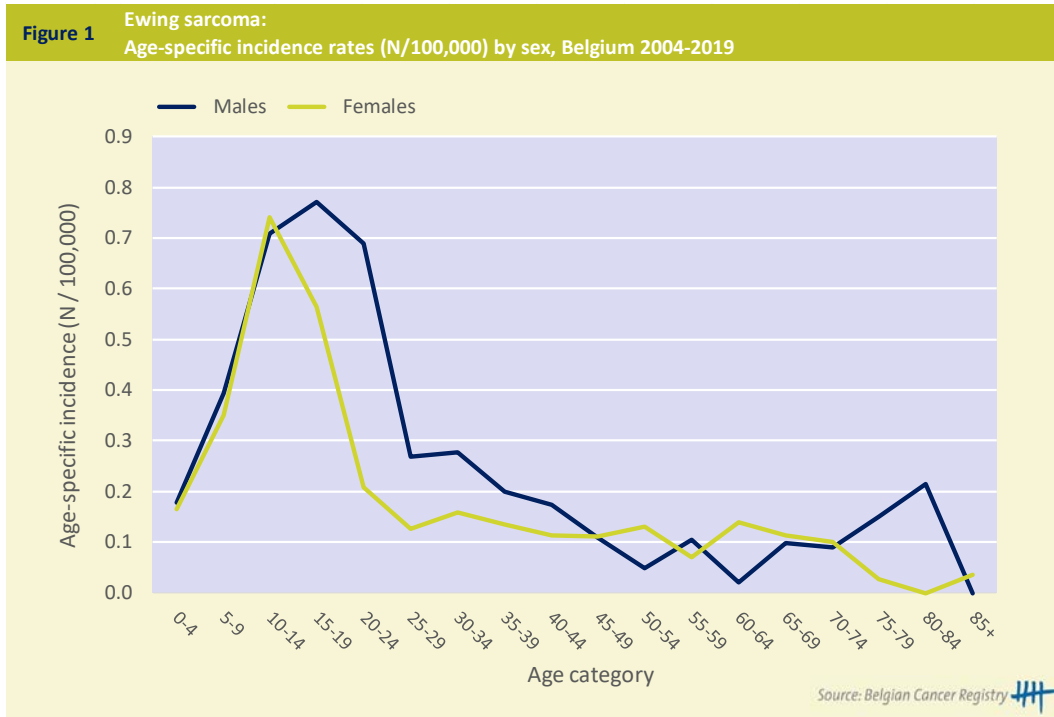


Figure 2 Ewing sarcoma: Incidence distribution by primary tumour location, Belgium 2004-2019

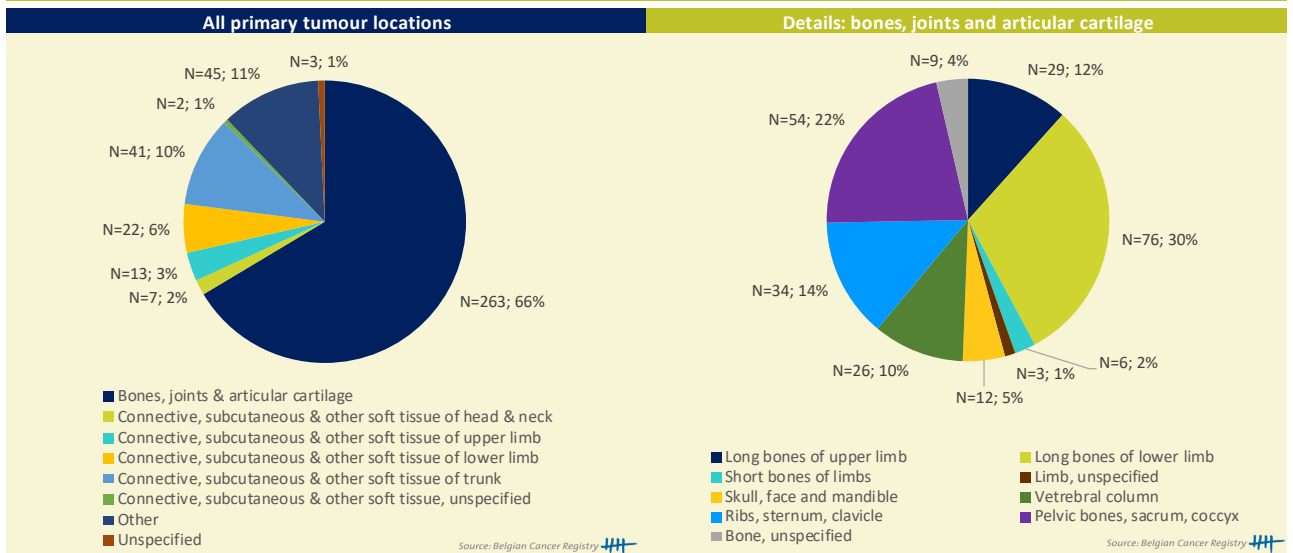
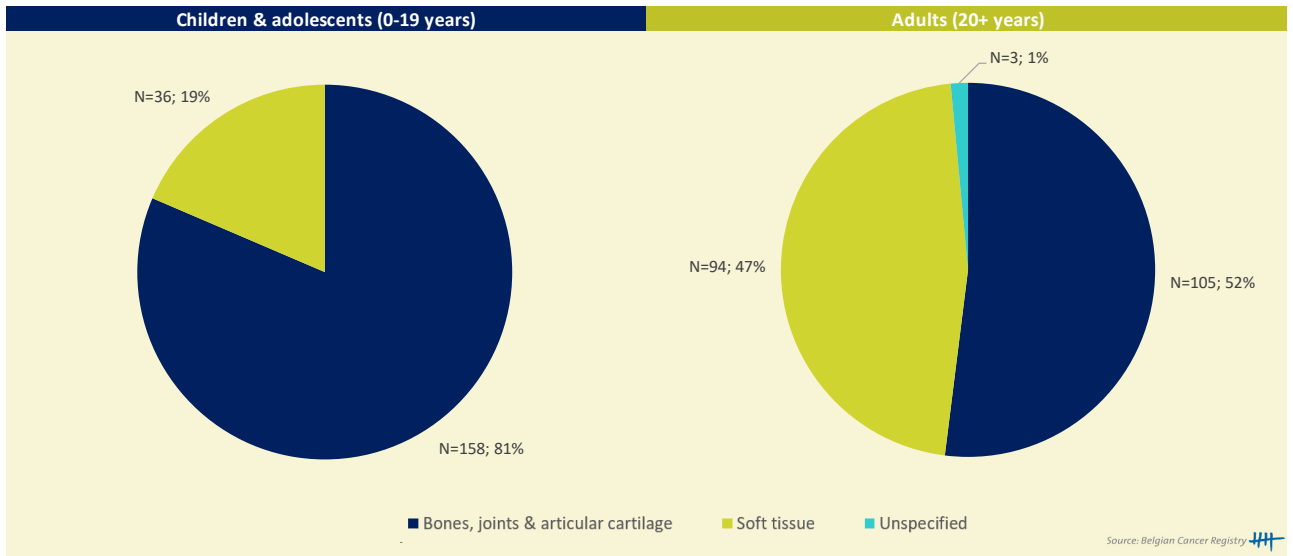
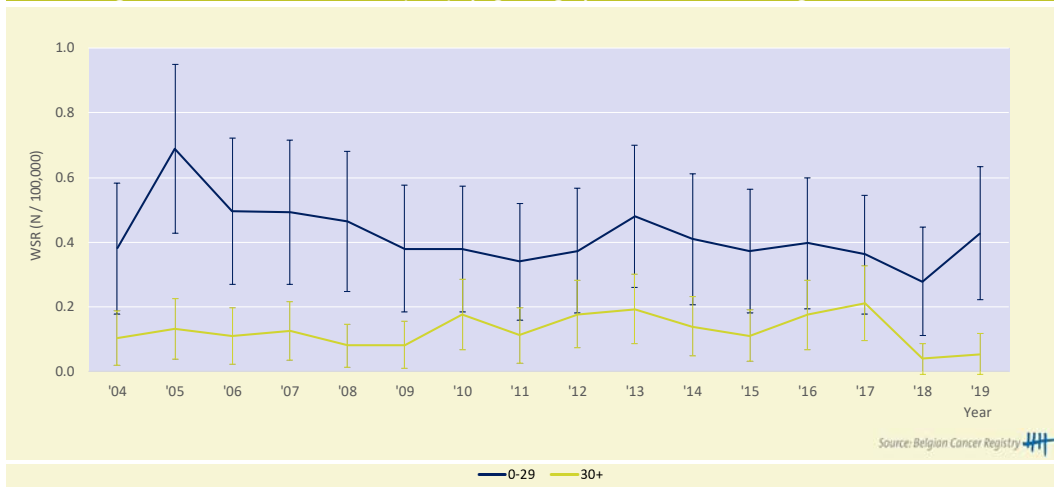


Figure 3 Ewing sarcoma: Incidence distribution by primary tumour location and age group, Belgium 2004-2019



Incidence trends

Figure 4 Ewing sarcoma: Age-standardised incidence rates* (WSR) by age category, males and females, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals.

Figure 5 Ewing sarcoma: Age-standardised incidence rates* (WSR) by sex, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals.

Table 2 Ewing sarcoma: Incidence trends by sex and age category in Belgium, 2004-2019

Age category	Males			Females		
	AAPC (%)	95%CI	Period	AAPC (%)	95%CI	Period
All ages	-2.1	[-4.5; 0.3]	2004-2019	-0.8	[-5.4; 4.1]	2004-2019
0 - 29 y	-3.1	[-5.8; -0.3]	2004-2019	-0.3	[-5.2; 5.0]	2004-2019
30+ y	0.1	[-6.6; 7.4]	2004-2019	-	-	-

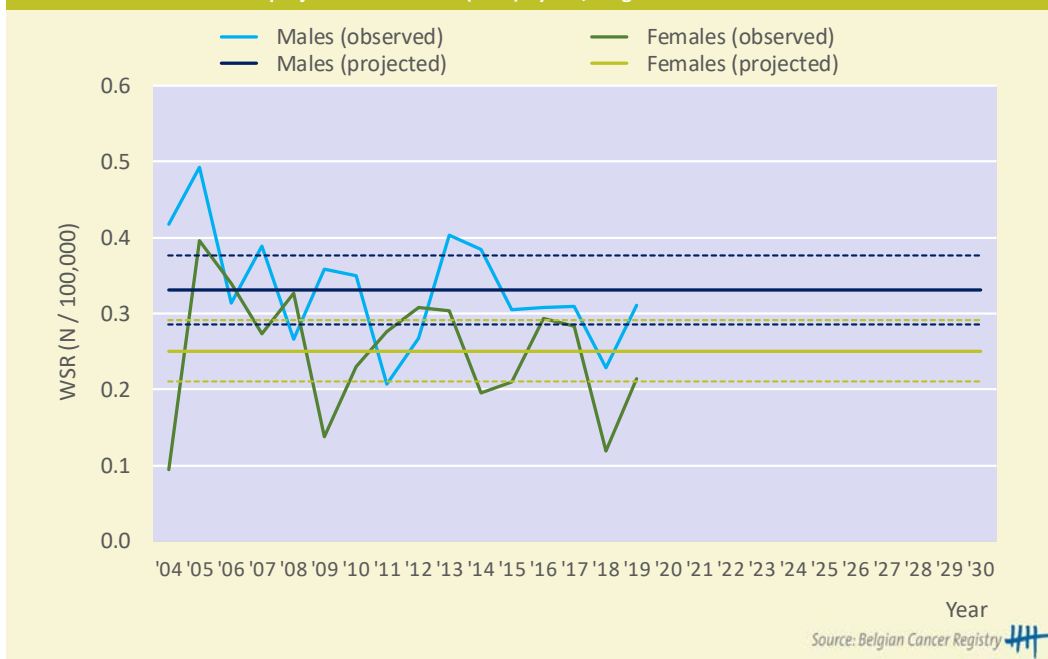
Source: Belgian Cancer Registry

AAPC: average annual percentage change

Period: when a joinpoint occurred, APC's are calculated for the period before and after the joinpoint. This column represents the corresponding time interval. AAPC's are always calculated over the entire study-period.

Incidence projections

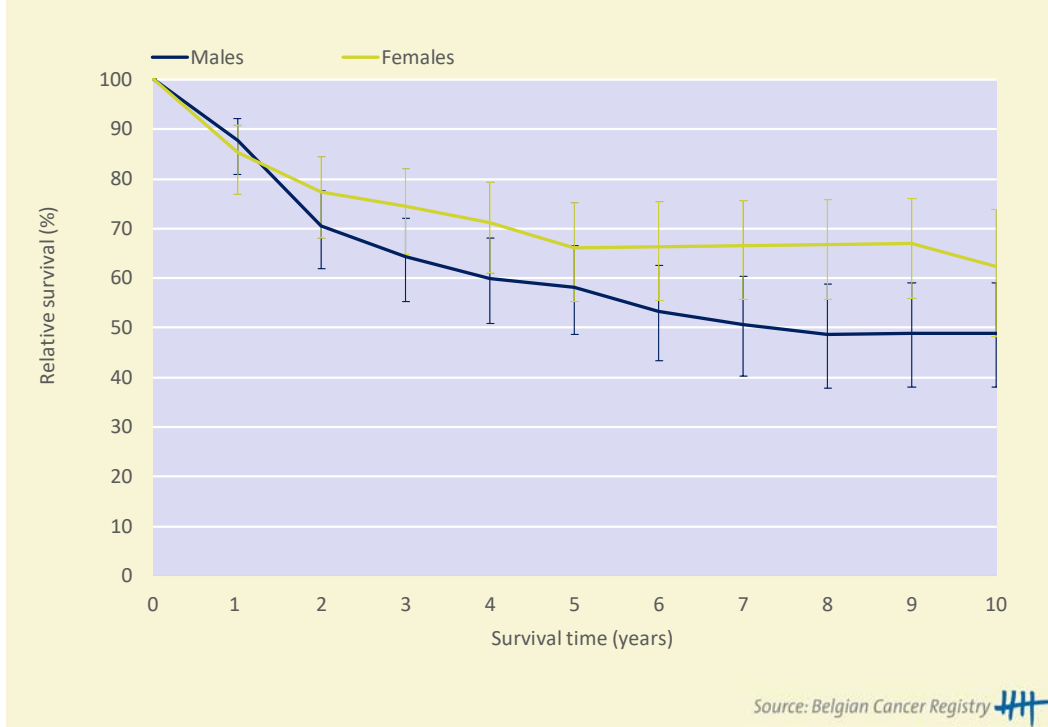
Figure 6 Ewing sarcoma: Observed and projected* incidence (WSR) by sex, Belgium 2004-2030



WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

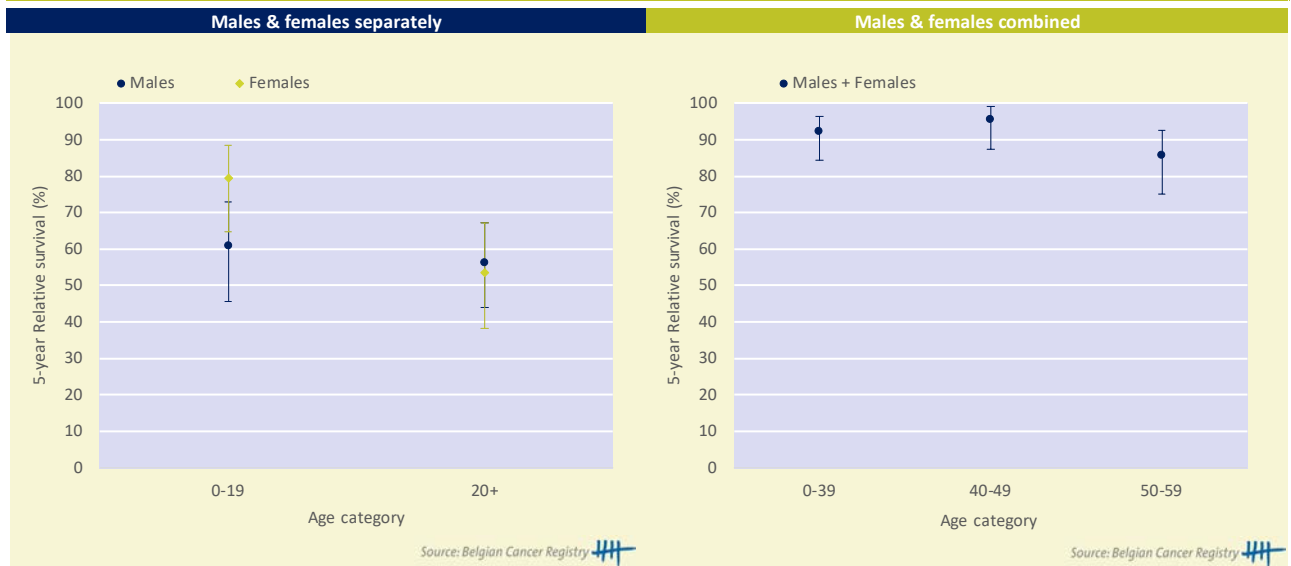
Survival

Figure 7 Ewing sarcoma: Relative survival* by sex, Belgium 2010-2019



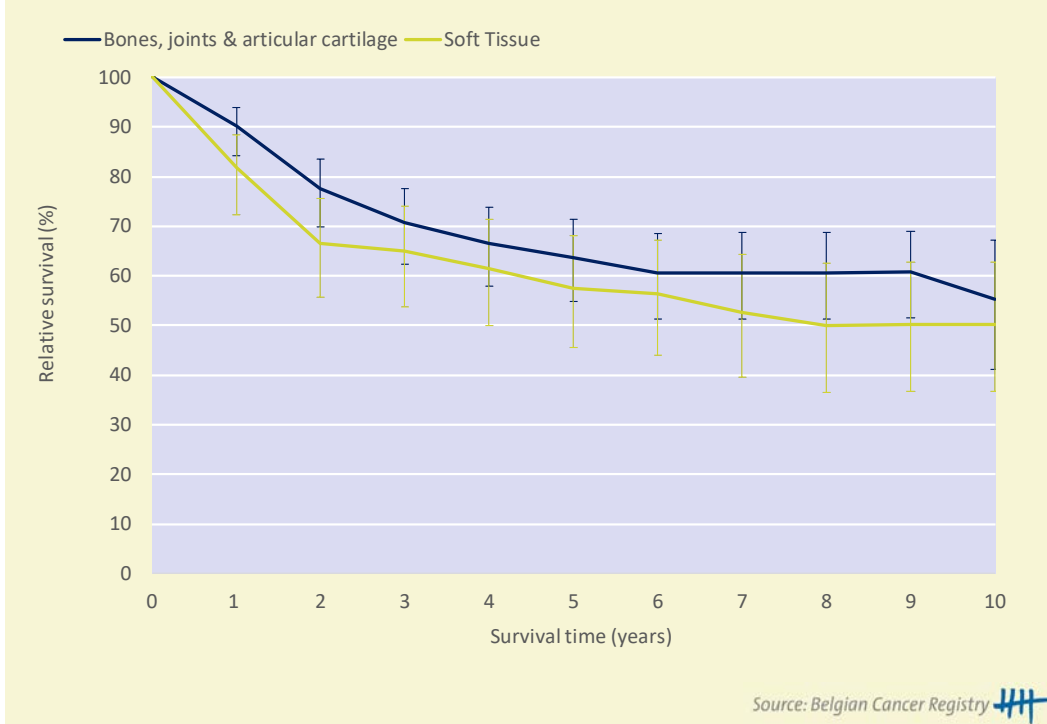
* The relative survival values are represented with 95% Confidence Intervals

Figure 8 Ewing sarcoma: 5-year relative survival* by age and sex, Belgium 2010-2019



* The relative survival values are represented with 95% Confidence Intervals

Figure 9 Ewing sarcoma: Relative survival* by primary tumour location, Belgium 2010-2019

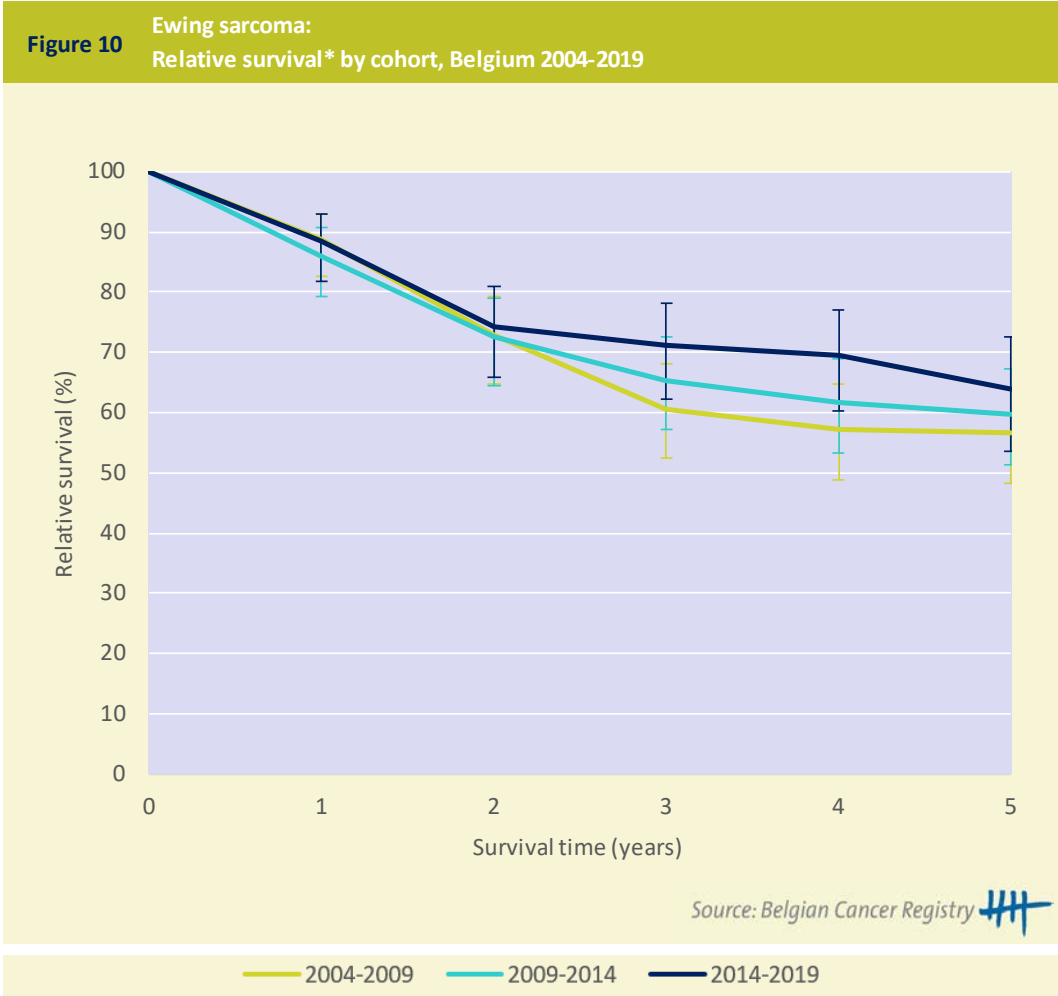


* The relative survival values are represented with 95% Confidence Intervals

X years since diagnosis	N at risk	%
1 year	206	68.2
2 year	163	78.4
3 year	138	82.6

* Unadjusted 5-yr relative survival probability conditional on surviving the first X years since diagnosis, %
 * Interpretation in lay-man's terms: Given that a patient has already survived X years, what is the relative survival probability 5 years later.

Survival trends



* The relative survival values are represented with 95% Confidence Intervals

3.2.11 CHONDROSARCOMA

MAIN SUBTYPES:

- *Chondrosarcoma, NOS grade 2 & 3 and periosteal chondrosarcoma*
- *Extra-skeletal myxoid chondrosarcoma*
- *Dedifferentiated chondrosarcoma*
- *Mesenchymal chondrosarcoma*
- *Clear cell chondrosarcoma*
- *Chondrosarcoma NOS*

KEYNOTES

Incidence (table 1-2; figure 1-6)

- Age-specific incidence rates show that chondrosarcoma is diagnosed in every age category, but with a general increasing incidence with age.
- Chondrosarcomas, NOS and periosteal chondrosarcomas represent the most important (83%) subtypes of chondrosarcomas.
- About half of the chondrosarcomas are diagnosed in the limbs, 31% in the lower limbs and 21% in the upper limbs.
- In 2019 in particular, a lower incidence is observed which could be related to the changes in the classification of sarcomas⁽¹⁻³⁾.
- The exhaustivity of registration of low grade (grade 1) chondrosarcoma is probably incomplete due to changes in the classification of low-grade chondrosarcoma and atypical cartilaginous tumours.

Survival (table 3; figure 7-11)

- The relative survival for males and females with chondrosarcomas is similar, with a 10-year relative survival around 83%.
- Given that a patient survives the first three years after diagnosis, the relative survival probability 5 years later is above 95%.
- The 5-year relative survival rate of chondrosarcomas:
 - Strongly decreases with age, from more than 90% under 40 years of age to 56% in patients older than 70 years.
 - Is higher for intraosseous tumours (85%) than extraosseous tumours (69%).

Table 1 Chondrosarcoma: Overview of incidence, prevalence and survival by sex, Belgium, 2010-2019

Males				
Incidence	N	CR	WSR	
Incidence, 2010-2019	233	0.4	0.3	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	92	1.6	1.2	
10-year prevalence, 31.12.2019	170	3.0	2.0	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	232	78.6	[71.3;84.8]	
10-year relative survival, 2010-2019	232	80.2	[70.2;88.7]	
Females				
Incidence	N	CR	WSR	
Incidence, 2010-2019	221	0.4	0.3	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	77	1.3	1.0	
10-year prevalence, 31.12.2019	181	3.1	2.3	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	221	87.0	[80.9;91.6]	
10-year relative survival, 2010-2019	221	85.4	[77.4;91.6]	
Median age at diagnosis, 2010-2019 (y)	53 [Q1: 41; Q3: 67]			
M/F-ratio	1.0			

Source: Belgian Cancer Registry 

N: number of new diagnoses

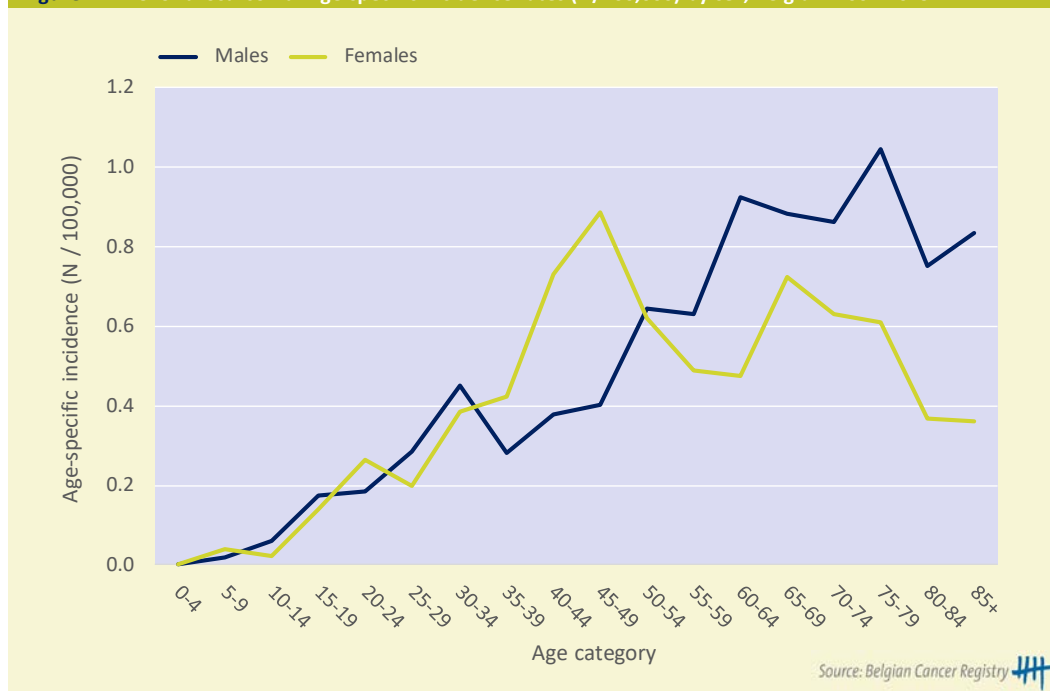
CR: crude (all ages) rate (N/100,000 person years)

WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

M/F-ratio: Male/Female ratio based on the age-standardised rates (2010-2019)

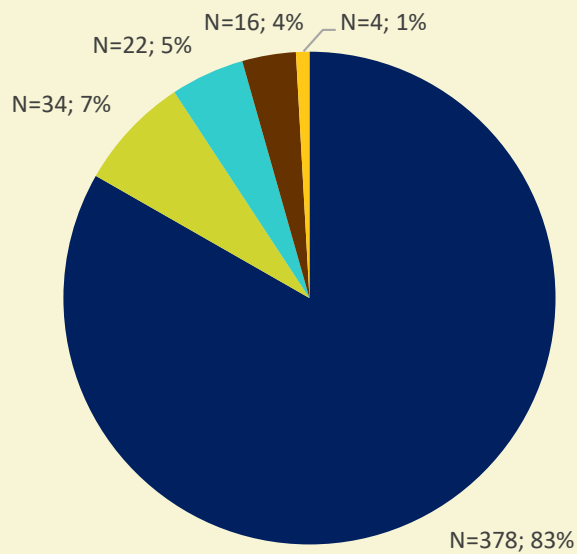
Incidence

Figure 1 Chondrosarcoma: Age-specific incidence rates (N/100,000) by sex, Belgium 2004-2019



Source: Belgian Cancer Registry 

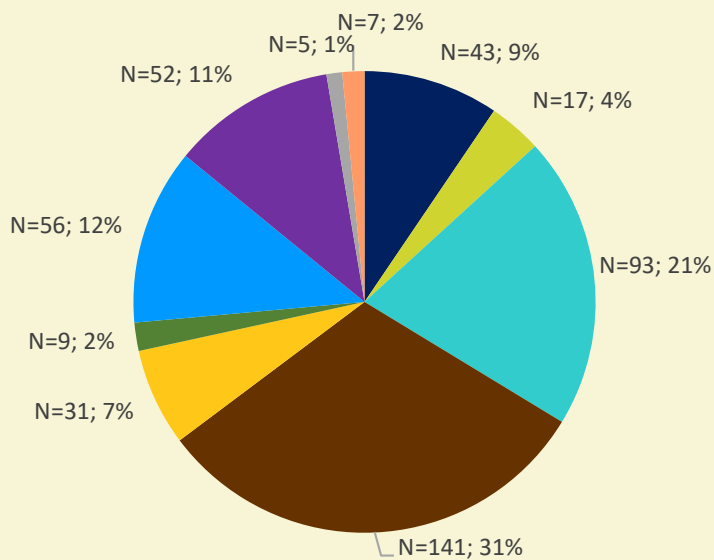
Figure 2 Chondrosarcoma: Incidence distribution by subtype, Belgium 2010-2019



- Chondrosarcoma, NOS grade 2 & 3 and periosteal chondrosarcoma
- Extraskeletal myxoid chondrosarcoma
- Dedifferentiated chondrosarcoma
- Mesenchymal chondrosarcoma
- Clear cell chondrosarcoma

Source: Belgian Cancer Registry

Figure 3 Chondrosarcoma: Incidence distribution by primary tumour location, Belgium 2010-2019

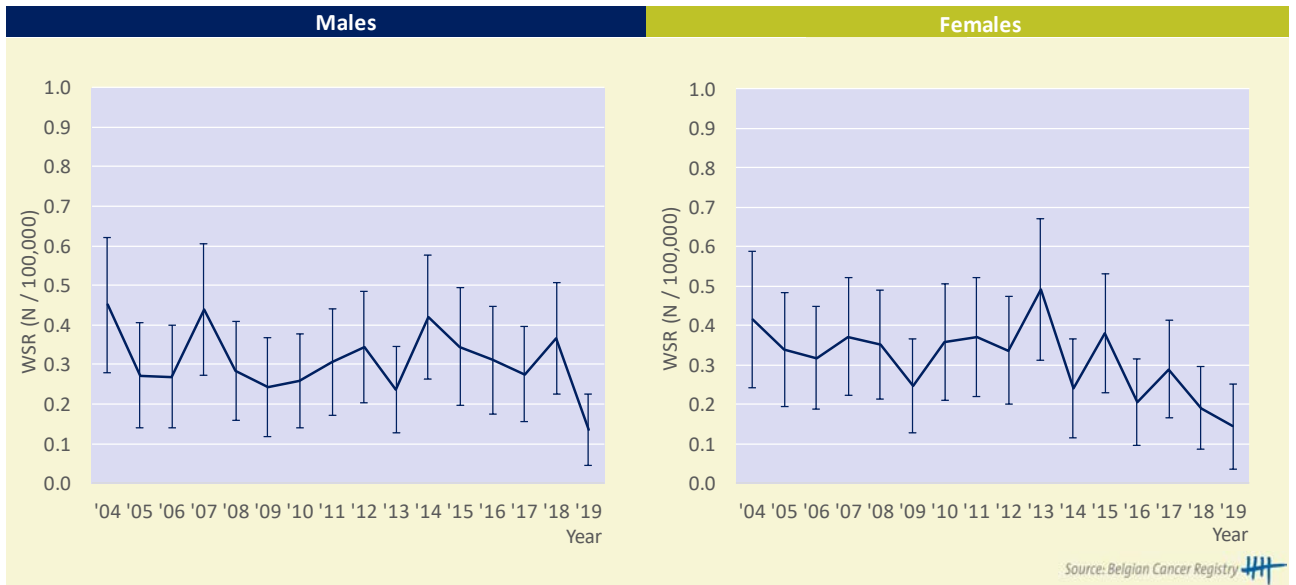


- Connective, subcutaneous & other soft tissue
- Bones of upper limb
- Bones of skull, face and mandible
- Rib, sternum, clavicle
- Bone, unspecified
- Laryngeal cartilage
- Bones of lower limb
- Vetrebral column
- Pelvic bones, sacrum, coccyx
- Other & unspecified

Source: Belgian Cancer Registry

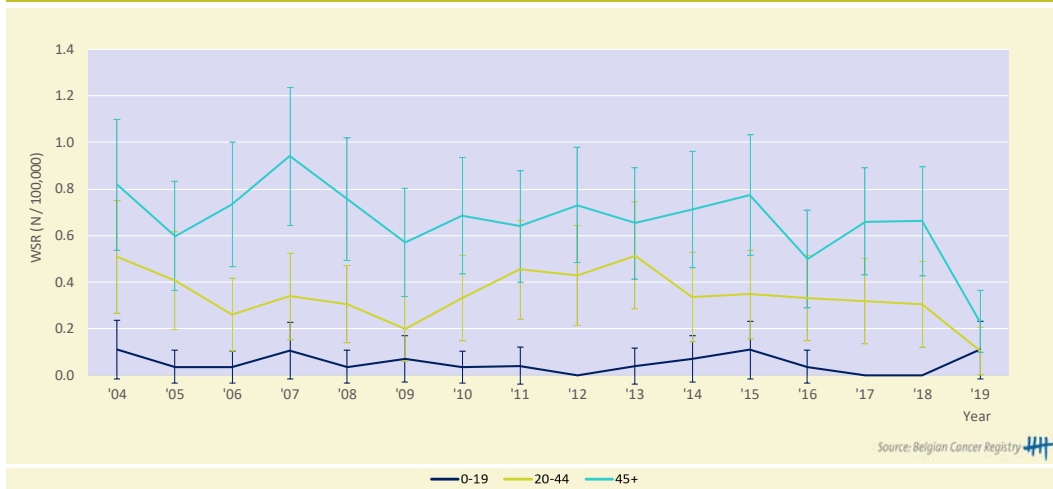
Incidence trends

Figure 4 Chondrosarcoma: Age-standardised incidence rates* (WSR) by sex, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals.

Figure 5 Chondrosarcoma: Age-standardised incidence rates* (WSR) by age category, males and females, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals.

Table 2 Chondrosarcoma: Incidence trends by sex and age category in Belgium, 2004-2019

Age category	Males			Females		
	AAPC (%)	95%CI	Period	AAPC (%)	95%CI	Period
All ages	-1.9	[-5.1; 1.4]	2004-2019	-5.4	[-7.9; -2.9]	2004-2019
				-0.7	[-4.2; 2.9]	2004-2015
				-17.3	[-26.1; -7.5]	2015-2019
0 - 19 y	-	-	-	-	-	-
20 - 44 y	-6.2	[-10.5; -1.8]	2004-2019	-5.3	[-10.2; -0.1]	2004-2019
	-10.2	[-18.6; -0.9]	2004-2011	5.5	[-3.8; 15.7]	2004-2012
	22.8	[4.2; 44.8]	2011-2015	-9.2	[-24.6; 9.4]	2012-2016
	-22.9	[-36.6; -6.2]	2015-2019	-24.8	[-44.6; 2.1]	2016-2019
45+ y	-1.4	[-5.2; 2.5]	2004-2019	-7.2	[-11.6; -2.6]	2004-2019
				2.1	[-18.2; 27.3]	2004-2007
				-3.2	[-9.0; 3.1]	2007-2016
				-25.7	[-40.4; -7.3]	2016-2019

Source: Belgian Cancer Registry

AAPC: average annual percentage change

Period: when a joinpoint occurred, APC's are calculated for the period before and after the joinpoint. This column represents the corresponding time interval. AAPC's are always calculated over the entire study-period.

Incidence projections

Figure 6 Chondrosarcoma: Observed and projected* incidence (WSR), Belgium 2004-2030

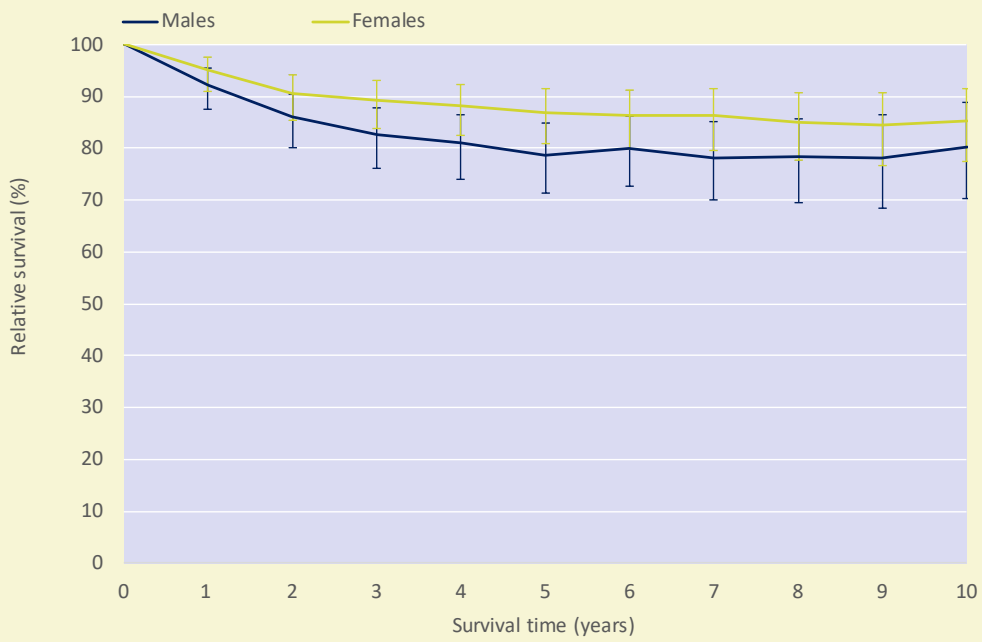


Source: Belgian Cancer Registry

WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

Survival

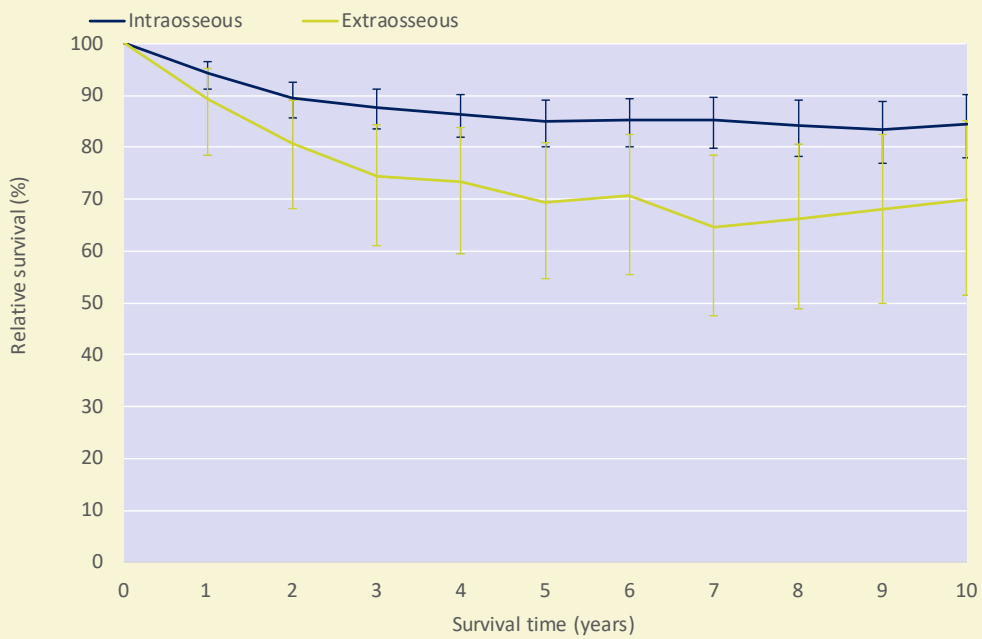
Figure 7 Chondrosarcoma: Relative survival* by sex, Belgium 2010-2019



Source: Belgian Cancer Registry

* The relative survival values are represented with 95% Confidence Intervals

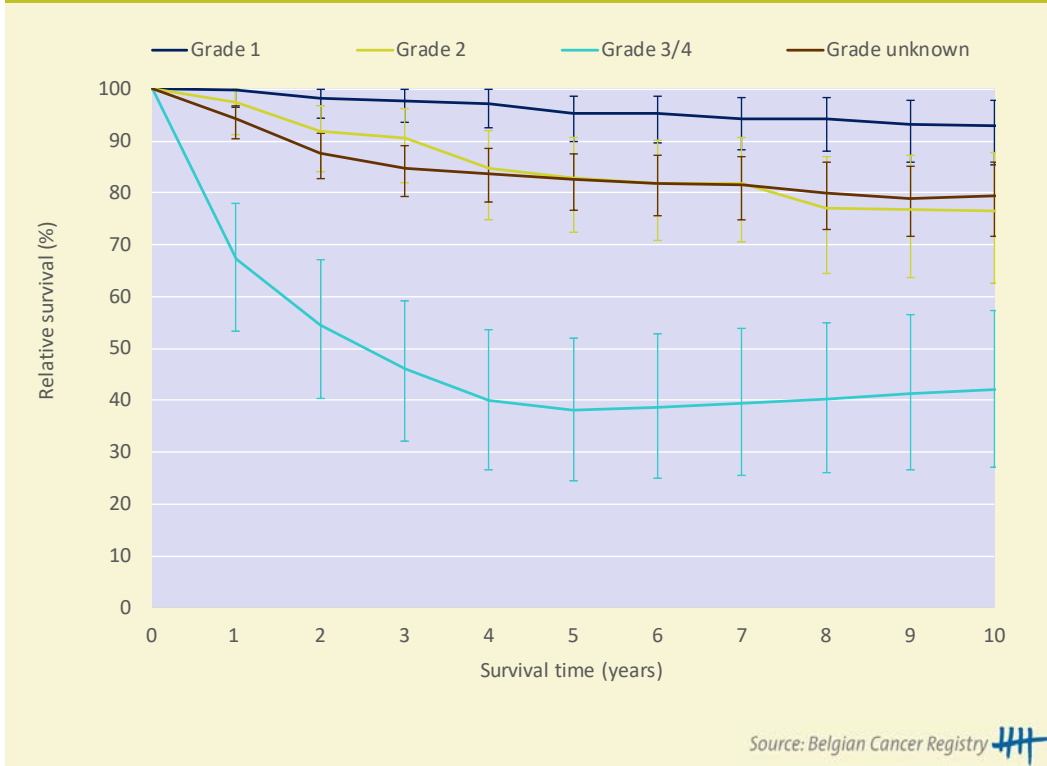
Figure 8 Chondrosarcoma: Relative survival* by primary tumour location, Belgium 2010-2019



Source: Belgian Cancer Registry

* The relative survival values are represented with 95% Confidence Intervals

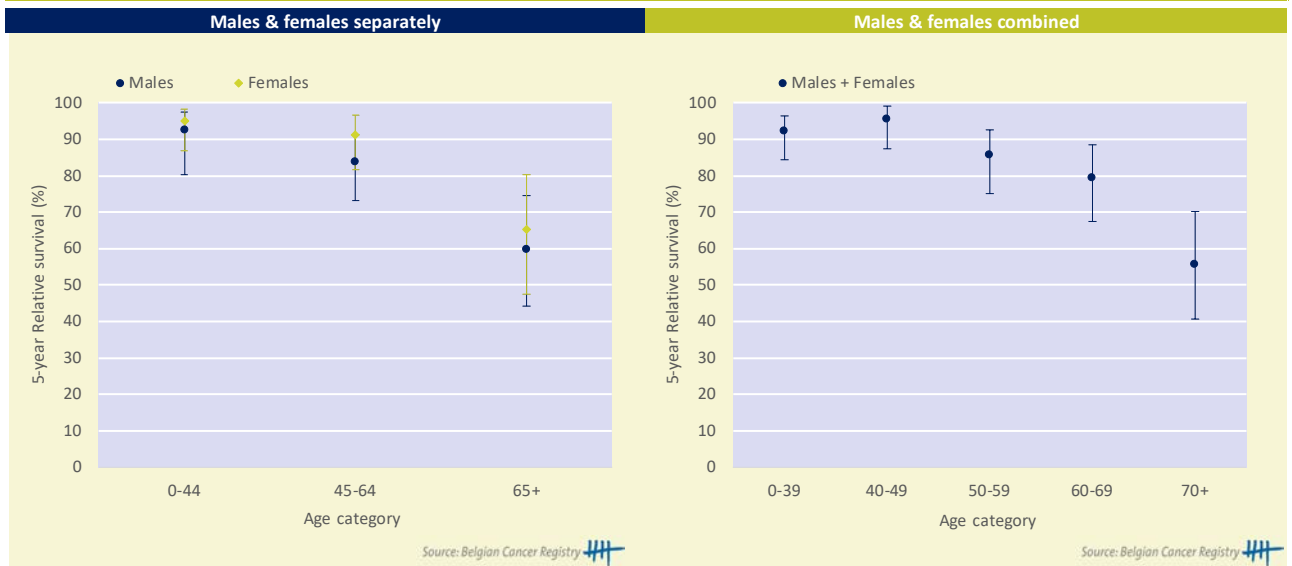
Figure 9 Intraosseous chondrosarcoma: Relative survival* by grade, Belgium 2004-2019



Grade based on the FNCLCC grading or the differentiation grade (WHO). Therefore grade 3 and 4 (which is not included in FNCLCC) were combined. Note that 45% of cases were registered without information on tumour grade (i.e. grade unknown).

* The relative survival values are represented with 95% Confidence Intervals

Figure 10 Chondrosarcoma: 5-year relative survival* by age and sex, Belgium 2010-2019



* The relative survival values are represented with 95% Confidence Intervals

Table 3 Chondrosarcoma: Conditional 5-year relative survival* in Belgium, 2010-2019

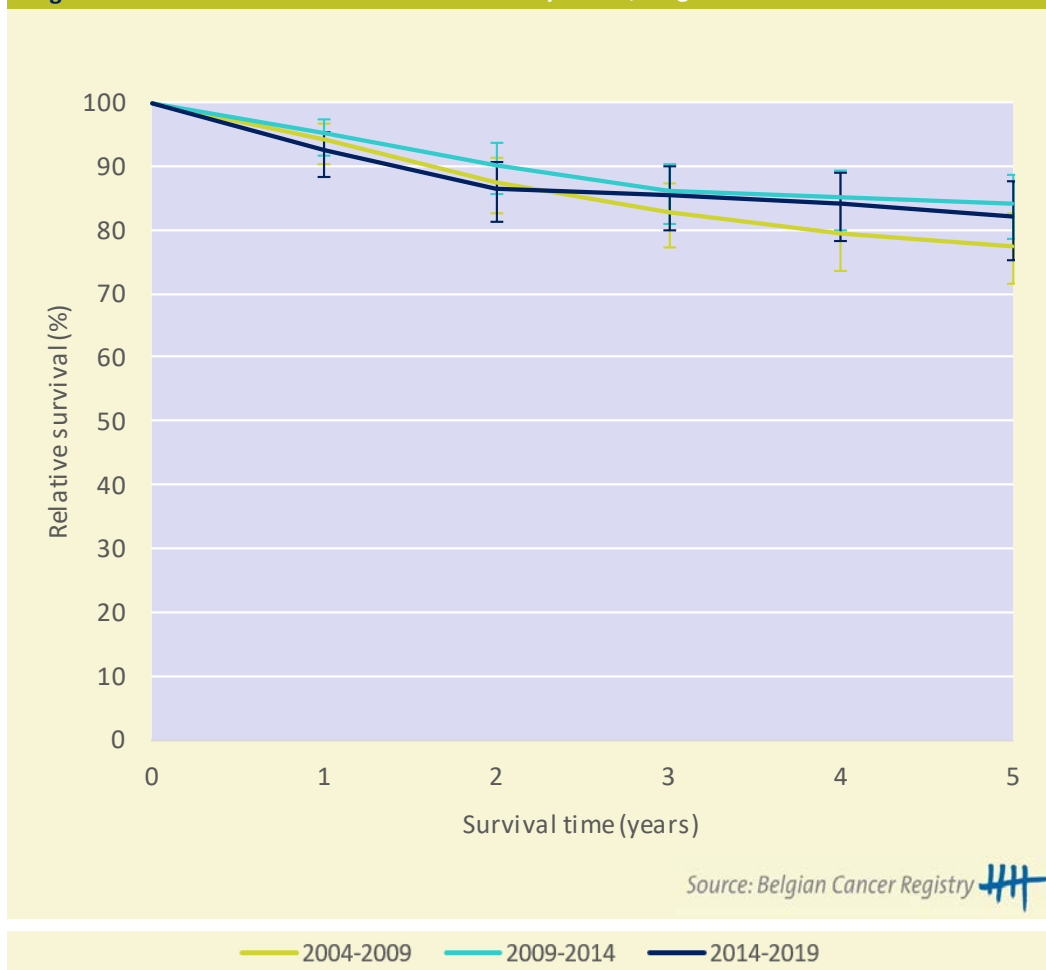
X years since diagnosis	N at risk	%
1 year	414	88.8
2 year	372	93.3
3 year	329	95.2

* Unadjusted 5-yr relative survival probability conditional on surviving the first X years since diagnosis, %

* Interpretation in lay-man's terms: Given that a patient has already survived X years, what is the relative survival probability 5 years later.

Survival trends

Figure 11 Chondrosarcoma: Relative survival* by cohort, Belgium 2004-2019



* The relative survival values are represented with 95% Confidence Intervals

3.2.12 OSTEOSARCOMA

KEYNOTES

Incidence (table 1-2; figure 1-5)

- The age-standardised incidence rate of osteosarcoma is characterised by two peaks:
 - one major peak in adolescence / under the age of 30 years
 - a smaller peak, in patients older than 60 years.
- Half (50%) of the osteosarcomas occur in the lower limbs.
- Osteosarcomas are also diagnosed in extra-skeletal locations, i.e. mostly in connective, subcutaneous and other soft tissues.

Survival (table 3; figure 6-8)

- The 5-year relative survival of patients with osteosarcoma: strongly decreases with age: from 73% in males and 84% in females under the age of 25 to 40% and 45% in males and females older than 60 years.
- The 10-year relative survival tends to be better in females (67%) than in males (55%).

Table 1 Osteosarcoma: Overview of incidence, prevalence and survival by sex in Belgium, 2010-2019

Males				
Incidence	N	CR	WSR	
Incidence, 2010-2019	222	0.4	0.4	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	78	1.4	1.5	
10-year prevalence, 31.12.2019	132	2.3	2.5	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	217	57.7	[50.0;64.7]	
10-year relative survival, 2010-2019	217	55.3	[46.2;63.7]	
Females				
Incidence	N	CR	WSR	
Incidence, 2010-2019	169	0.3	0.3	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	59	1.0	1.0	
10-year prevalence, 31.12.2019	112	1.9	1.9	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	165	67.1	[58.5;74.6]	
10-year relative survival, 2010-2019	165	67.0	[57.5;75.3]	
Median age at diagnosis, 2010-2019 (y)	33 [Q1: 17; Q3: 63]			
M/F-ratio	1.4			

Source: Belgian Cancer Registry 

N: number of new diagnoses

CR: crude (all ages) rate (N/100,000 person years)

WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

M/F-ratio: Male/Female ratio based on the age-standardised rates (2010-2019)

Incidence

Figure 1 Osteosarcoma: Age-specific incidence rates (N/100,000) by sex, Belgium 2004-2019

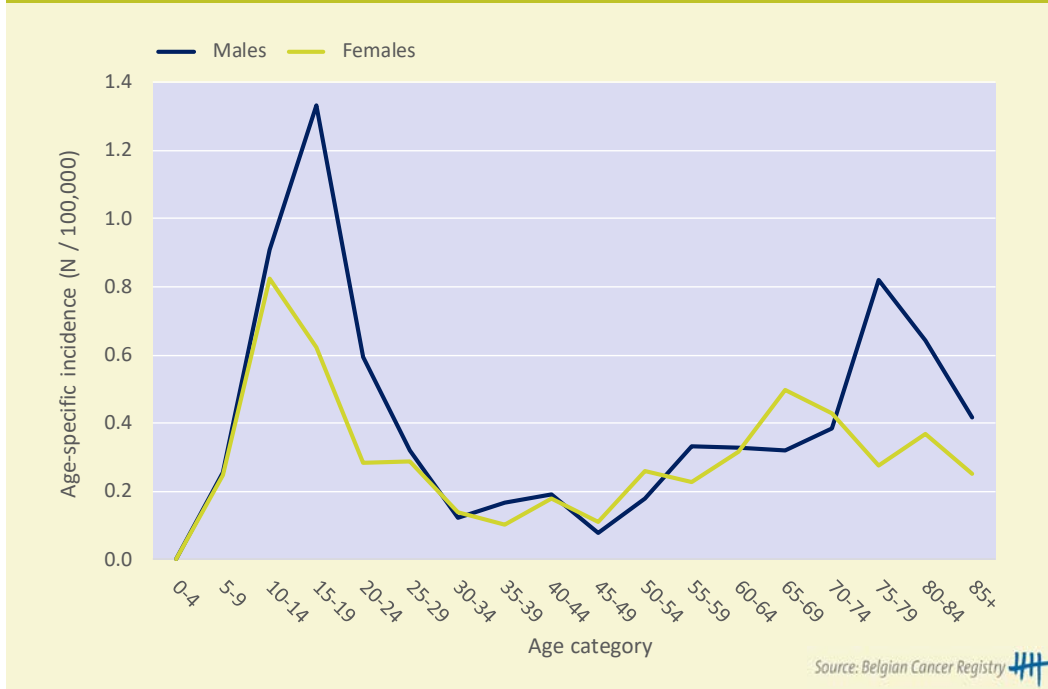
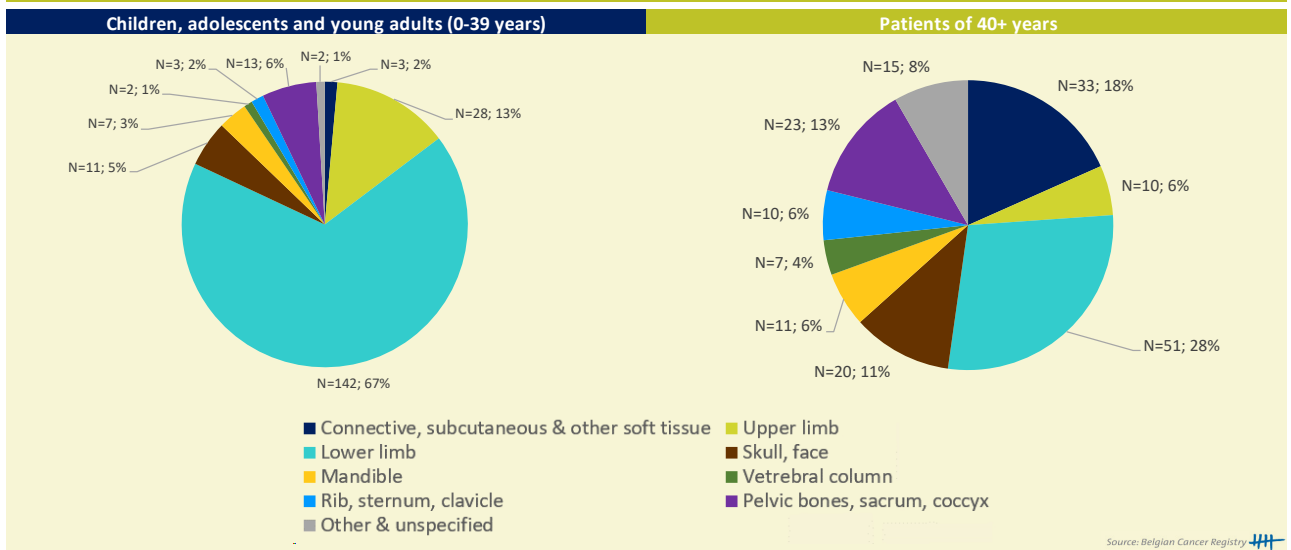
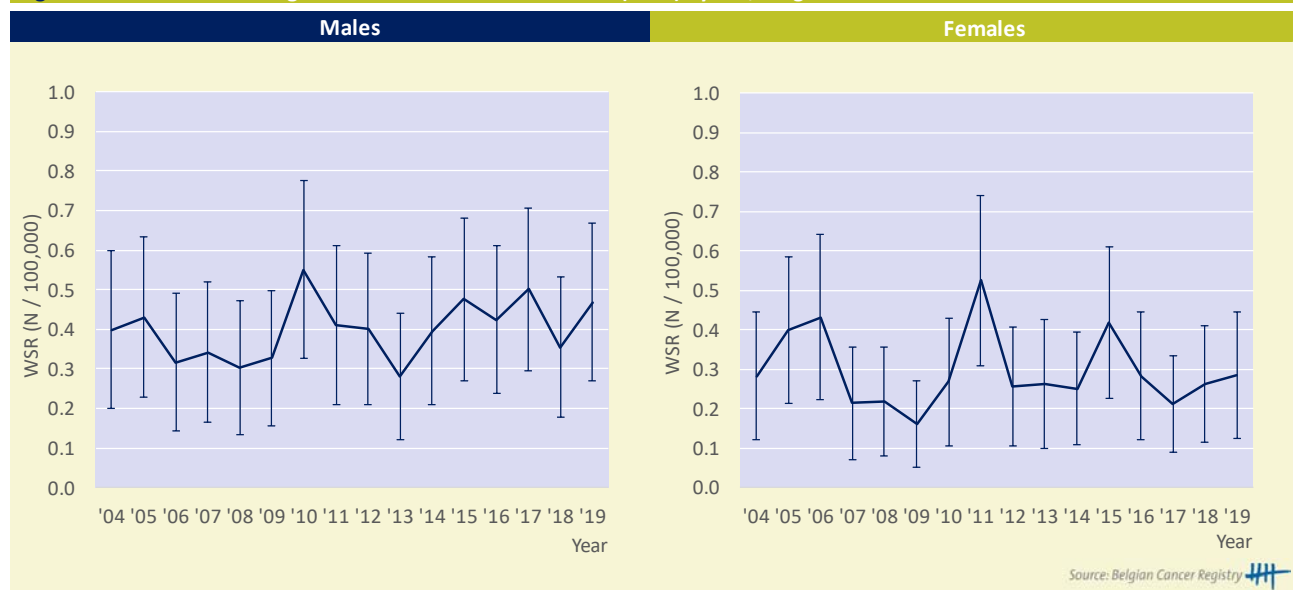


Figure 2 Osteosarcoma: Incidence distribution by primary tumour location, Belgium 2010-2019



Incidence trends

Figure 3 Osteosarcoma: Age-standardised incidence rates* (WSR) by sex, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals.

Figure 4 Osteosarcoma: Age-standardised incidence rates* (WSR) by age category, males and females, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals.

Table 2 Osteosarcoma: Incidence trends by sex and age category in Belgium, 2004-2019

Age category	Males			Females		
	AAPC (%)	95%CI	Period	AAPC (%)	95%CI	Period
All ages	1.3	[-0.8; 3.5]	2004-2019	-0.7	[-4.2; 3.0]	2004-2019
0 - 29 y	0.3	[-1.7; 2.3]	2004-2019	-1.2	[-5.8; 3.7]	2004-2019
30 - 59 y	4.8	[-3.3; 13.5]	2004-2019	-0.7	[-7.8; 6.8]	2004-2019
60+ y	5.1	[0.0; 10.6]	2004-2019	2.1	[-7.8; 13.0]	2004-2019

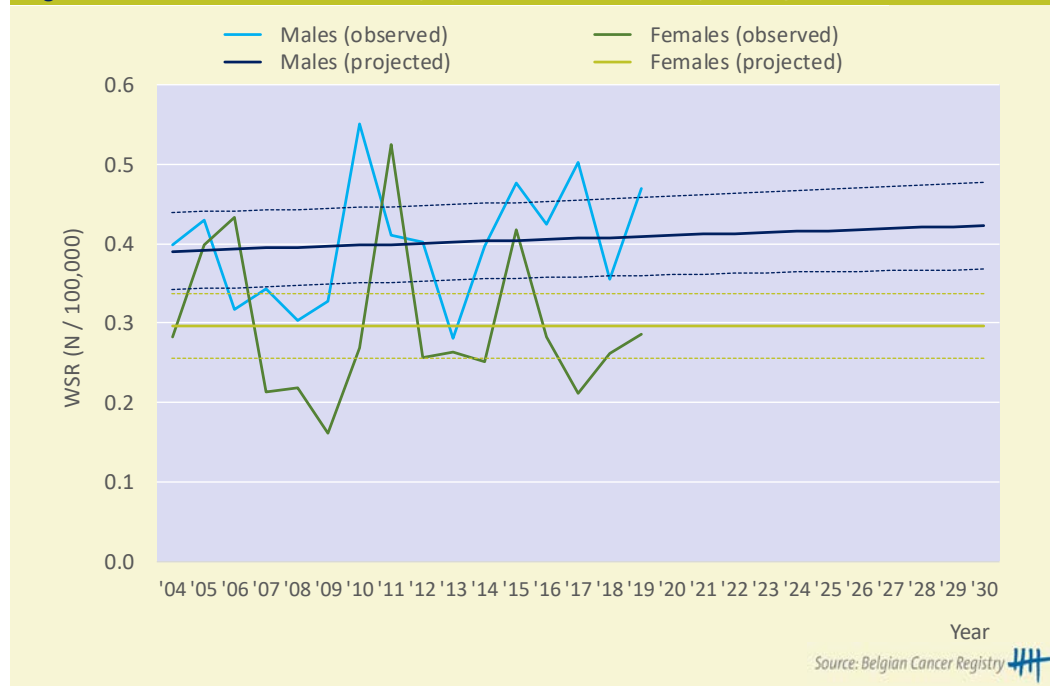
Source: Belgian Cancer Registry

AAPC: average annual percentage change

Period: when a joinpoint occurred, APC's are calculated for the period before and after the joinpoint. This column represents the corresponding time interval. AAPC's are always calculated over the entire study-period.

Incidence projections

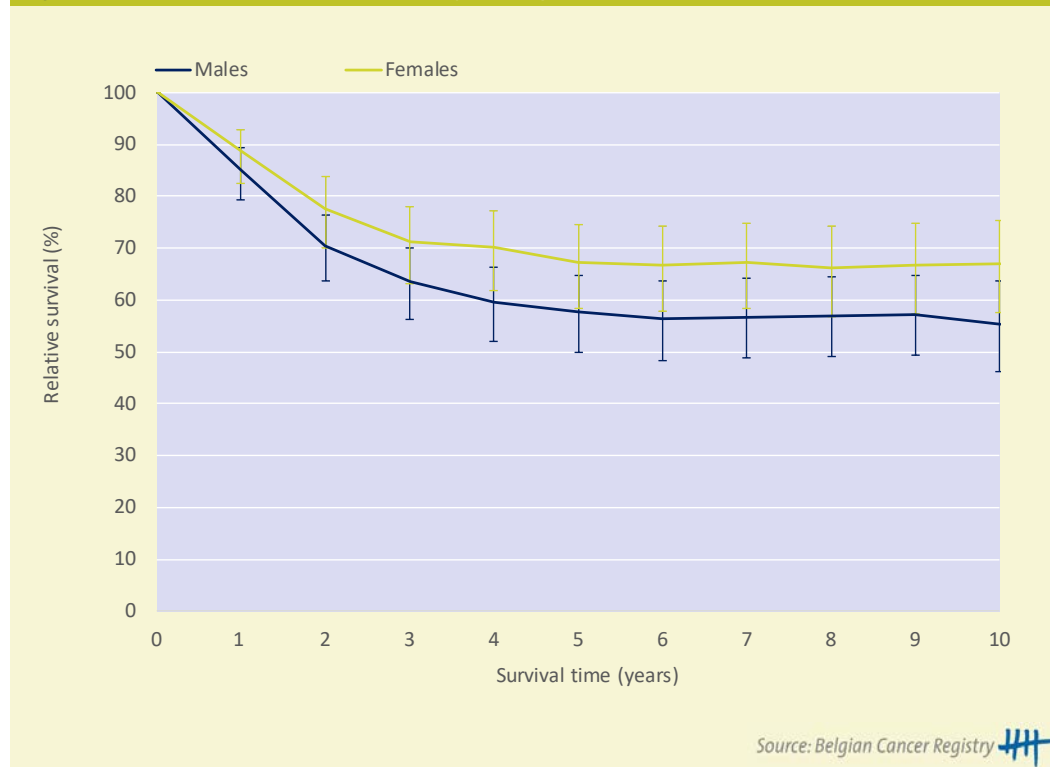
Figure 5 Osteosarcoma: Observed and projected* incidence (WSR) by sex, Belgium 2004-2030



WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

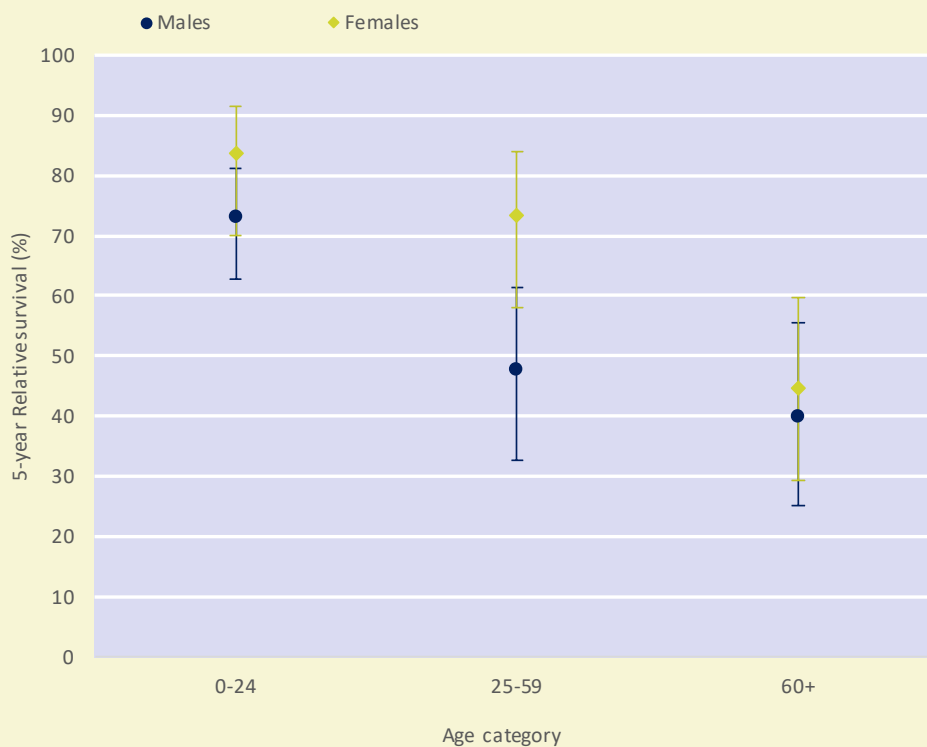
Survival

Figure 6 Osteosarcoma: Relative survival* by sex, Belgium 2010-2019



* The relative survival values are represented with 95% Confidence Intervals

Figure 7 Osteosarcoma:
5-year relative survival* by age and sex, Belgium 2010-2019



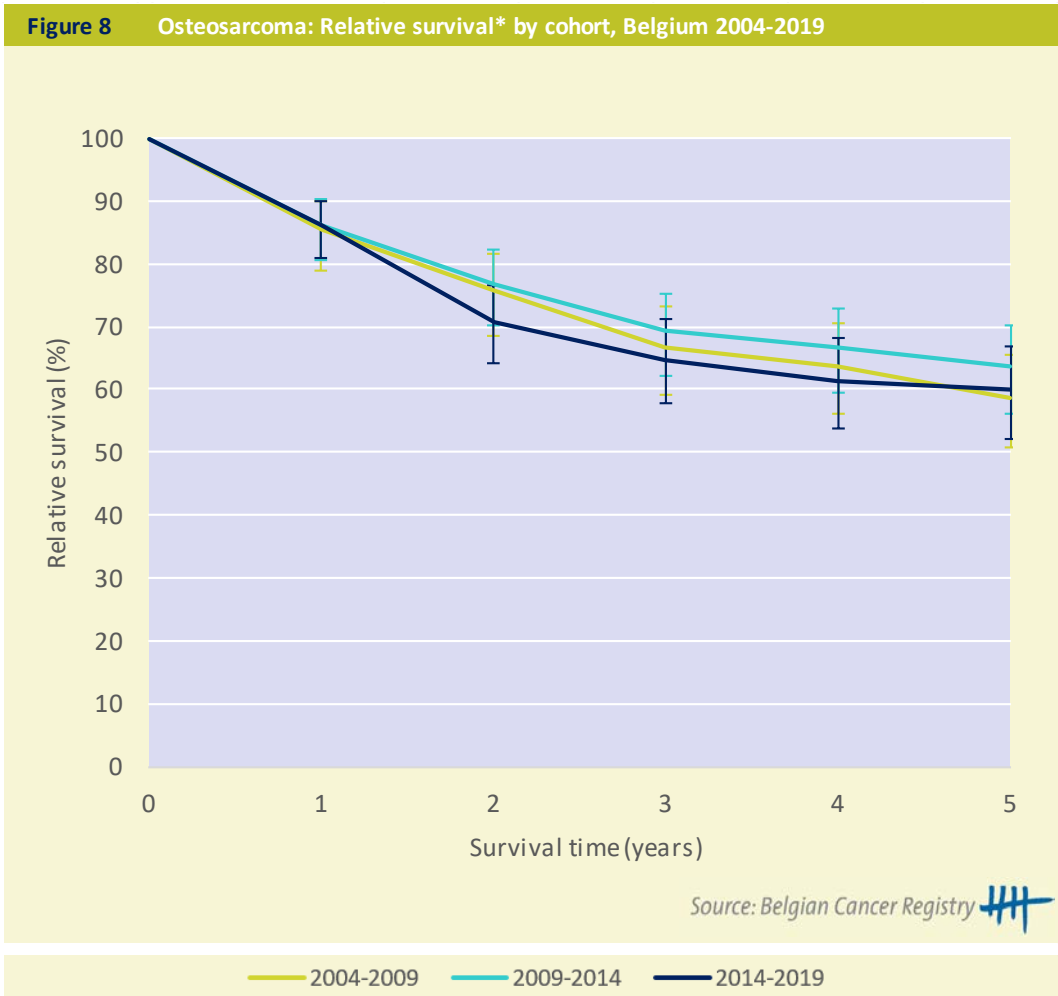
Source: Belgian Cancer Registry

* The relative survival values are represented with 95% Confidence Intervals

X years since diagnosis	N at risk	%
1 year	324	70.2
2 year	248	83.4
3 year	206	91.1

* Unadjusted 5-yr relative survival probability conditional on surviving the first X years since diagnosis, %
* Interpretation in lay-man's terms: Given that a patient has already survived X years, what is the relative survival probability 5 years later.

Survival trends



* The relative survival values are represented with 95% Confidence Intervals

3.2.13 OTHER BONE TUMOURS OF UNCERTAIN DIFFERENTIATION

MAIN SUBTYPES:

- Adamantinoma of long bones
- Chordoma

KEYNOTES

Incidence (table 1-2; figure 1-7)

- The age-specific incidence rate of other bone tumours of uncertain differentiation shows two incidence peaks:
 - A peak in adolescents and young adults (under the age of 30 years)
 - A second higher peak in adults older than 70 years of age
- The incidence of chordomas increases with age, conversely, the incidence of adamantinomas of long bones decreases with age.
- About a third (33%) of the other bone tumours of uncertain differentiation are diagnosed in the bones of the skull and face.

Survival (table 3; figure 8-10)

- The relative survival of patients with other bone tumours of uncertain differentiation does not show a consistent difference between males and females.
- The 5-year relative survival does not show a consistent improvement over time (2004-2019).

Table 1 Other bone tumours of uncertain differentiation: Overview of incidence, prevalence and survival by sex in Belgium, 2010-2019				
Males				
Incidence	N	CR	WSR	
Incidence, 2010-2019	76	0.1	0.1	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	38	0.7	0.4	
10-year prevalence, 31.12.2019	55	1.0	0.6	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	74	82.6	[68.5;92.8]	
10-year relative survival, 2010-2019	74	64.7	[32.7;90.6]	
Females				
Incidence	N	CR	WSR	
Incidence, 2010-2019	65	0.1	0.1	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	29	0.5	0.4	
10-year prevalence, 31.12.2019	52	0.9	0.7	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	65	86.0	[71.4;94.9]	
10-year relative survival, 2010-2019	65	76.5	[56.8;90.5]	
Median age at diagnosis, 2010-2019 (y)	60 [Q1: 44; Q3: 71]			
M/F-ratio	1.0			

Source: Belgian Cancer Registry 

N: number of new diagnoses

CR: crude (all ages) rate (N/100,000 person years)

WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

M/F-ratio: Male/Female ratio based on the age-standardised rates (2010-2019)

Incidence

Figure 1

Other bone tumours of uncertain differentiation:
Age-specific incidence rates (N/100,000) by sex, Belgium 2004-2019

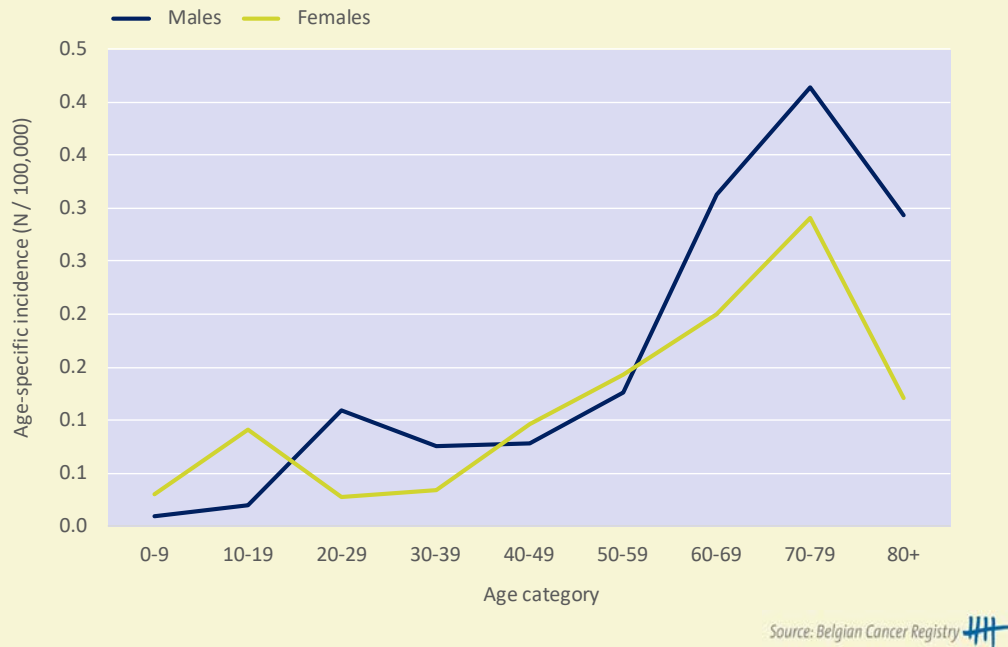


Figure 2

Other bone tumours of uncertain differentiation:
Incidence distribution by primary tumour location, Belgium 2004-2019

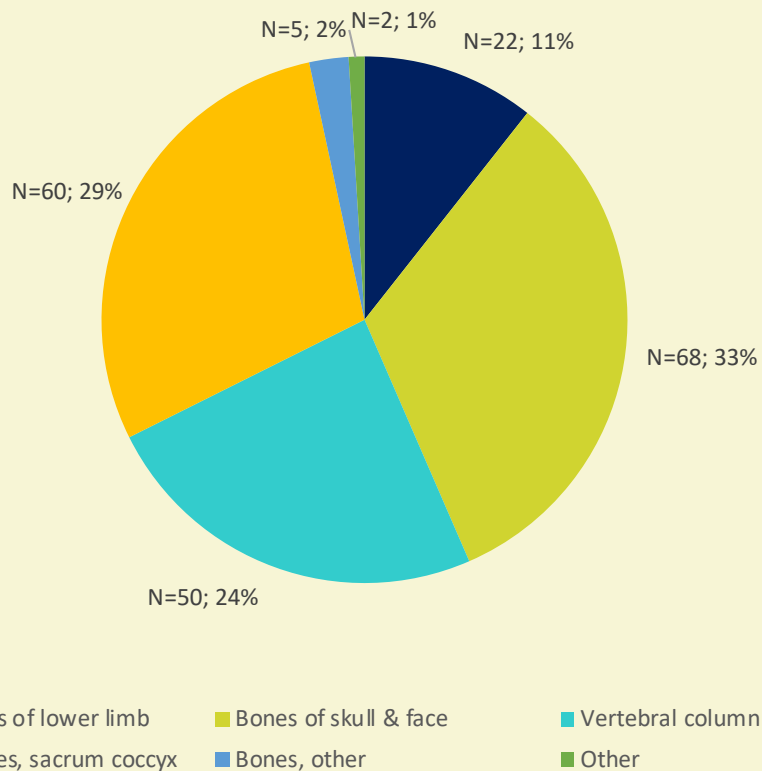


Figure 3 Other bone tumours of uncertain differentiation: Age-specific incidence rates (N/100,000) by subtype, Belgium 2004-2019

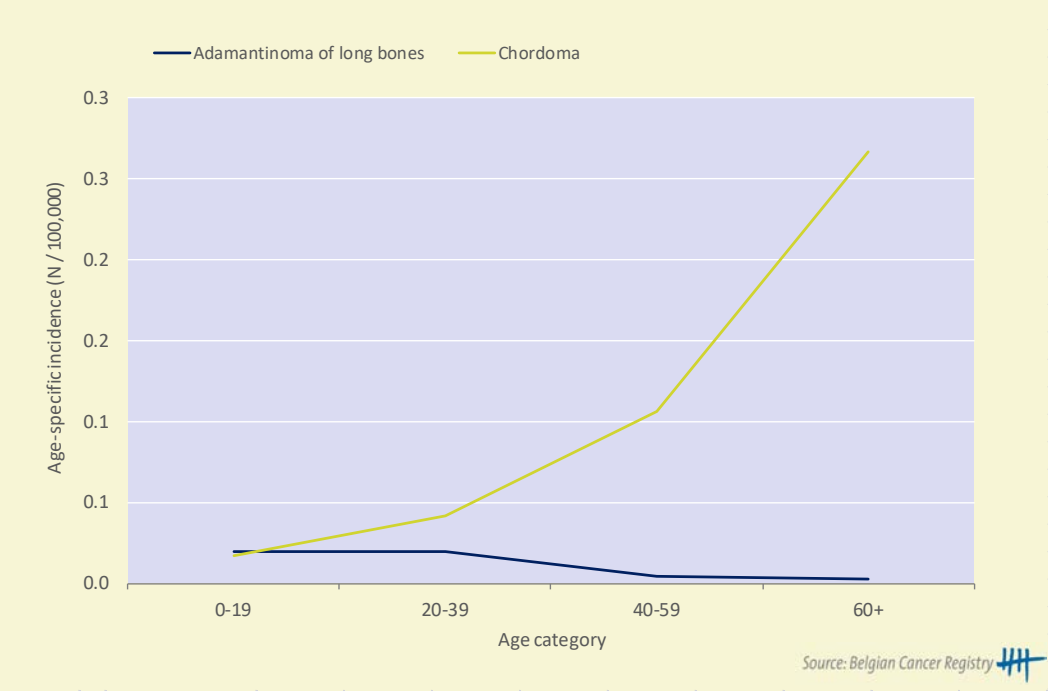
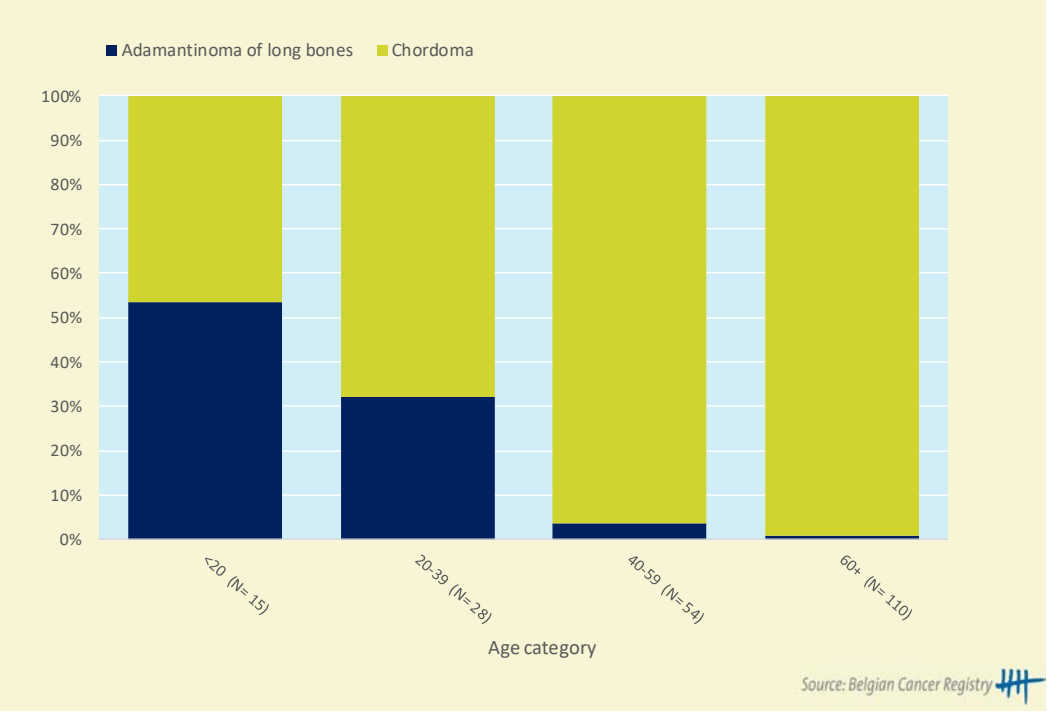
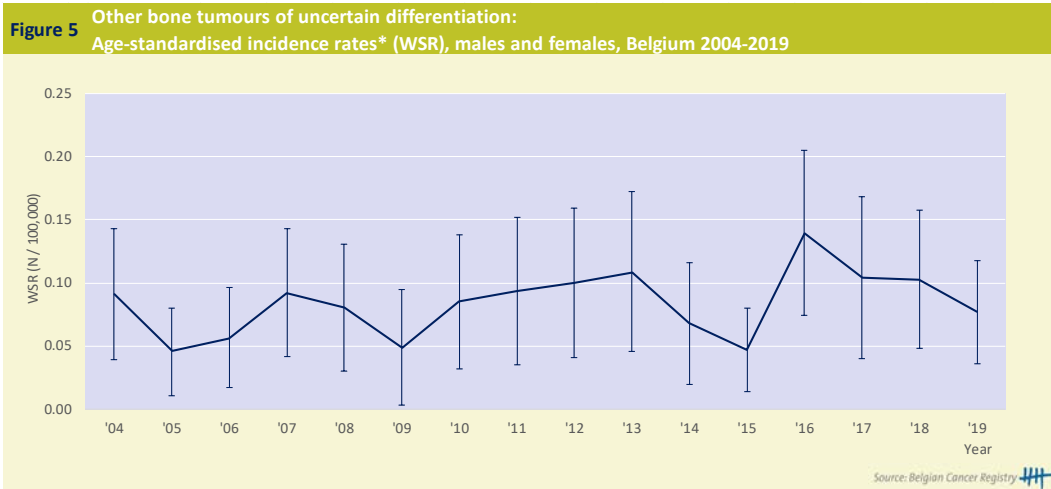


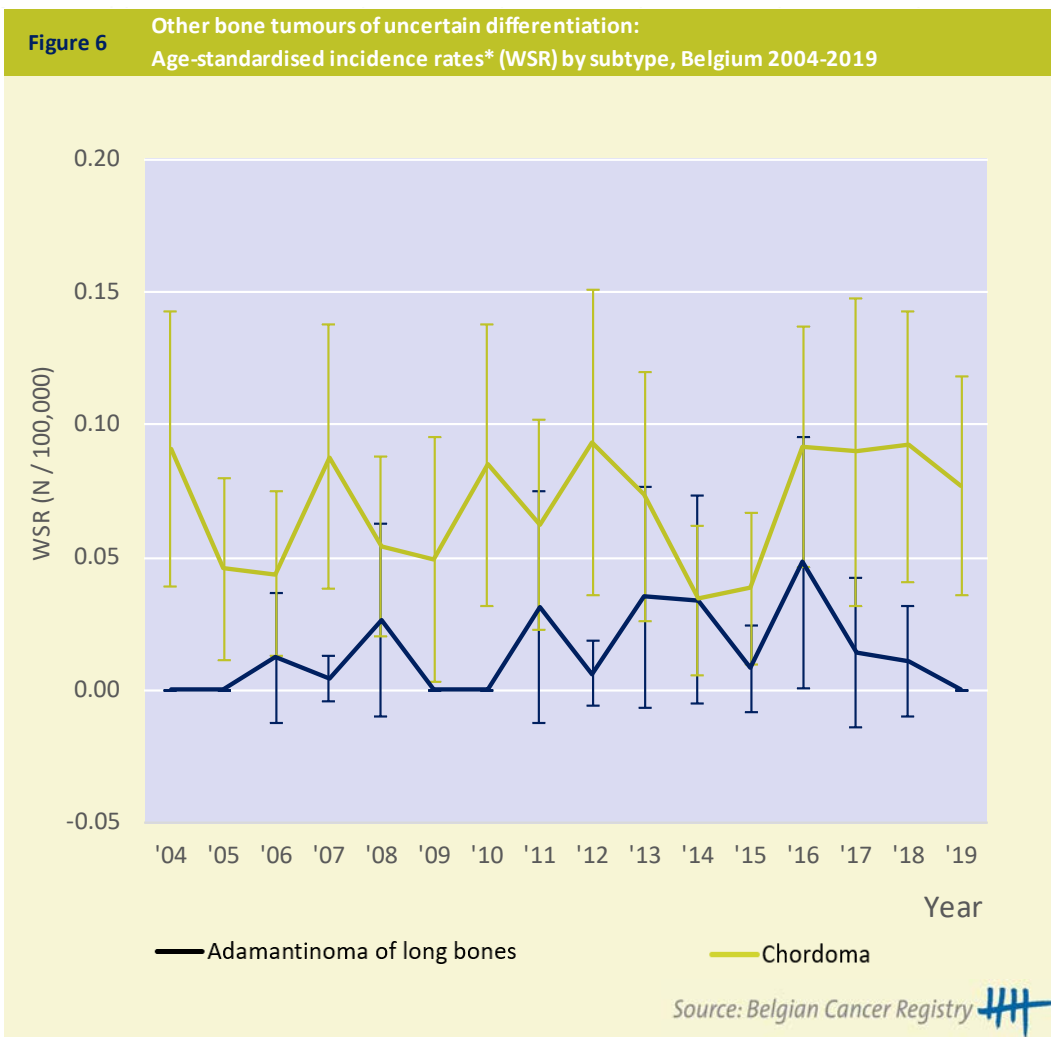
Figure 4 Other bone tumours of uncertain differentiation: Subtype incidence distribution (%) by age category, Belgium 2004-2019



Incidence trends



* The age-standardised incidence rates are represented with 95% Confidence Intervals.



* The age-standardised incidence rates are represented with 95% Confidence Intervals.

Table 2 Other bone tumours of uncertain differentiation:
Incidence trends by sex in Belgium, 2004-2019

Age category	Males			Females		
	AAPC (%)	95%CI	Period	AAPC (%)	95%CI	Period
All ages	3.0	[-1.2; 7.4]	2004-2019	1.5	[-7.4; 11.3]	2004-2019

Source: Belgian Cancer Registry

AAPC: average annual percentage change

Period: when a joinpoint occurred, APC's are calculated for the period before and after the joinpoint. This column represents the corresponding time interval. AAPC's are always calculated over the entire study-period.

Incidence projections

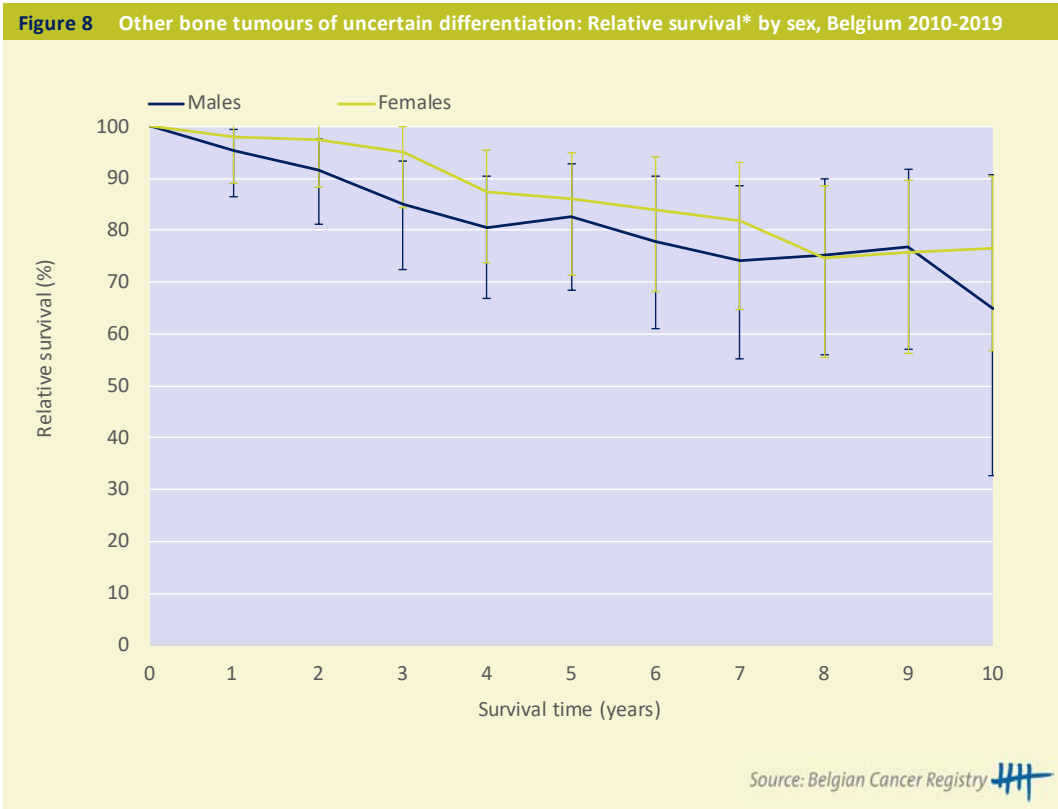
Figure 7 Other bone tumours of uncertain differentiation:
Observed and projected* incidence (WSR), Belgium 2004-2030



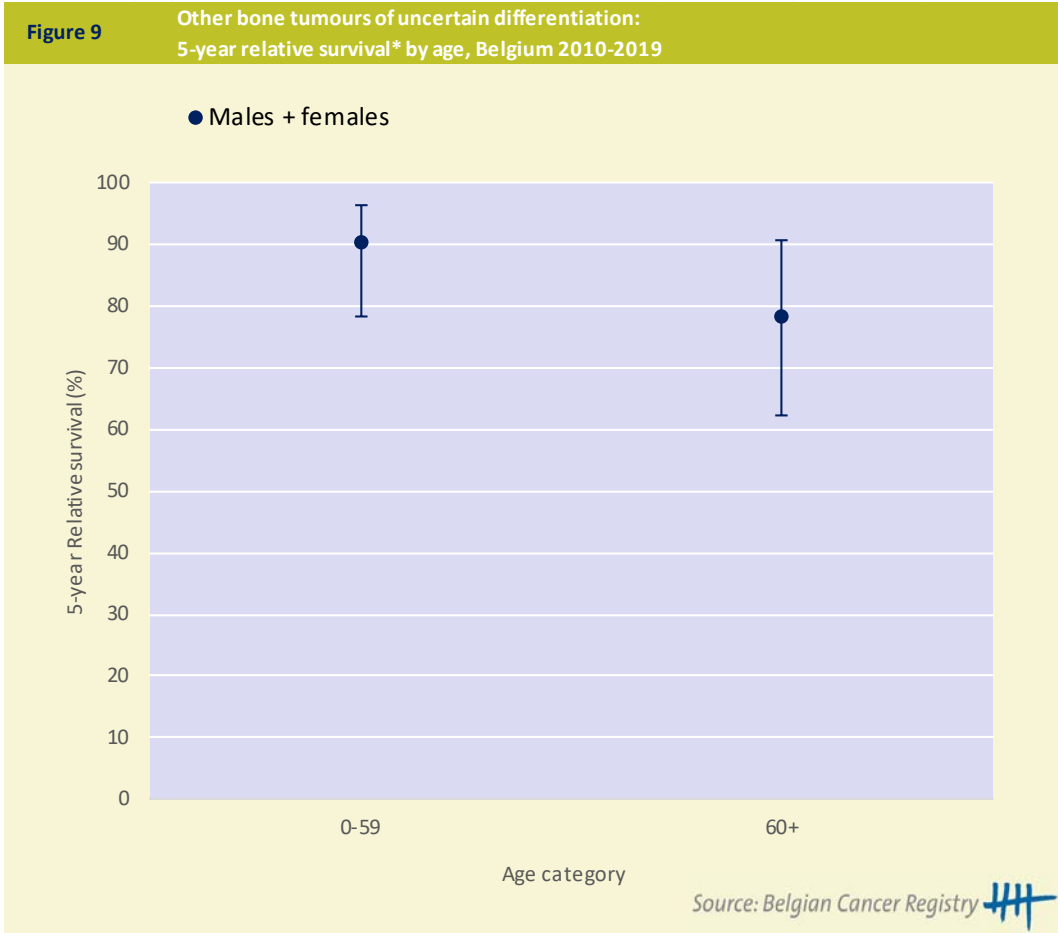
Source: Belgian Cancer Registry

WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

Survival



* The relative survival values are represented with 95% Confidence Intervals



* The relative survival values are represented with 95% Confidence Intervals

Other bone tumours of uncertain differentiation:

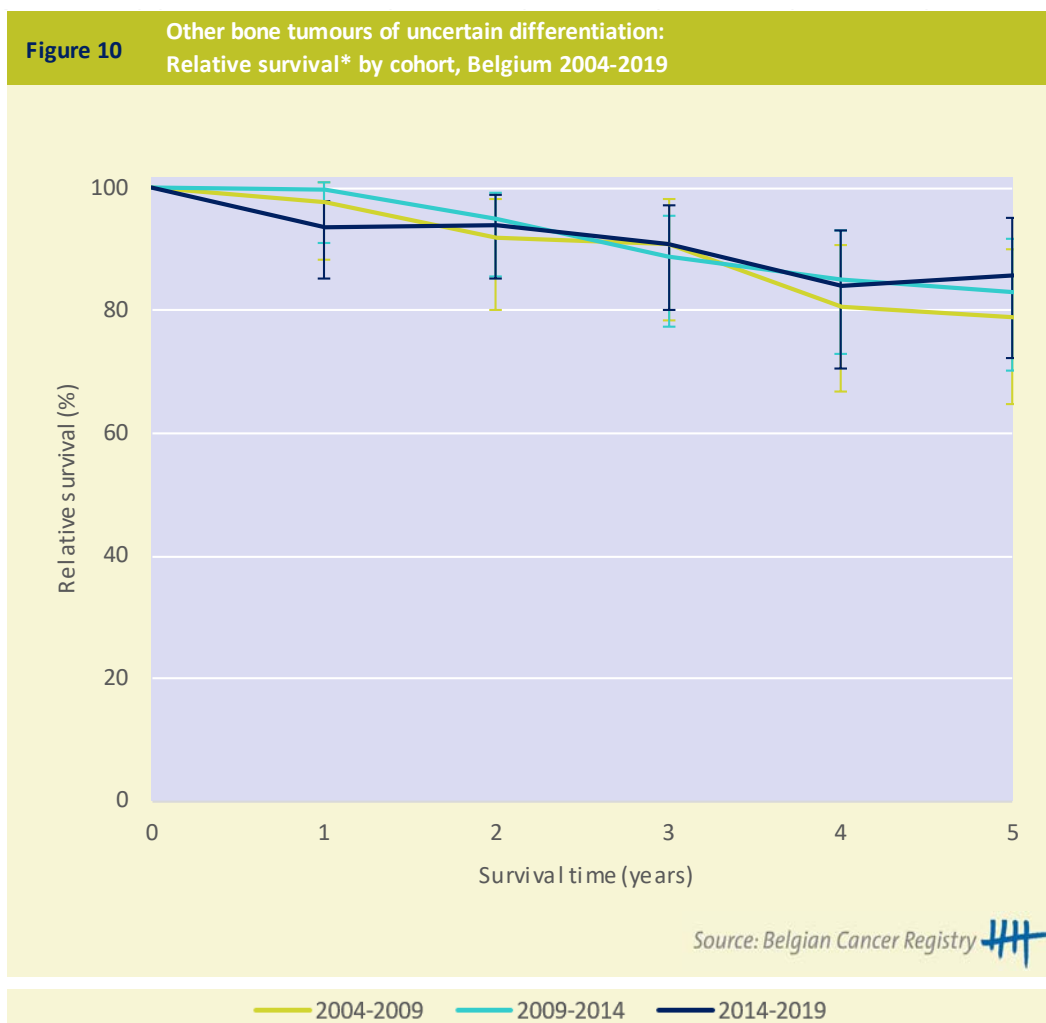
Table 3 Conditional 5-year relative survival*, Belgium, 2010-2019

X years since diagnosis	N at risk	%
1 year	132	83.8
2 year	117	82.7
3 year	95	82.6

* Unadjusted 5-yr relative survival probability conditional on surviving the first X years since diagnosis, %

* Interpretation in lay-man's terms: Given that a patient has already survived X years, what is the relative survival probability 5 years later.

Survival trends



* The relative survival values are represented with 95% Confidence Intervals

3.2.13.1 CHORDOMA

KEYNOTES

Incidence (table 1-2; figure 1-4)

- Chordoma is mainly diagnosed in adults, the incidence increases with age starting around the age of 40 years, reaching a peak at the age of 70-79 years.
- Chordoma occurs in bones, especially the bones of skull and face (36%), the pelvic bones, sacrum & coccyx (32%), and in the vertebral column (27%).

Survival (table 3; figure 5-7)

- The 5-year relative survival does not show a consistent difference between male and female patients with chordoma (81-83%).
- The 5-year relative survival does not show a consistent improvement over time (2004-2019).

Table 1 Chordoma: Overview of incidence, prevalence and survival by sex in Belgium, 2010-2019

Males				
Incidence	N	CR	WSR	
Incidence, 2010-2019	70	0.1	0.1	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	35	0.6	0.3	
10-year prevalence, 31.12.2019	49	0.9	0.5	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	68	80.7	[65.2;92.0]	
10-year relative survival, 2010-2019	68	62.3	[31.2;88.7]	
Females				
Incidence	N	CR	WSR	
Incidence, 2010-2019	55	0.1	0.1	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	25	0.4	0.3	
10-year prevalence, 31.12.2019	42	0.7	0.5	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	55	82.7	[65.5;93.5]	
10-year relative survival, 2010-2019	55	70.6	[47.9;87.7]	
Median age at diagnosis, 2010-2019 (y)	64 [Q1: 50; Q3: 72]			
M/F-ratio	1.2			

Source: Belgian Cancer Registry 

N: number of new diagnoses

CR: crude (all ages) rate (N/100,000 person years)

WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

M/F-ratio: Male/Female ratio based on the age-standardised rates (2010-2019)

Incidence

Figure 1 Chordoma: Age-specific incidence rates (N/100,000) by sex, Belgium 2004-2019

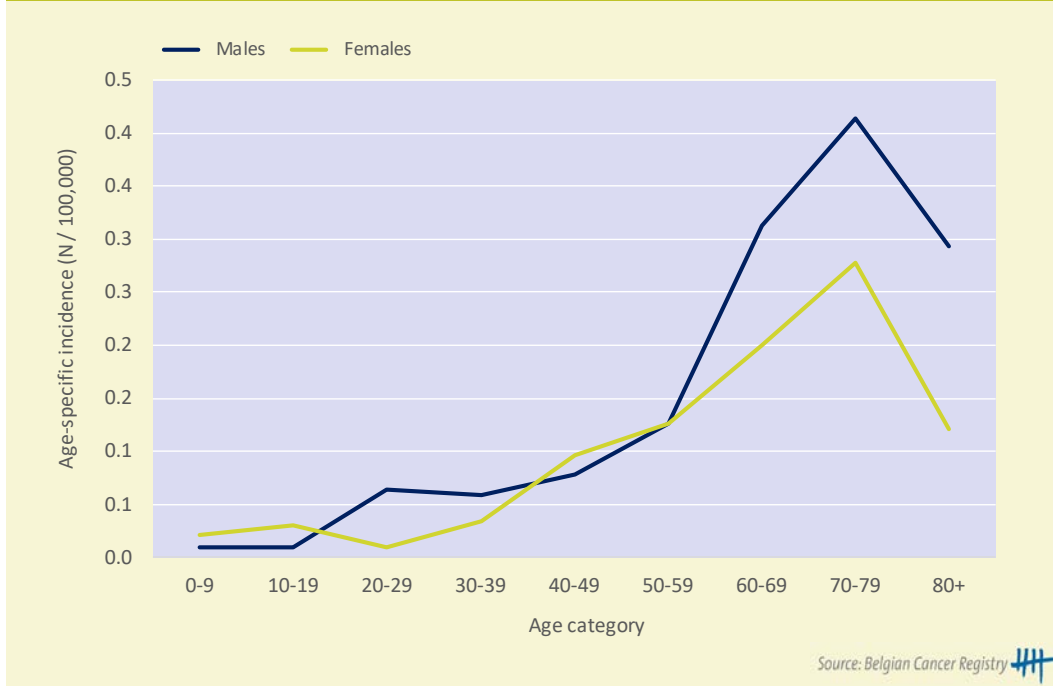
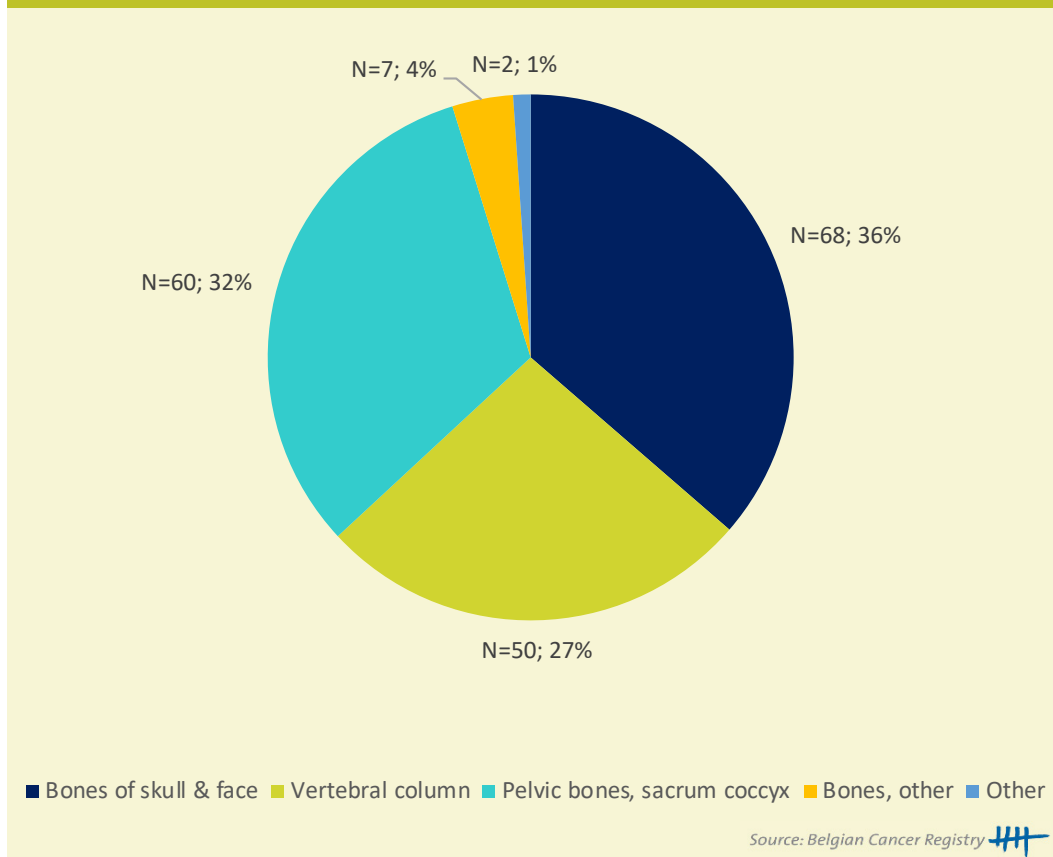
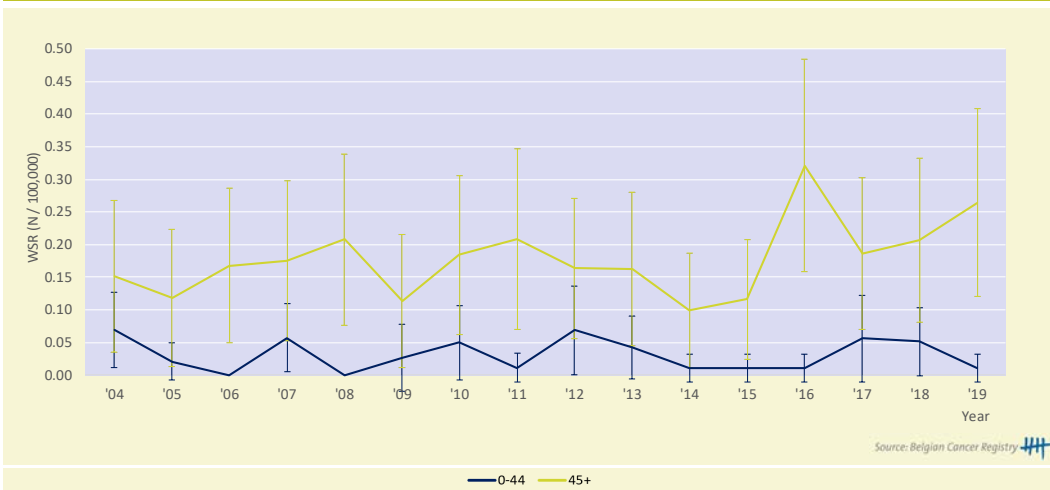


Figure 2 Chordoma: Incidence distribution by primary tumour location, Belgium 2004-2019



Incidence trends

Figure 3 Chordoma: Age-standardised incidence rates* (WSR) by age category, males and females, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals

Table 2 Chordoma: Incidence trends by sex in Belgium, 2004-2019

Age category	Males			Females		
	AAPC (%)	95%CI	Period	AAPC (%)	95%CI	Period
All ages	2.5	[-1.6; 6.9]	2004-2019	0.6	[-7.1; 8.8]	2004-2019

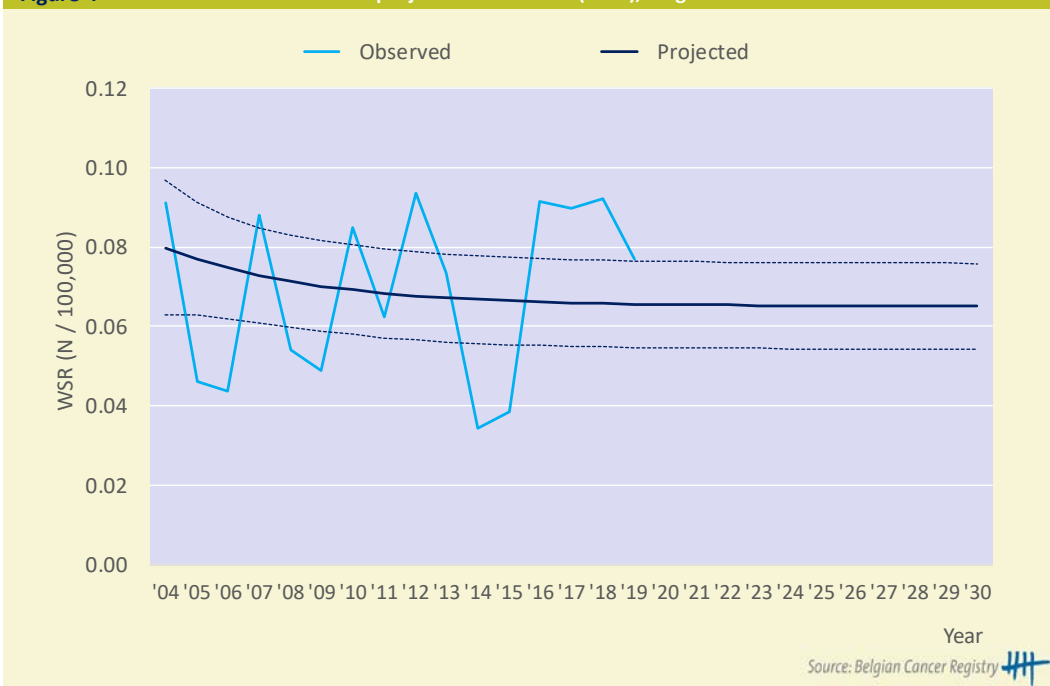
Source: Belgian Cancer Registry

AAPC: average annual percentage change

Period: when a joinpoint occurred, APC's are calculated for the period before and after the joinpoint. This column represents the corresponding time interval. AAPC's are always calculated over the entire study-period.

Incidence projections

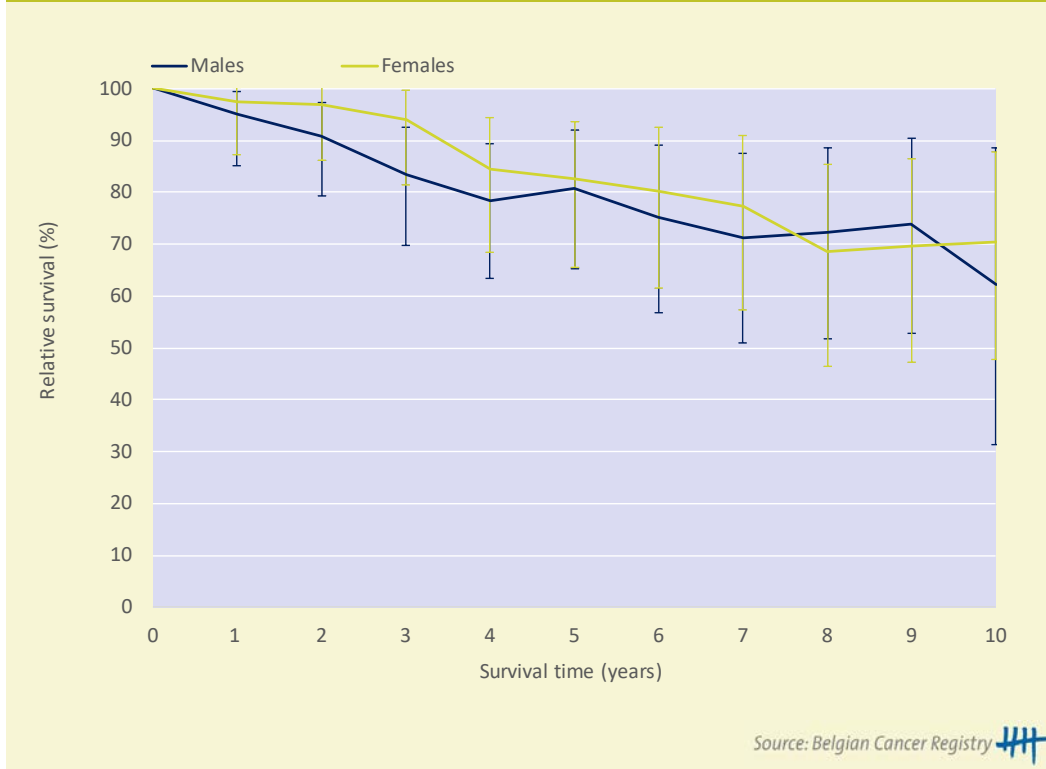
Figure 4 Chordoma: Observed and projected* incidence (WSR), Belgium 2004-2030



WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

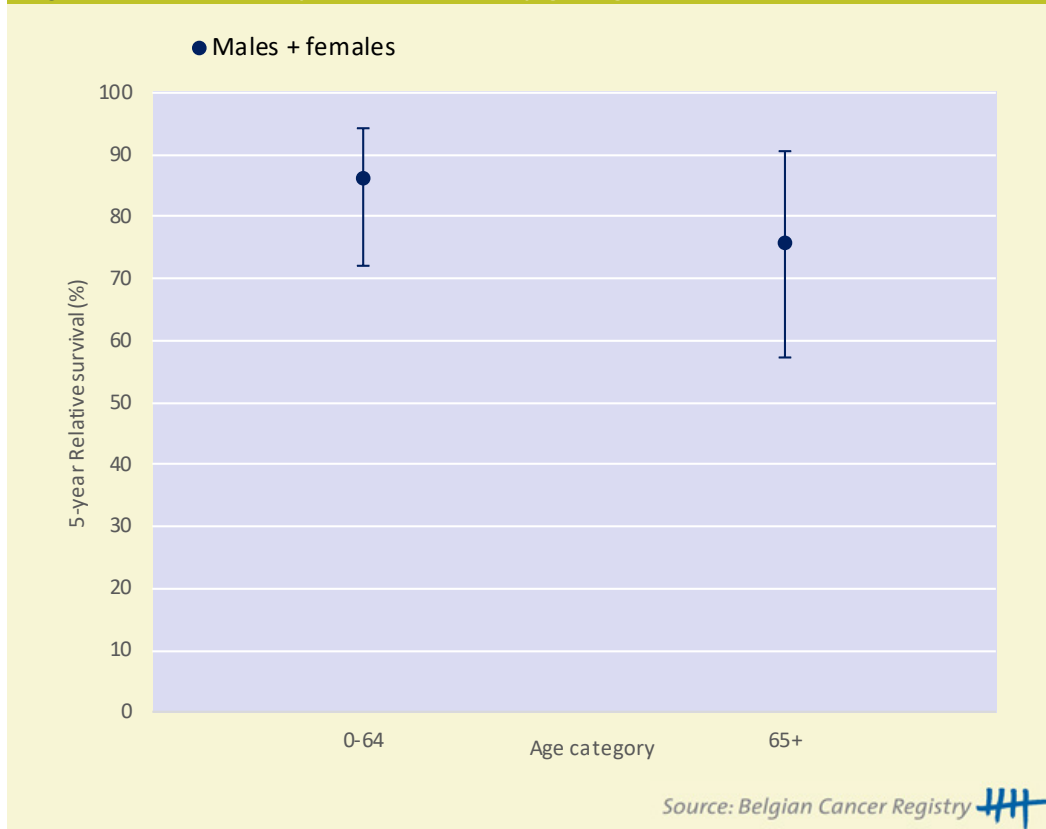
Survival

Figure 5 Chordoma: Relative survival* by sex, Belgium 2010-2019



* The relative survival values are represented with 95% Confidence Intervals

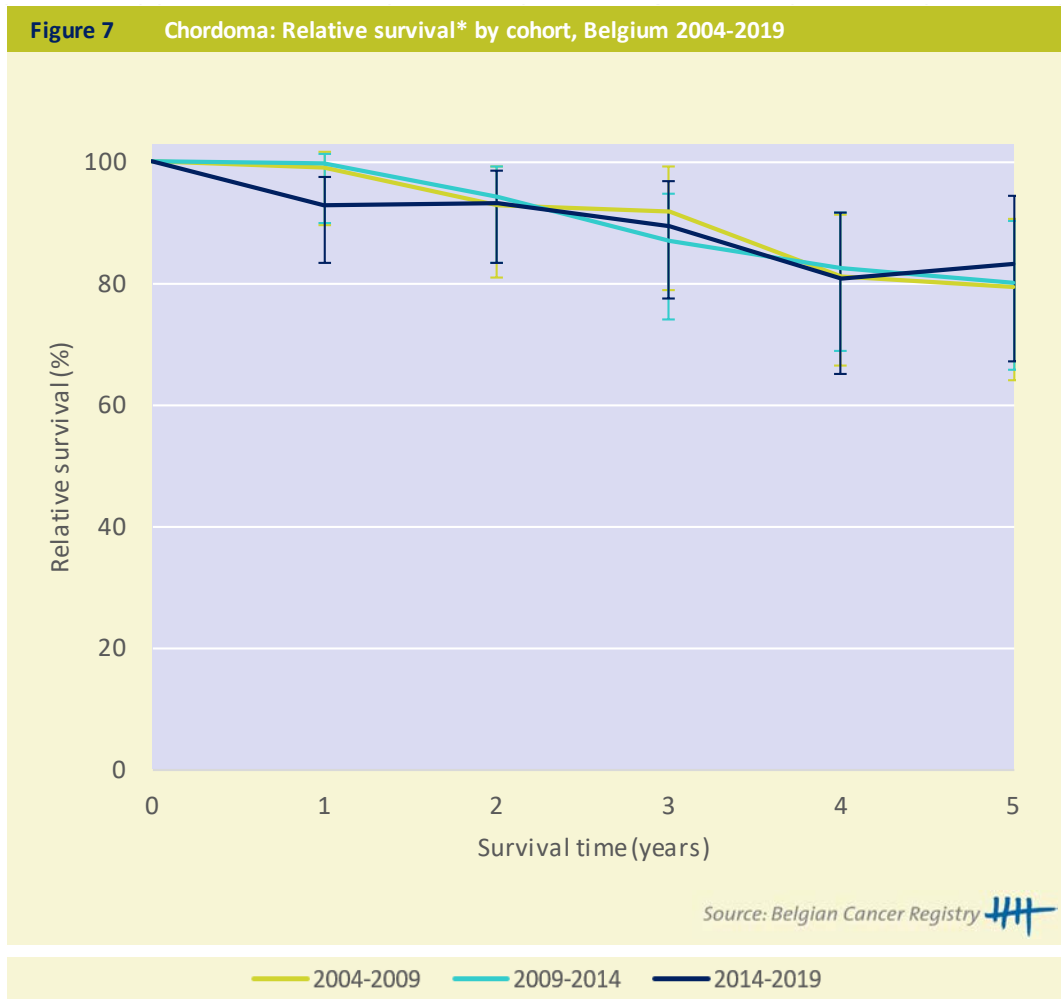
Figure 6 Chordoma: 5-year relative survival* by age, Belgium 2010-2019



* The relative survival values are represented with 95% Confidence Intervals

Chordoma: Conditional 5-year relative survival* in Belgium, 2010-2019		
X years since diagnosis	N at risk	%
1 year	116	80.7
2 year	101	79.3
3 year	80	79.1

* Unadjusted 5-yr relative survival probability conditional on surviving the first X years since diagnosis, %
 * Interpretation in lay-man's terms: Given that a patient has already survived X years, what is the relative survival probability 5 years later.



* The relative survival values are represented with 95% Confidence Intervals

3.2.14 UNCLASSIFIED AND POORLY CHARACTERISED SARCOMA

MAIN SUBTYPES: Entities which are not well specified or not elsewhere classified

- *Small cell sarcoma*
- *Undifferentiated sarcoma*
- *Sarcoma not otherwise specified, stromal sarcoma*
- *Spindle cell sarcoma*
- *Pleomorphic or giant cell sarcoma*

KEYNOTES

Incidence (table 1-2; figure 1-6)

- Unclassified and poorly characterised sarcomas are mostly diagnosed in the older population (60+) and more frequently in males than in females (male/female ratio of 1.5).
- About half (51%) of these unclassified diagnoses concern pleomorphic (or giant cell) sarcoma.

Survival (table 3; figure 7-9)

- The 5-year relative survival of patients with unclassified and poorly characterised sarcoma decreases from 66% under the age of 40 years to 50% in patients older than 80 years. A slight improvement of the survival is observed over time (from 45% in 2004-2009 to 52% in 2014-2019).
- The 10-year relative survival rate is 43% with no major difference between male and female patients.

Table 1 Unclassified and poorly characterised sarcoma: Overview of incidence, prevalence and survival by sex in Belgium, 2010-2019				
Males				
Incidence	N	CR	WSR	
Incidence, 2010-2019	541	1.0	0.5	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	174	3.1	1.5	
10-year prevalence, 31.12.2019	245	4.3	2.1	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	532	52.3	[46.3;58.2]	
10-year relative survival, 2010-2019	532	44.8	[34.0;56.2]	
Females				
Incidence	N	CR	WSR	
Incidence, 2010-2019	409	0.7	0.3	
Prevalence	N	CR	WSR	
5-year prevalence, 31.12.2019	126	2.2	1.1	
10-year prevalence, 31.12.2019	180	3.1	1.6	
Relative survival	N at risk	%	95%CI	
5-year relative survival, 2010-2019	398	46.8	[40.6;52.9]	
10-year relative survival, 2010-2019	398	42.0	[33.4;50.9]	
Median age at diagnosis, 2010-2019 (y)	73 [Q1: 60; Q3: 81]			
M/F-ratio	1.5			

Source: Belgian Cancer Registry 

N: number of new diagnoses

CR: crude (all ages) rate (N/100,000 person years)

WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

M/F-ratio: Male/Female ratio based on the age-standardised rates (2010-2019)

Incidence

Figure 1 Unclassified and poorly characterised sarcoma: Age-specific incidence rates (N/100,000) by sex, Belgium 2004-2019

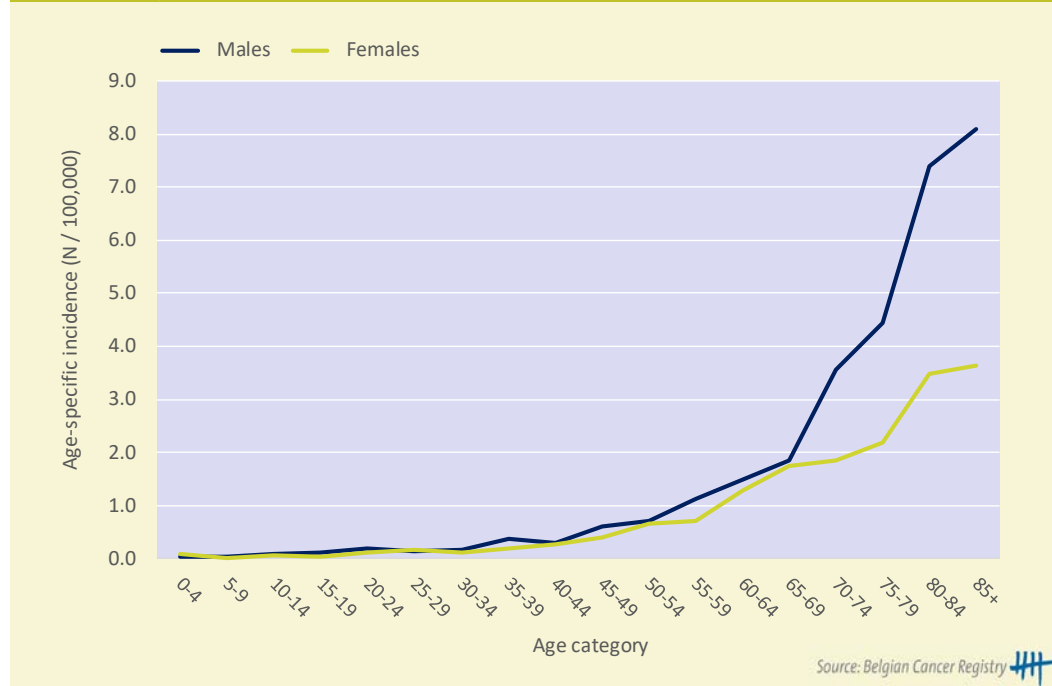


Figure 2 Unclassified and poorly characterised sarcoma: Incidence distribution by subtype, Belgium 2010-2019

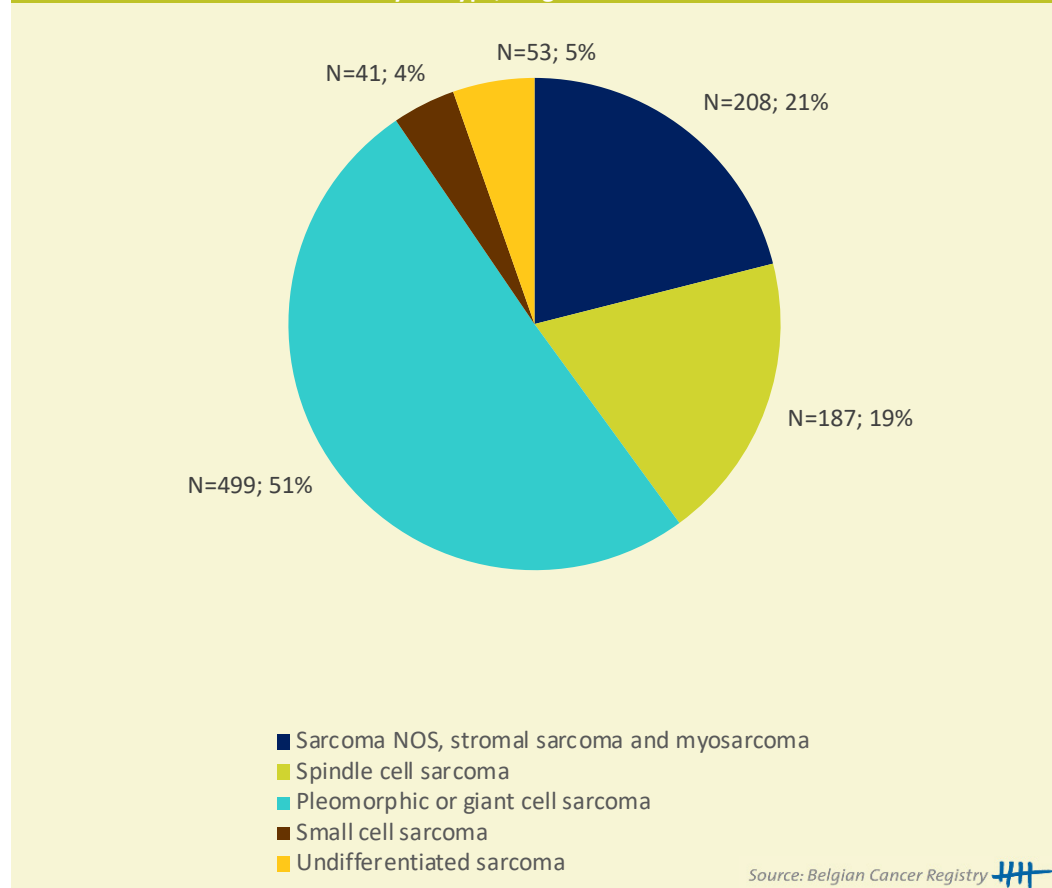
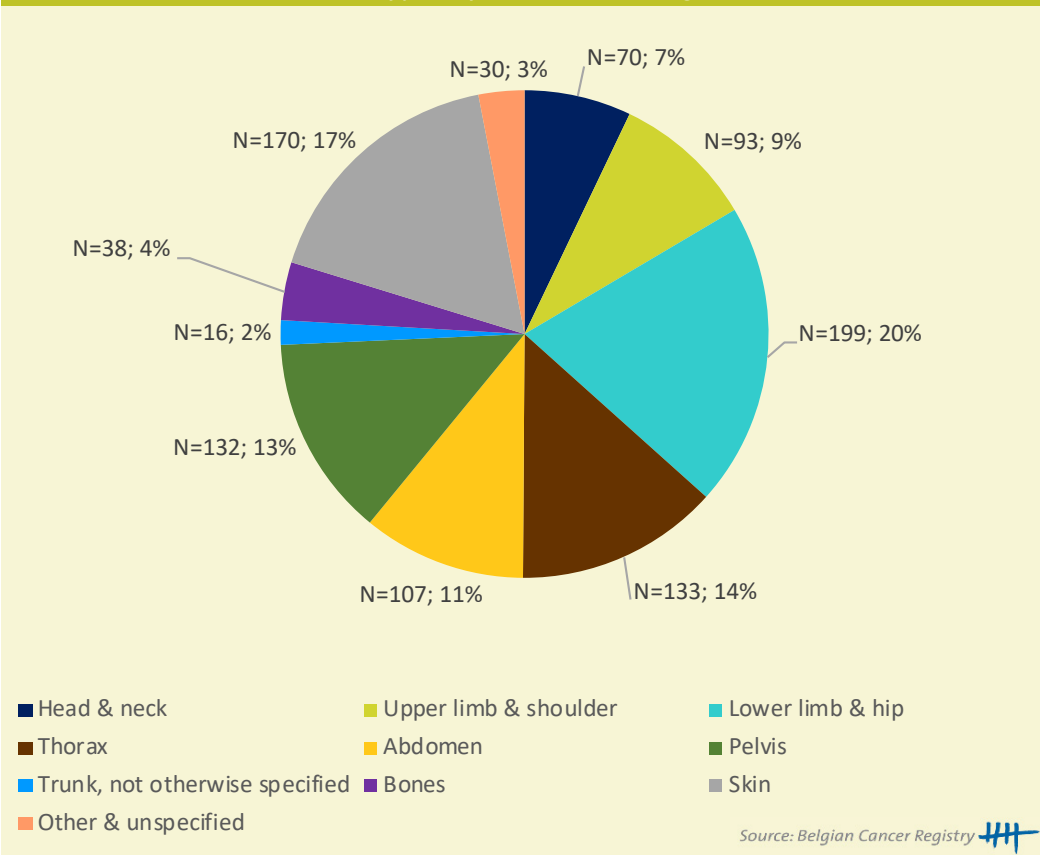


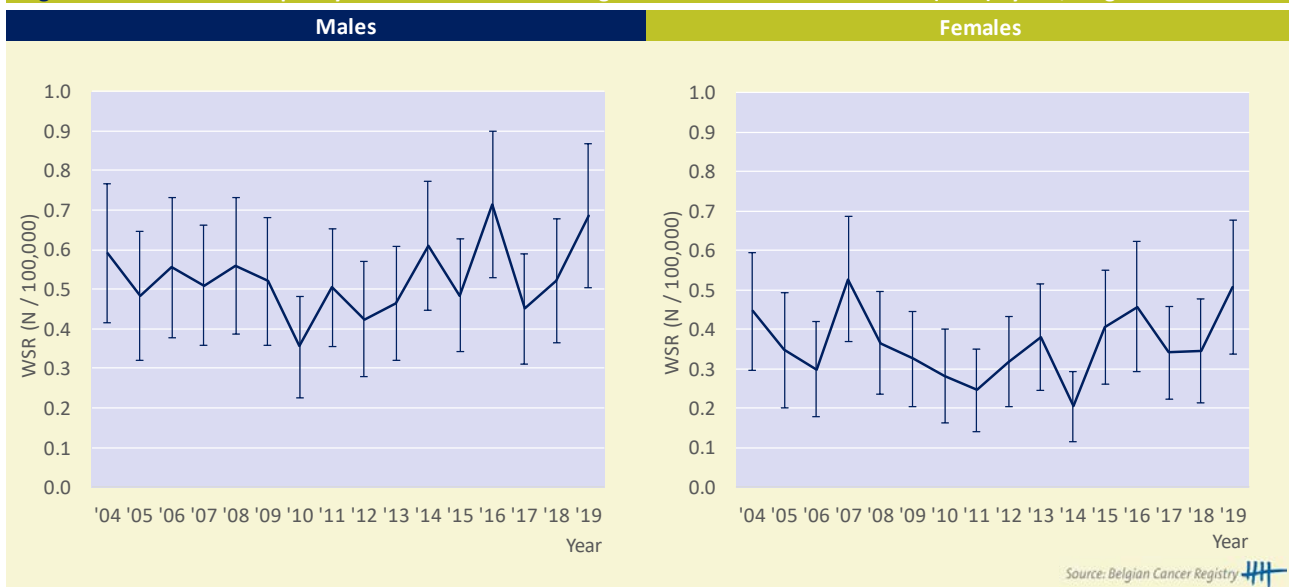
Figure 3

**Unclassified and poorly characterised sarcoma:
Incidence distribution by primary tumour location, Belgium 2010-2019**

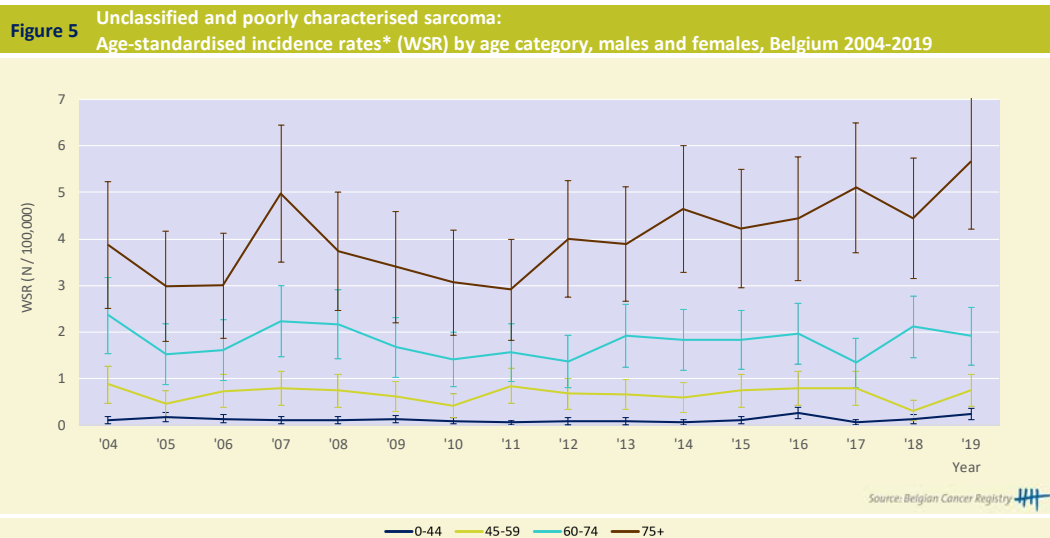


Incidence trends

Figure 4 Unclassified and poorly characterised sarcoma: Age-standardised incidence rates* (WSR) by sex, Belgium 2004-2019



* The age-standardised incidence rates are represented with 95% Confidence Intervals.



* The age-standardised incidence rates are represented with 95% Confidence Intervals.

Table 2 Unclassified and poorly characterised sarcoma:
Incidence trends by sex and age category in Belgium, 2004-2019

Age category	Males			Females		
	AAPC (%)	95%CI	Period	AAPC (%)	95%CI	Period
All ages	0.7	[-1.4; 2.8]	2004-2019	0.2	[-2.7; 3.3]	2004-2019
0 - 44 y	1.0	[-4.3; 6.6]	2004-2019	0.1	[-6.3; 6.9]	2004-2019
	-10.8	[-20.3; -0.2]	2004-2012			
45 - 59 y	16.5	[2.2; 32.7]	2012-2019	-0.2	[-7.2; 7.3]	2004-2019
	-1.4	[-5.3; 2.5]	2004-2019			
60 - 74 y	0.1	[-3.1; 3.4]	2004-2019	-0.7	[-4.5; 3.3]	2004-2019
75+ y	2.7	[0.1; 5.3]	2004-2019	2.5	[-1.2; 6.2]	2004-2019
	-6.1	[-12.6; 0.9]	2004-2010			
	8.9	[4.1; 14.0]	2010-2019			

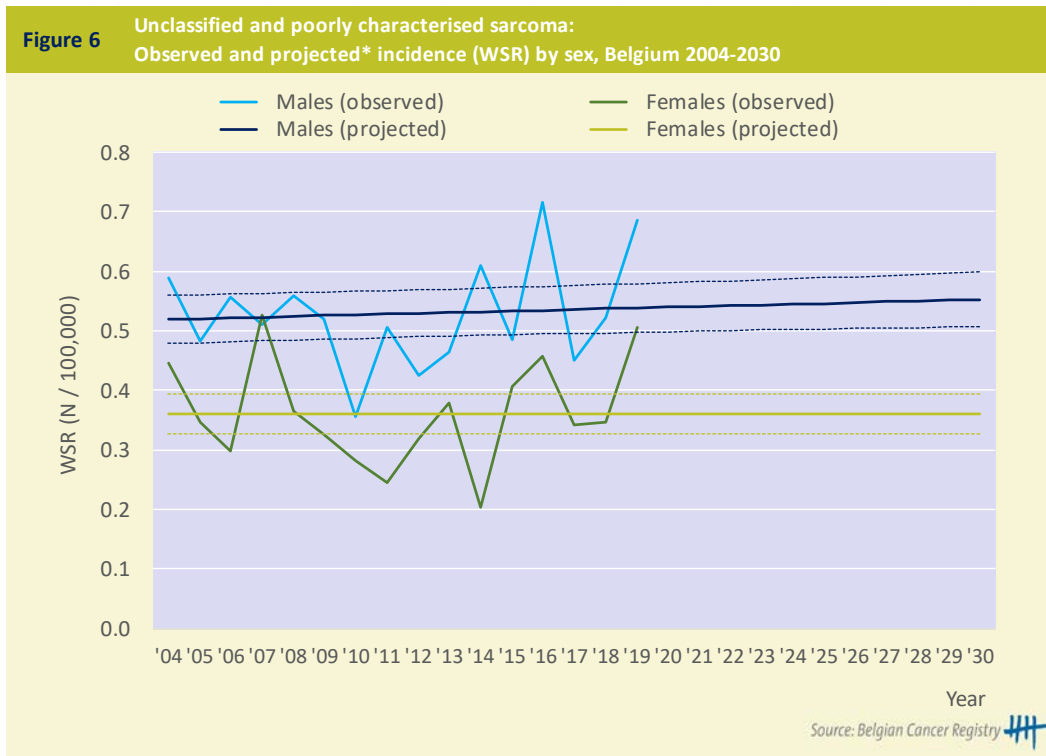
Source: Belgian Cancer Registry

AAPC: average annual percentage change

Period: when a joinpoint occurred, APC's are calculated for the period before and after the joinpoint. This column represents the corresponding time interval. AAPC's are always calculated over the entire study-period.

Incidence projections

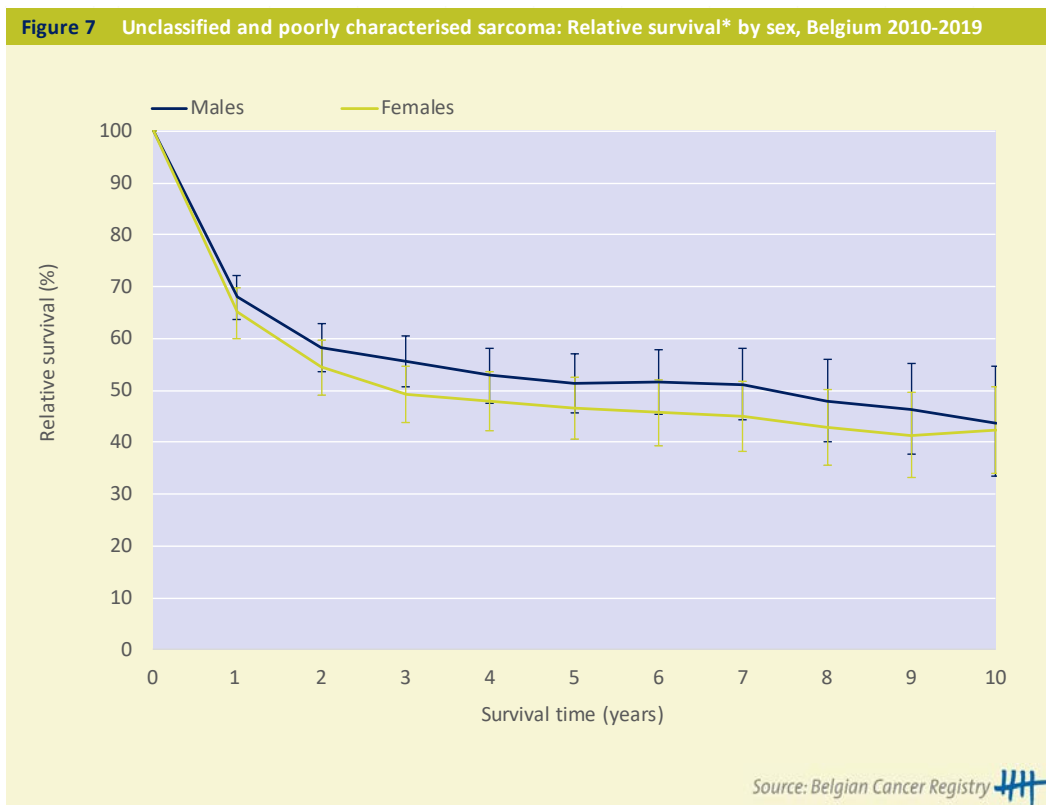
Figure 6 Unclassified and poorly characterised sarcoma: Observed and projected* incidence (WSR) by sex, Belgium 2004-2030



WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

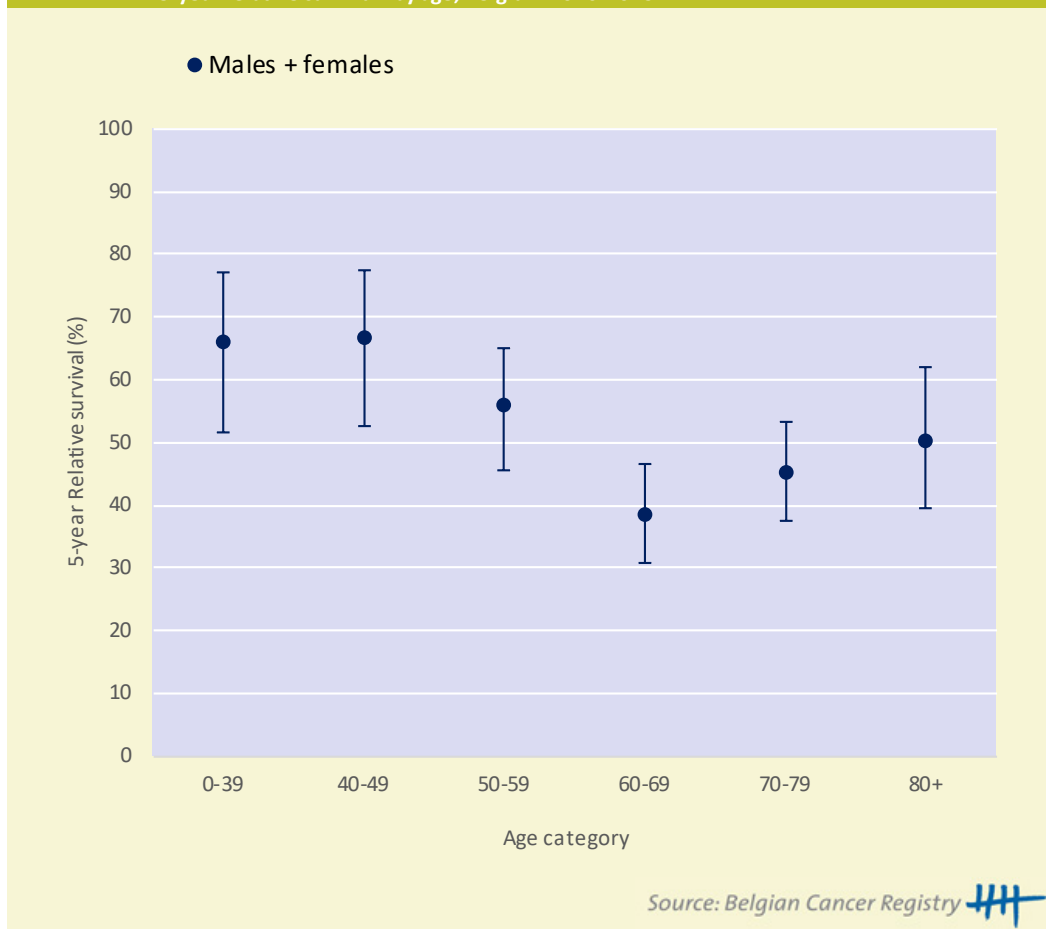
Survival

Figure 7 Unclassified and poorly characterised sarcoma: Relative survival* by sex, Belgium 2010-2019



* The relative survival values are represented with 95% Confidence Intervals

Figure 8 Unclassified and poorly characterised sarcoma:
5-year relative survival* by age, Belgium 2010-2019



* The relative survival values are represented with 95% Confidence Intervals

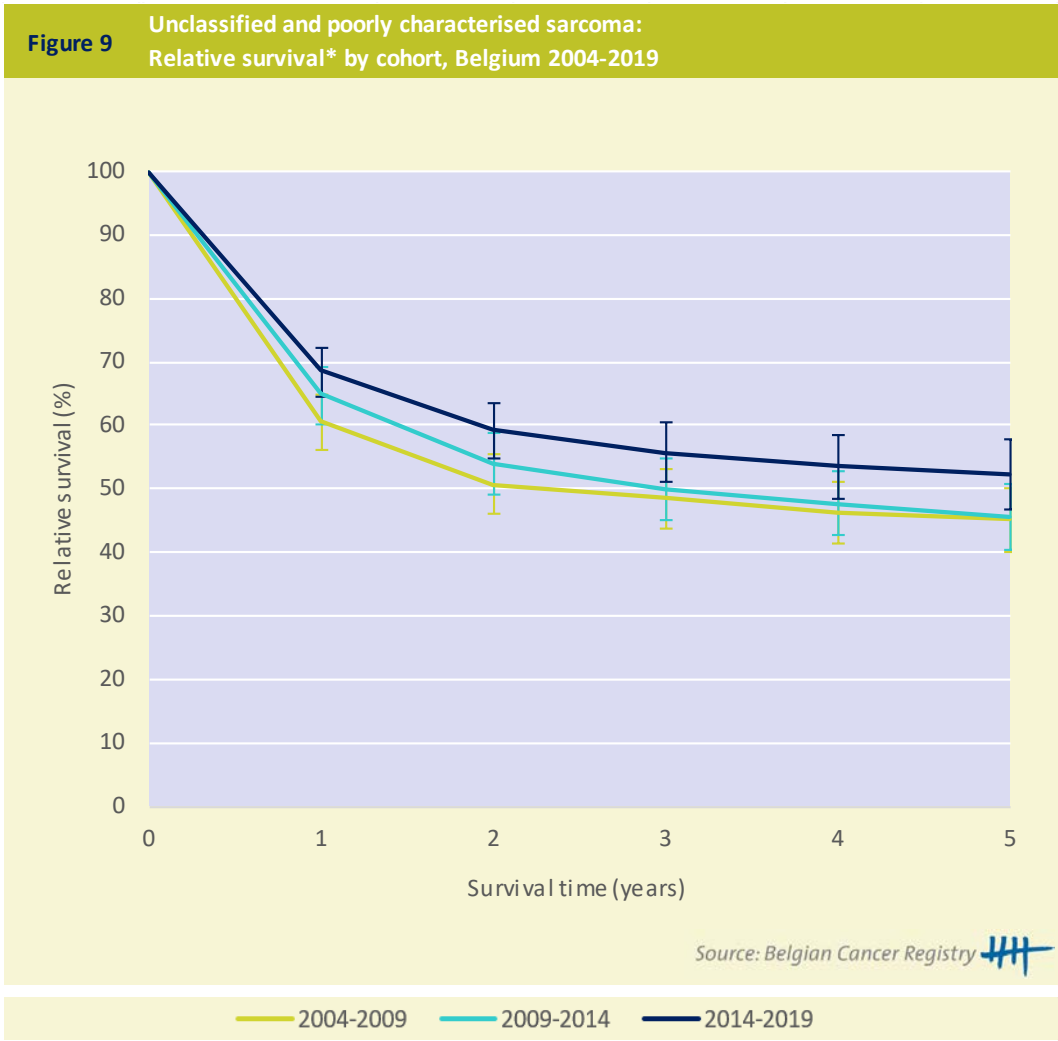
Unclassified and poorly characterised sarcoma:

Table 3 Conditional 5-year relative survival*, Belgium, 2010-2019

X years since diagnosis	N at risk	%
1 year	633	73.3
2 year	450	85.5
3 year	348	86.5

* Unadjusted 5-yr relative survival probability conditional on surviving the first X years since diagnosis, %
* Interpretation in lay-man's terms: Given that a patient has already survived X years, what is the relative survival probability 5 years later.

Survival trends



* The relative survival values are represented with 95% Confidence Intervals

4 INTERNATIONAL COMPARISON

KEYNOTES

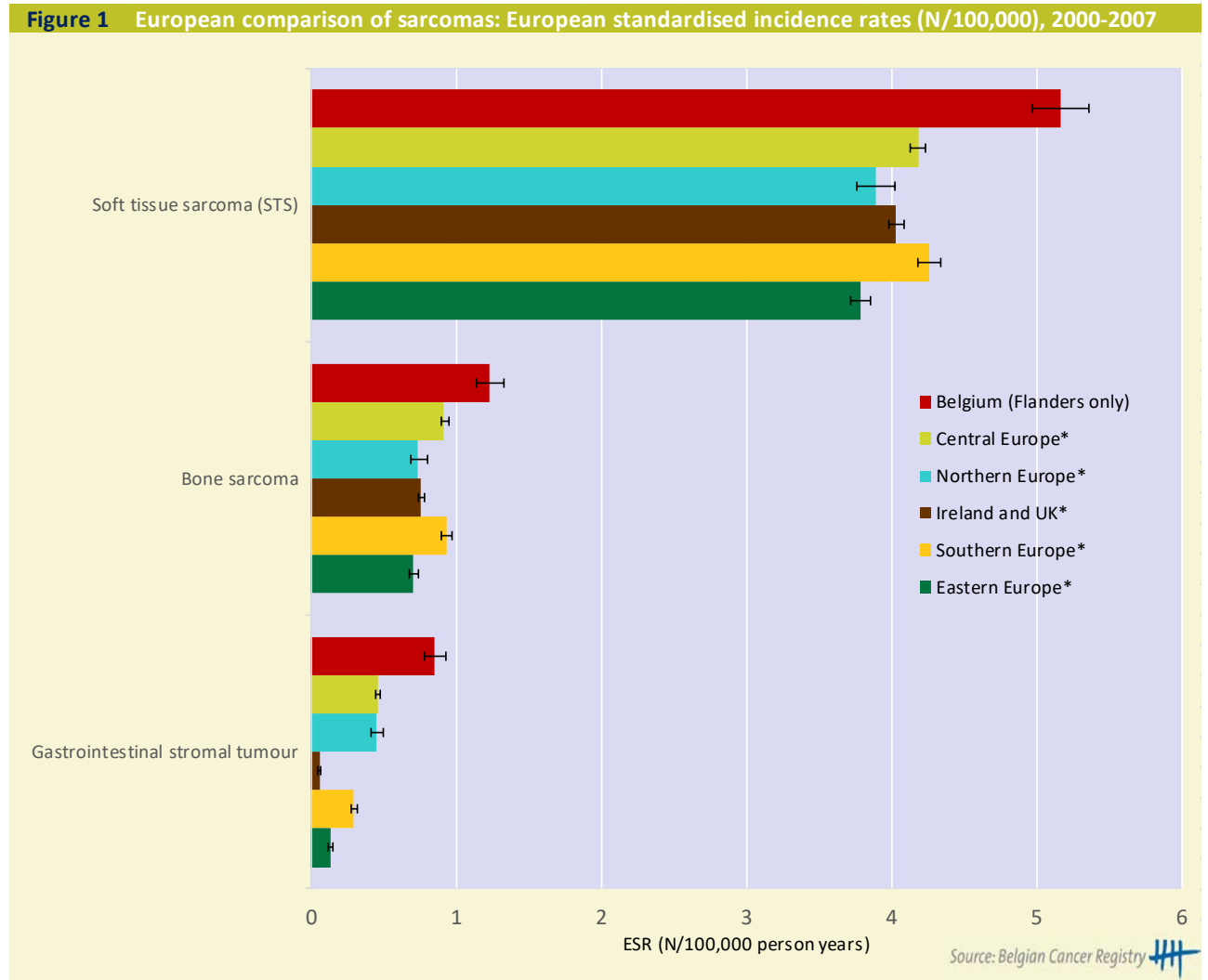
Incidence (figure 1-6; table 1-2)

- Overall sarcoma incidence is consistent between Belgium and the different European regions, and more precisely, the neighbouring countries, France and the Netherlands.
- The main discrepancy is observed for gastrointestinal stromal tumours (GIST) which are much more frequent in Belgium than in the various European regions, and specially in France and the Netherlands. This can be explained by different inclusion criteria : all GIST, whatever the grade, the behaviour and the size (including incidental micro-GIST), are registered in Belgium (according to TNM Classification, 8th edition), while other countries, such as France, only register high-grade GIST and exclude micro-GIST.
- The excess incidence of bone tumours in Belgium compared to the various European regions can be relativized by the fact that the main subtypes of bone tumours are in the same order of magnitude as the neighbouring countries, France and the Netherlands, or even lower for chondrosarcomas when comparing to the Netherlands.
- A potential explanation for the different incidence of chondrosarcomas between Belgium, France and the Netherlands could be differences in the exhaustivity of registration in relation to the changes in the classification of low-grade chondrosarcoma and atypical cartilaginous tumours.

Survival (figure 7-10; table 3-5)

- Overall, the 5-year relative survival of patients diagnosed with sarcoma in Belgium is in the same range (or even slightly better) than in the United States of America (USA), different parts of Europe, and two neighbouring countries, France and the Netherlands.
- As expected, the only major difference is the higher 5-year relative survival of GIST, especially during the period 2000-2007 (Flanders compared to European regions).

Incidence



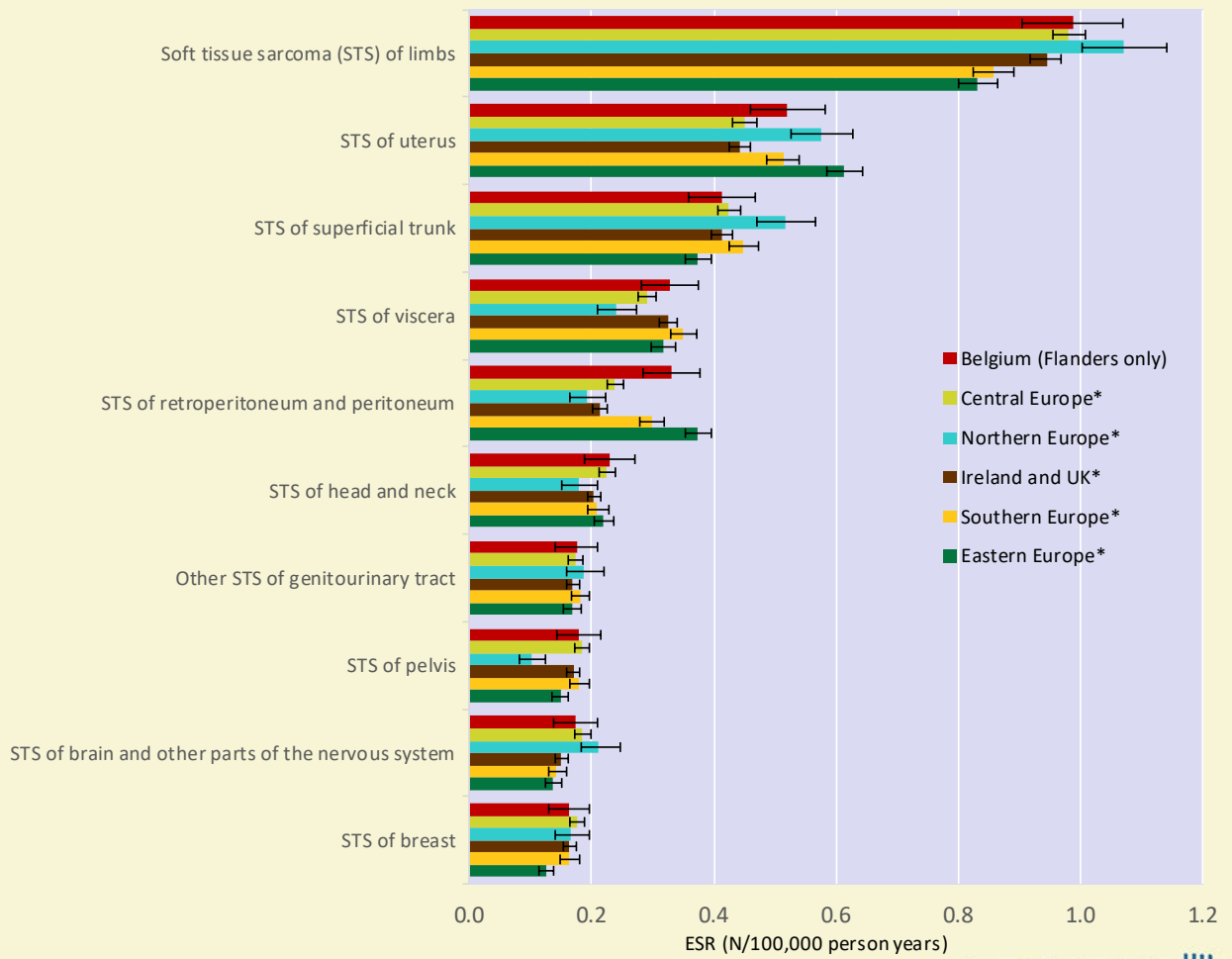
ESR: age-standardised rate using the 1976 European Standard Population (N/100,000 person years)

The age-specific incidence rates are represented with 95% Confidence Intervals

* Data from RARECARENet - Online Analysis (istitutotumori.mi.it)⁽²⁹⁾

Figure 2

European comparison of ten most frequent soft tissue sarcomas: European standardised incidence rates (N/100,000), 2000-2007



Source: Belgian Cancer Registry

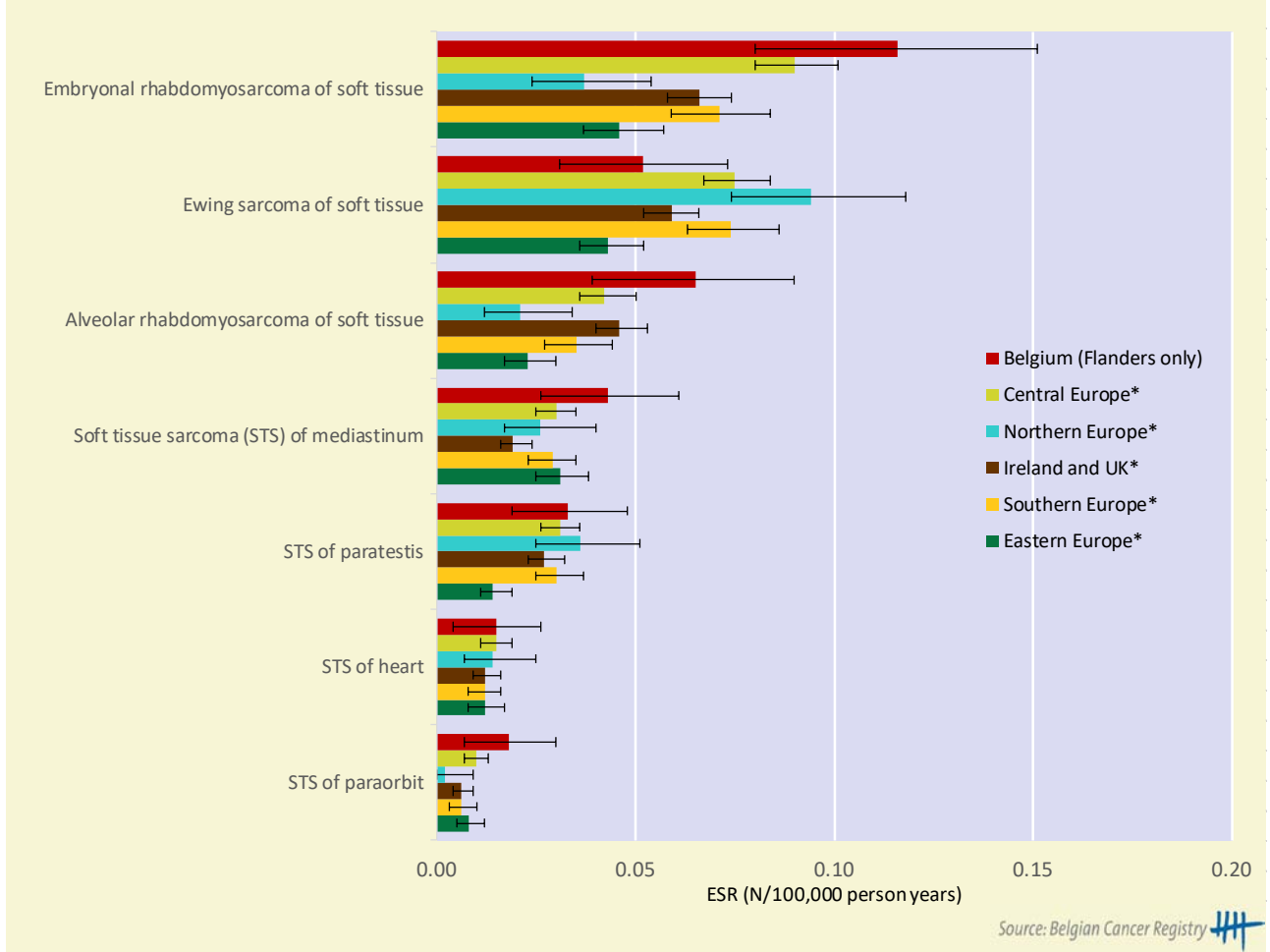
ESR: age-standardised rate using the 1976 European Standard Population (N/100,000 person years)

The age-specific incidence rates are represented with 95% Confidence Intervals

* Data from RARECARENet - On line Analysis (istitutotumori.mi.it)⁽²⁹⁾

Figure 3

European comparison of less frequent soft tissue sarcomas: European standardised incidence rates (N/100,000), 2000-2007



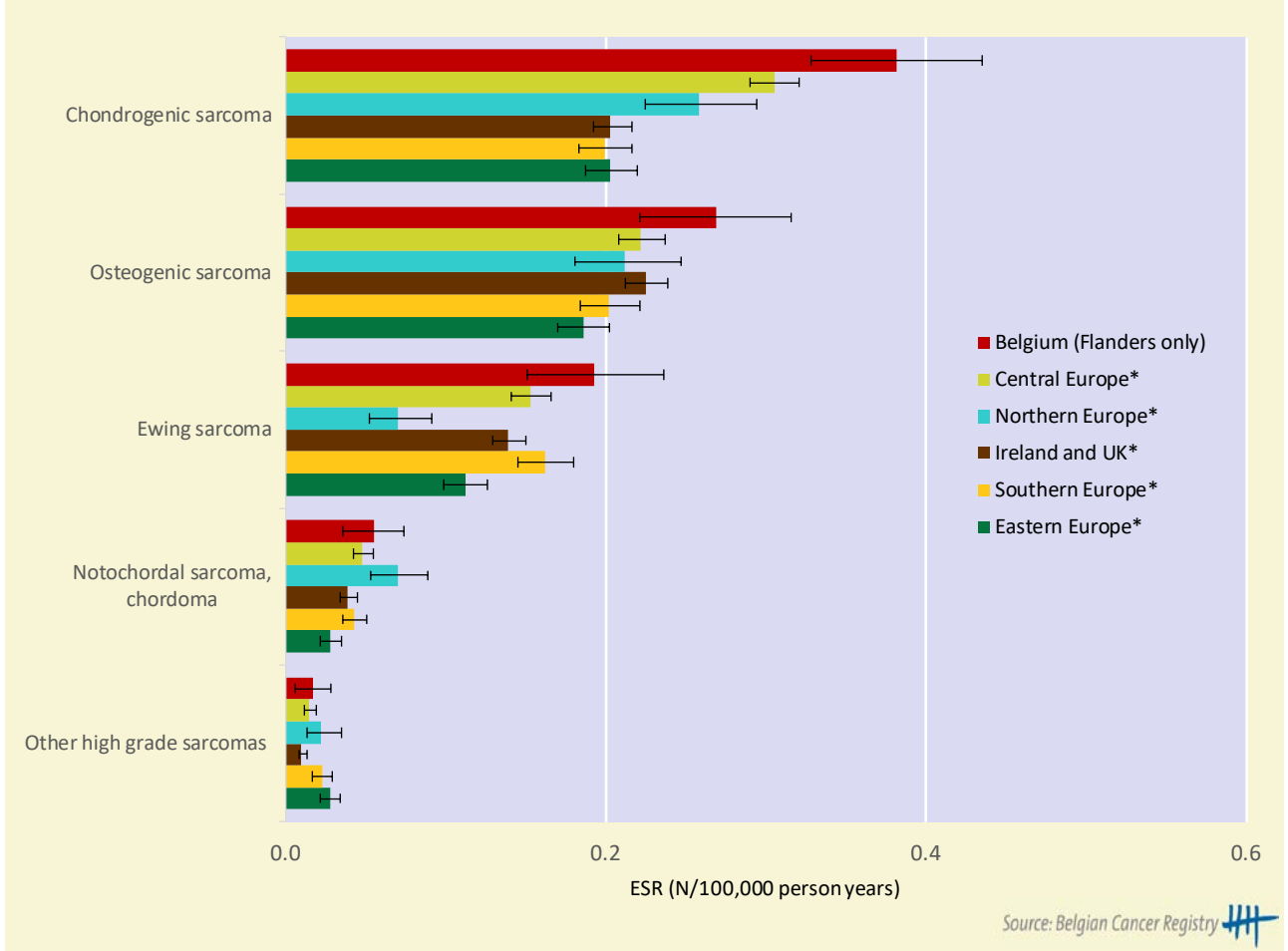
ESR: age-standardised rate using the 1976 European Standard Population (N/100,000 person years)

The age-specific incidence rates are represented with 95% Confidence Intervals

* Data from RARECARENet - On line Analysis (istitutotumori.mi.it)⁽²⁹⁾

Figure 4

European comparison of bone sarcoma: European standardised incidence rates (N/100,000), 2000-2007



ESR: age-standardised rate using the 1976 European Standard Population (N/100,000 person years)

The age-specific incidence rates are represented with 95% Confidence Intervals

* Data from RARECARENet - On line Analysis (istitutotumori.mi.it)⁽²⁹⁾

Source: Belgian Cancer Registry

Table 1 European comparison of sarcoma European Standardised incidence Rates (N/100,000), 2000-2007

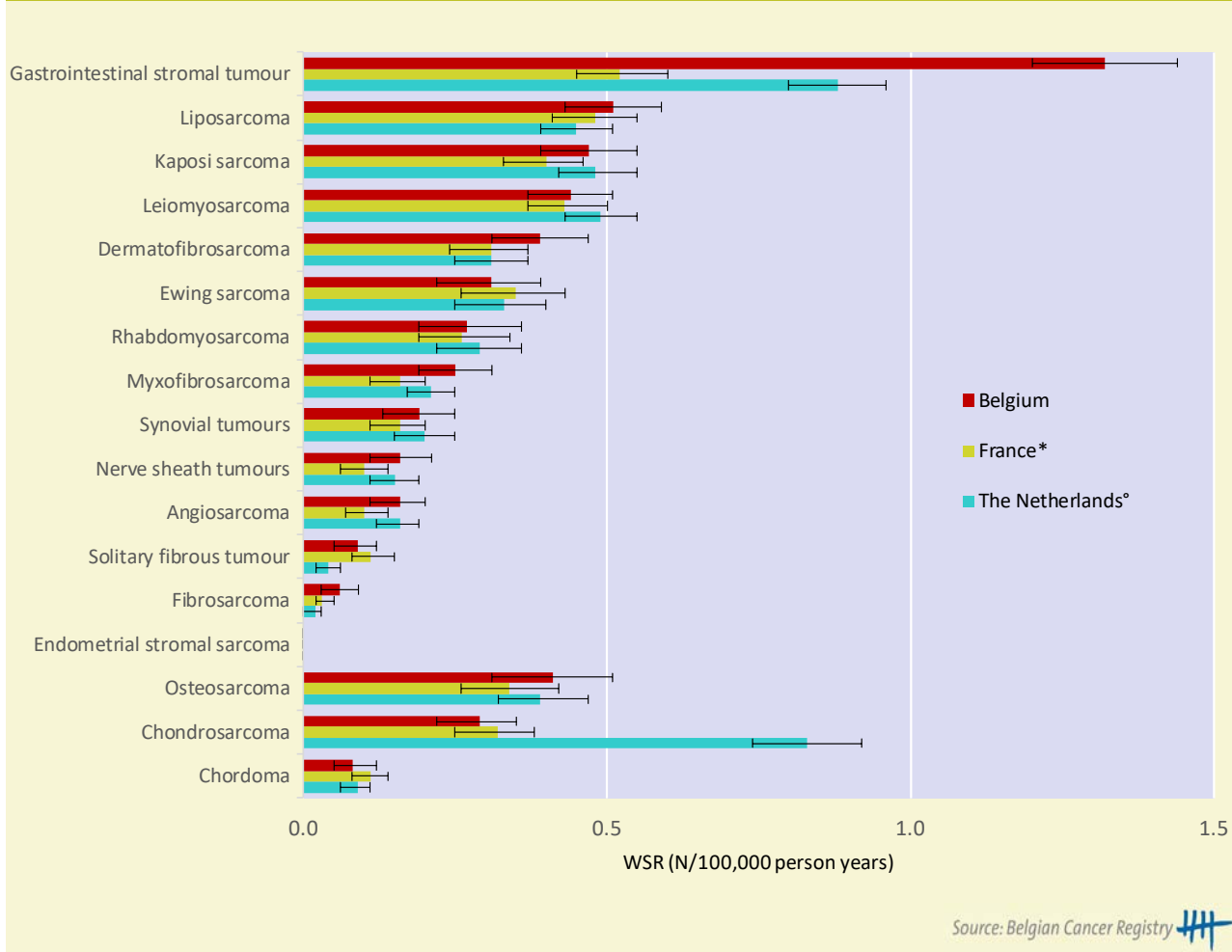
	Belgium (Flanders)		Central Europe*		Northern Europe*		Ireland and UK*		Southern Europe*		Northern Europe*	
	ESR [95% CI]		ESR [95% CI]		ESR [95% CI]		ESR [95% CI]		ESR [95% CI]		ESR [95% CI]	
Soft tissue sarcoma (STS)	5.17 [4.97;5.36]	4.18 [4.12;4.24]	3.89 [3.76;4.02]	4.03 [3.98;4.09]	4.26 [4.18;4.33]	3.78 [3.71;3.85]						
STS of limbs	0.99 [0.90;1.07]	0.98 [0.95;1.01]	1.07 [1.00;1.14]	0.94 [0.92;0.97]	0.86 [0.82;0.89]	0.83 [0.80;0.86]						
STS of uterus	0.52 [0.46;0.58]	0.45 [0.43;0.47]	0.57 [0.53;0.63]	0.44 [0.42;0.46]	0.51 [0.49;0.54]	0.61 [0.59;0.64]						
STS of superficial trunk	0.41 [0.36;0.47]	0.42 [0.41;0.44]	0.52 [0.47;0.56]	0.41 [0.40;0.43]	0.45 [0.42;0.47]	0.37 [0.35;0.40]						
Gastrointestinal stromal tumour	0.85 [0.78;0.93]	0.46 [0.44;0.48]	0.45 [0.41;0.50]	0.05 [0.05;0.06]	0.29 [0.27;0.31]	0.13 [0.12;0.15]						
STS of viscera	0.33 [0.28;0.37]	0.29 [0.28;0.31]	0.24 [0.21;0.27]	0.33 [0.31;0.34]	0.35 [0.33;0.37]	0.32 [0.30;0.34]						
STS of retroperitoneum and peritoneum	0.33 [0.28;0.38]	0.24 [0.22;0.25]	0.19 [0.16;0.22]	0.21 [0.20;0.23]	0.30 [0.28;0.32]	0.37 [0.35;0.40]						
STS of head and neck	0.23 [0.19;0.27]	0.22 [0.21;0.24]	0.18 [0.15;0.21]	0.20 [0.19;0.22]	0.21 [0.19;0.23]	0.22 [0.20;0.24]						
Other STS of genitourinary tract	0.18 [0.14;0.21]	0.17 [0.16;0.18]	0.19 [0.16;0.22]	0.17 [0.16;0.18]	0.18 [0.17;0.20]	0.17 [0.15;0.18]						
STS of pelvis	0.18 [0.14;0.21]	0.18 [0.17;0.19]	0.10 [0.08;0.12]	0.17 [0.16;0.18]	0.18 [0.16;0.20]	0.15 [0.13;0.16]						
STS of brain and other parts of the nervous system	0.17 [0.14;0.21]	0.18 [0.17;0.20]	0.21 [0.18;0.25]	0.15 [0.14;0.16]	0.14 [0.13;0.16]	0.14 [0.12;0.15]						
STS of breast	0.16 [0.13;0.20]	0.18 [0.16;0.19]	0.17 [0.14;0.20]	0.16 [0.15;0.17]	0.16 [0.15;0.18]	0.12 [0.11;0.14]						
Embryonal rhabdomyosarcoma of soft tissue	0.12 [0.08;0.15]	0.09 [0.08;0.10]	0.04 [0.02;0.05]	0.07 [0.06;0.07]	0.07 [0.06;0.08]	0.05 [0.04;0.06]						
Ewing sarcoma of soft tissue	0.05 [0.03;0.07]	0.07 [0.07;0.08]	0.09 [0.07;0.12]	0.06 [0.05;0.07]	0.07 [0.06;0.09]	0.04 [0.04;0.05]						
Alveolar rhabdomyosarcoma of soft tissue	0.06 [0.04;0.09]	0.04 [0.04;0.05]	0.02 [0.01;0.03]	0.05 [0.04;0.05]	0.03 [0.03;0.04]	0.02 [0.02;0.03]						
STS of mediastinum	0.04 [0.03;0.06]	0.03 [0.02;0.03]	0.03 [0.02;0.04]	0.02 [0.02;0.02]	0.03 [0.02;0.03]	0.03 [0.02;0.04]						
STS of para testis	0.03 [0.02;0.05]	0.03 [0.03;0.04]	0.04 [0.02;0.05]	0.03 [0.02;0.03]	0.03 [0.02;0.04]	0.01 [0.01;0.02]						
STS of heart	0.01 [0.00;0.03]	0.01 [0.01;0.02]	0.01 [0.01;0.02]	0.01 [0.01;0.02]	0.01 [0.01;0.02]	0.01 [0.01;0.02]						
STS of paraorbit	0.02 [0.01;0.03]	0.01 [0.01;0.01]	0.00 [0.00;0.01]	0.01 [0.00;0.01]	0.01 [0.00;0.01]	0.01 [0.00;0.01]						
Bone sarcoma	1.22 [1.13;1.32]	0.91 [0.89;0.94]	0.73 [0.68;0.80]	0.75 [0.73;0.78]	0.93 [0.89;0.97]	0.70 [0.67;0.73]						
Chondrogenic sarcoma	0.38 [0.33;0.43]	0.30 [0.29;0.32]	0.26 [0.22;0.29]	0.20 [0.19;0.22]	0.20 [0.18;0.22]	0.20 [0.19;0.22]						
Osteogenic sarcoma	0.27 [0.22;0.32]	0.22 [0.21;0.24]	0.21 [0.18;0.25]	0.22 [0.21;0.24]	0.20 [0.18;0.22]	0.19 [0.17;0.20]						
Ewing sarcoma of bone	0.19 [0.15;0.24]	0.15 [0.14;0.17]	0.07 [0.05;0.09]	0.14 [0.13;0.15]	0.16 [0.14;0.18]	0.11 [0.10;0.13]						
Notochordal sarcoma, chordoma	0.05 [0.04;0.07]	0.05 [0.04;0.05]	0.07 [0.05;0.09]	0.04 [0.03;0.04]	0.04 [0.04;0.05]	0.03 [0.02;0.03]						
Other high grade bone sarcomas (fibrosarcoma, malignant fibrous histiocytoma)	0.02 [0.01;0.03]	0.01 [0.01;0.02]	0.02 [0.01;0.03]	0.01 [0.01;0.01]	0.02 [0.02;0.03]	0.03 [0.02;0.03]						

ESR: age-standardised rate using the European Standard Population of 1976 (N/100,000 person years)

* Data from RARECARENet - On line Analysis (istitutotumori.mi.it) ⁽²⁹⁾

Source: Belgian Cancer Registry ^{†††}

Figure 5 Comparison of male sarcoma World standardised incidence rates (N/100,000) with neighbouring countries, 2010-2013



WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

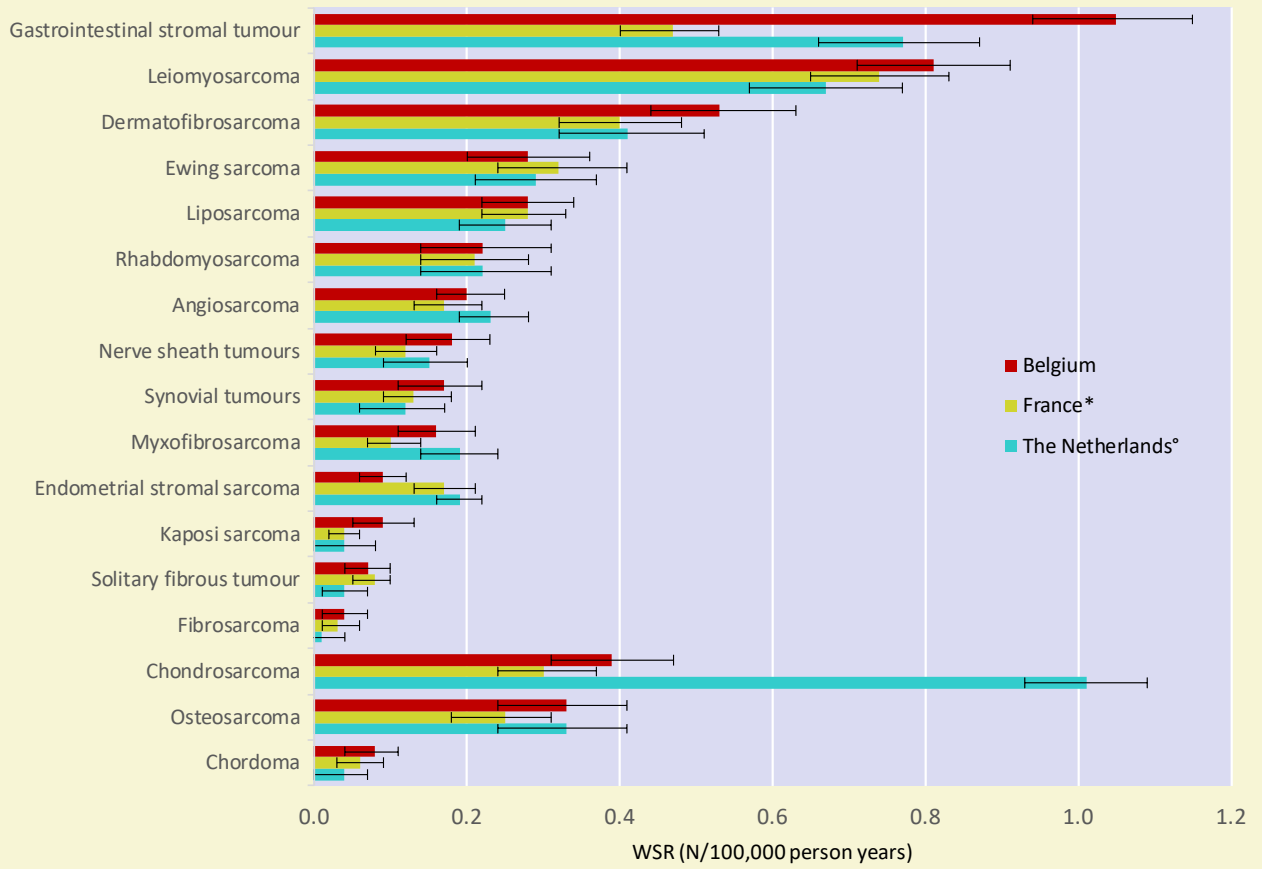
The age-specific incidence rate values are represented with 95% Confidence Intervals

* Data from Amadeo et al., 2020⁽¹⁶⁾

° Data from Dutch Cancer Registry administrated by IKNL (Integraal Kankercentrum Nederland)

Figure 6

Comparison of female sarcoma World standardised incidence rates (N/100,000) with neighbouring countries, 2010-2013



Source: Belgian Cancer Registry 

WSR: age-standardised rate using the World Standard Population (N/100,000 person years)


The age-specific incidence rate values are represented with 95% Confidence Intervals

* Data from Amadeo et al., 2020⁽¹⁶⁾

° Data from Dutch Cancer Registry administrated by IKNL (Integraal Kankercentrum Nederland)

Table 2 Comparison of sarcoma World Standardised incidence Rates (N/100,000) with neighbouring countries, 2010-2013

	Belgium	France*	The Netherlands°
Male	WSR [95% CI]	WSR [95% CI]	WSR [95% CI]
Soft tissue sarcoma			
Gastrointestinal stromal tumour	1.32 [1.20;1.44]	0.52 [0.45;0.60]	0.88 [0.80;0.96]
Liposarcoma	0.51 [0.43;0.59]	0.48 [0.41;0.55]	0.45 [0.39;0.51]
Kaposi sarcoma	0.47 [0.39;0.55]	0.40 [0.33;0.46]	0.48 [0.42;0.55]
Leiomyosarcoma	0.44 [0.37;0.51]	0.43 [0.37;0.50]	0.49 [0.43;0.55]
Dermatofibrosarcoma	0.39 [0.31;0.47]	0.31 [0.24;0.37]	0.31 [0.25;0.37]
Ewing sarcoma	0.31 [0.22;0.39]	0.35 [0.26;0.43]	0.33 [0.25;0.40]
Rhabdomyosarcoma	0.27 [0.19;0.36]	0.26 [0.19;0.34]	0.29 [0.22;0.36]
Myxofibrosarcoma	0.25 [0.19;0.31]	0.16 [0.11;0.20]	0.21 [0.17;0.25]
Synovial tumours	0.19 [0.13;0.25]	0.16 [0.11;0.20]	0.20 [0.15;0.25]
Nerve sheath tumours	0.16 [0.11;0.21]	0.10 [0.06;0.14]	0.15 [0.11;0.19]
Angiosarcoma	0.16 [0.11;0.20]	0.10 [0.07;0.14]	0.16 [0.12;0.19]
Solitary fibrous tumour	0.09 [0.05;0.12]	0.11 [0.08;0.15]	0.04 [0.02;0.06]
Fibrosarcoma	0.06 [0.03;0.09]	0.03 [0.02;0.05]	0.02 [0.00;0.03]
Endometrial stromal sarcoma	-	-	-
Bone sarcoma			
Osteosarcoma	0.41 [0.31;0.51]	0.34 [0.26;0.42]	0.39 [0.32;0.47]
Chondrosarcoma	0.29 [0.22;0.35]	0.32 [0.25;0.38]	0.83 [0.74;0.92]
Chordoma	0.08 [0.05;0.12]	0.11 [0.08;0.14]	0.09 [0.06;0.11]
Female	WSR [95% CI]	WSR [95% CI]	WSR [95% CI]
Soft tissue sarcoma			
Gastrointestinal stromal tumour	1.05 [0.94;1.15]	0.47 [0.40;0.53]	0.77 [0.69;0.84]
Leiomyosarcoma	0.81 [0.71;0.91]	0.74 [0.65;0.83]	0.67 [0.60;0.74]
Dermatofibrosarcoma	0.53 [0.44;0.63]	0.40 [0.32;0.48]	0.41 [0.34;0.48]
Ewing sarcoma	0.28 [0.20;0.36]	0.32 [0.24;0.41]	0.29 [0.22;0.36]
Liposarcoma	0.28 [0.22;0.34]	0.28 [0.22;0.33]	0.25 [0.20;0.30]
Rhabdomyosarcoma	0.22 [0.14;0.31]	0.21 [0.14;0.28]	0.22 [0.16;0.28]
Angiosarcoma	0.20 [0.16;0.25]	0.17 [0.13;0.22]	0.23 [0.19;0.27]
Nerve sheath tumours	0.18 [0.12;0.23]	0.12 [0.08;0.16]	0.15 [0.11;0.19]
Synovial tumours	0.17 [0.11;0.22]	0.13 [0.09;0.18]	0.12 [0.08;0.16]
Myxofibrosarcoma	0.16 [0.11;0.21]	0.10 [0.07;0.14]	0.19 [0.15;0.23]
Endometrial stromal sarcoma	0.09 [0.06;0.12]	0.17 [0.13;0.21]	0.19 [0.15;0.22]
Kaposi sarcoma	0.09 [0.05;0.13]	0.04 [0.02;0.06]	0.04 [0.02;0.06]
Solitary fibrous tumour	0.07 [0.04;0.10]	0.08 [0.05;0.10]	0.04 [0.02;0.06]
Fibrosarcoma	0.04 [0.01;0.07]	0.03 [0.01;0.06]	0.01 [0.00;0.02]
Bone sarcoma			
Chondrosarcoma	0.39 [0.31;0.47]	0.30 [0.24;0.37]	1.01 [0.91;1.11]
Osteosarcoma	0.33 [0.24;0.41]	0.25 [0.18;0.31]	0.33 [0.26;0.40]
Chordoma	0.08 [0.04;0.11]	0.06 [0.03;0.09]	0.04 [0.02;0.06]

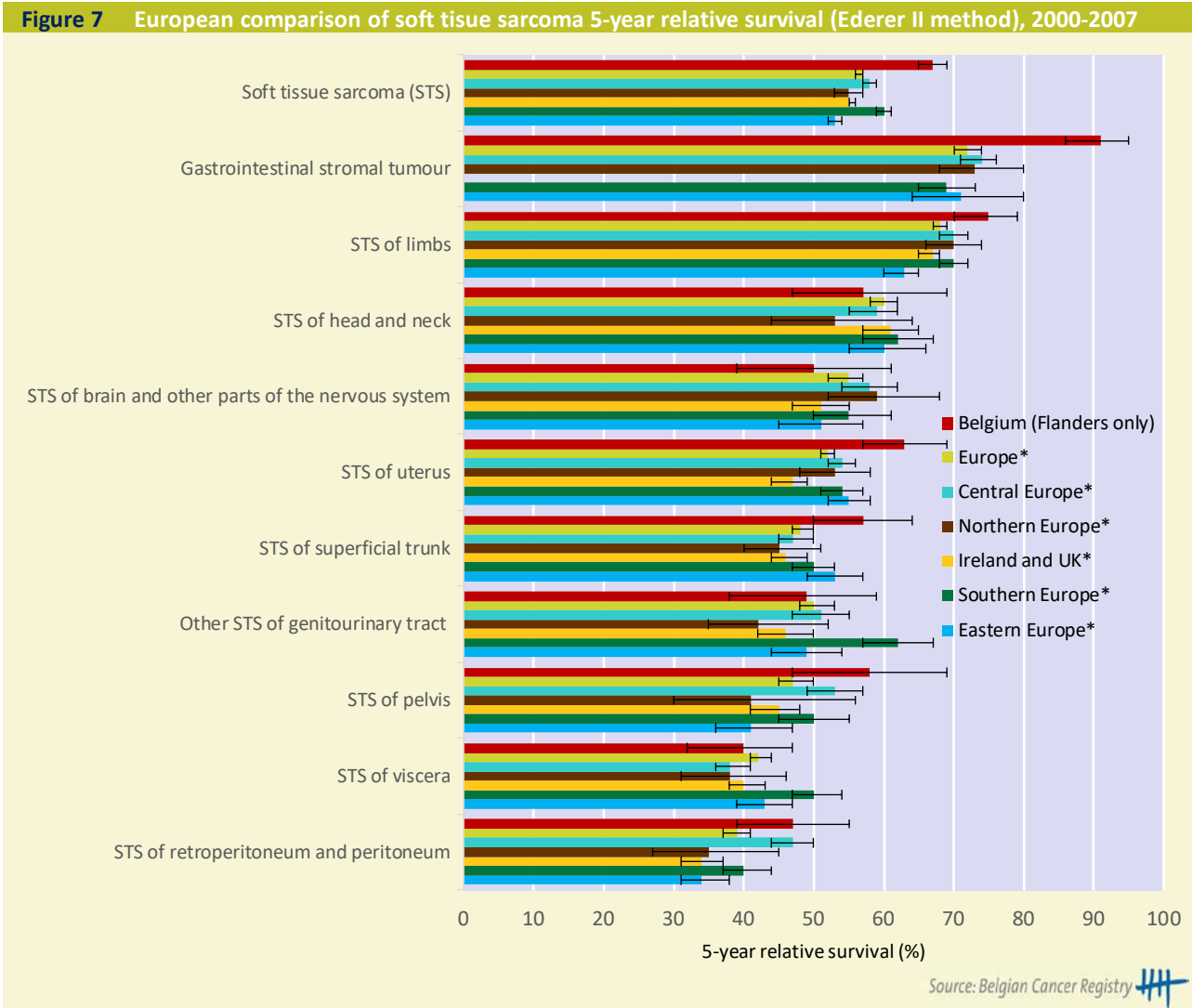
Source: Belgian Cancer Registry 

WSR: age-standardised rate using the World Standard Population (N/100,000 person years)

* Data from Amadeo et al., 2020⁽¹⁶⁾

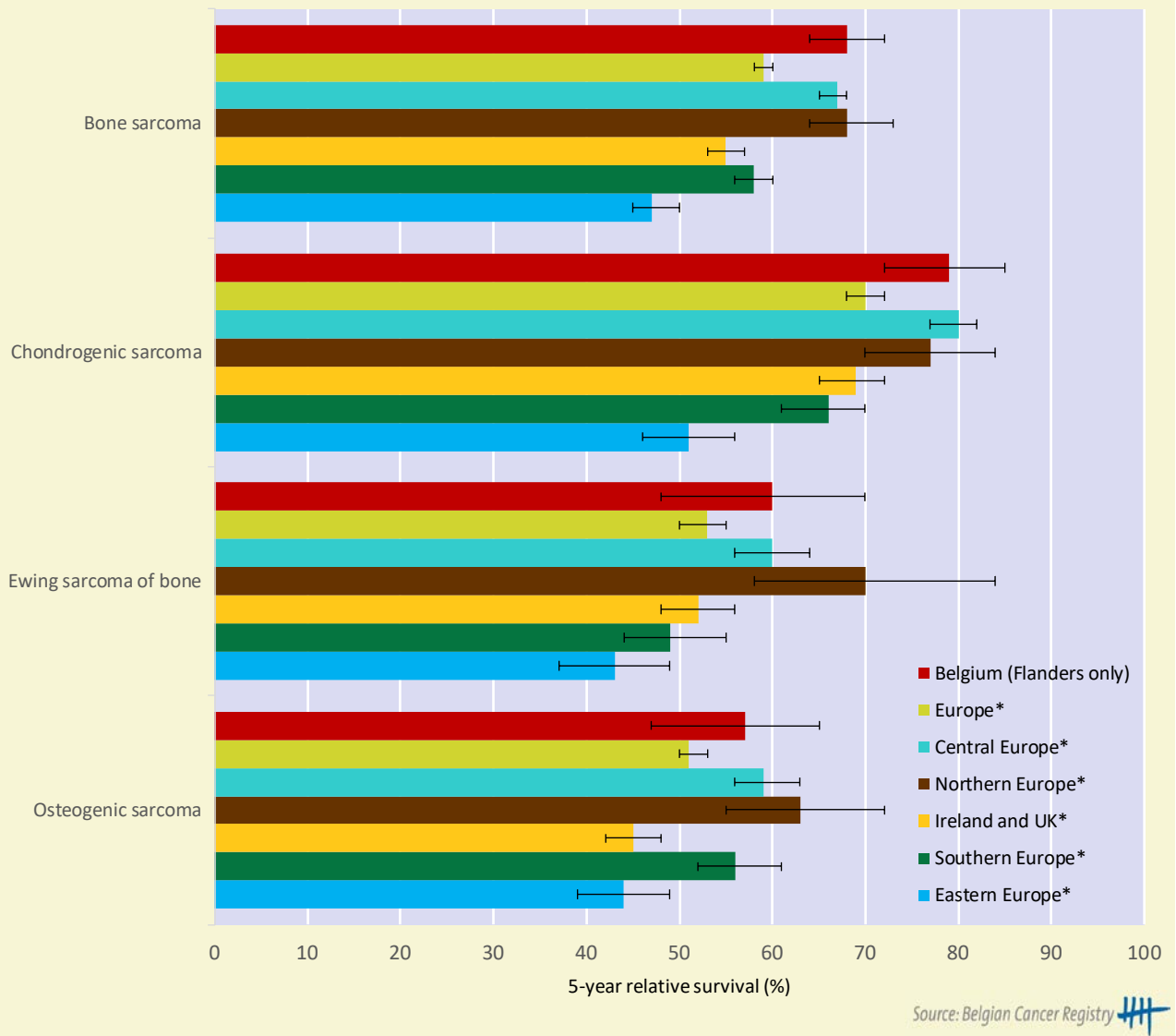
° Data from Dutch Cancer Registry administrated by IKNL (Integraal Kankercentrum Nederland)

Survival



The relative survival values are represented with 95% Confidence Intervals
 * Data from RARECARENet - On line Analysis (istitutotumori.mi.it)⁽²⁹⁾

Figure 8 European comparison of bone sarcoma 5-year relative survival (Ederer II method), 2000-2007



The relative survival values are represented with 95% Confidence Intervals

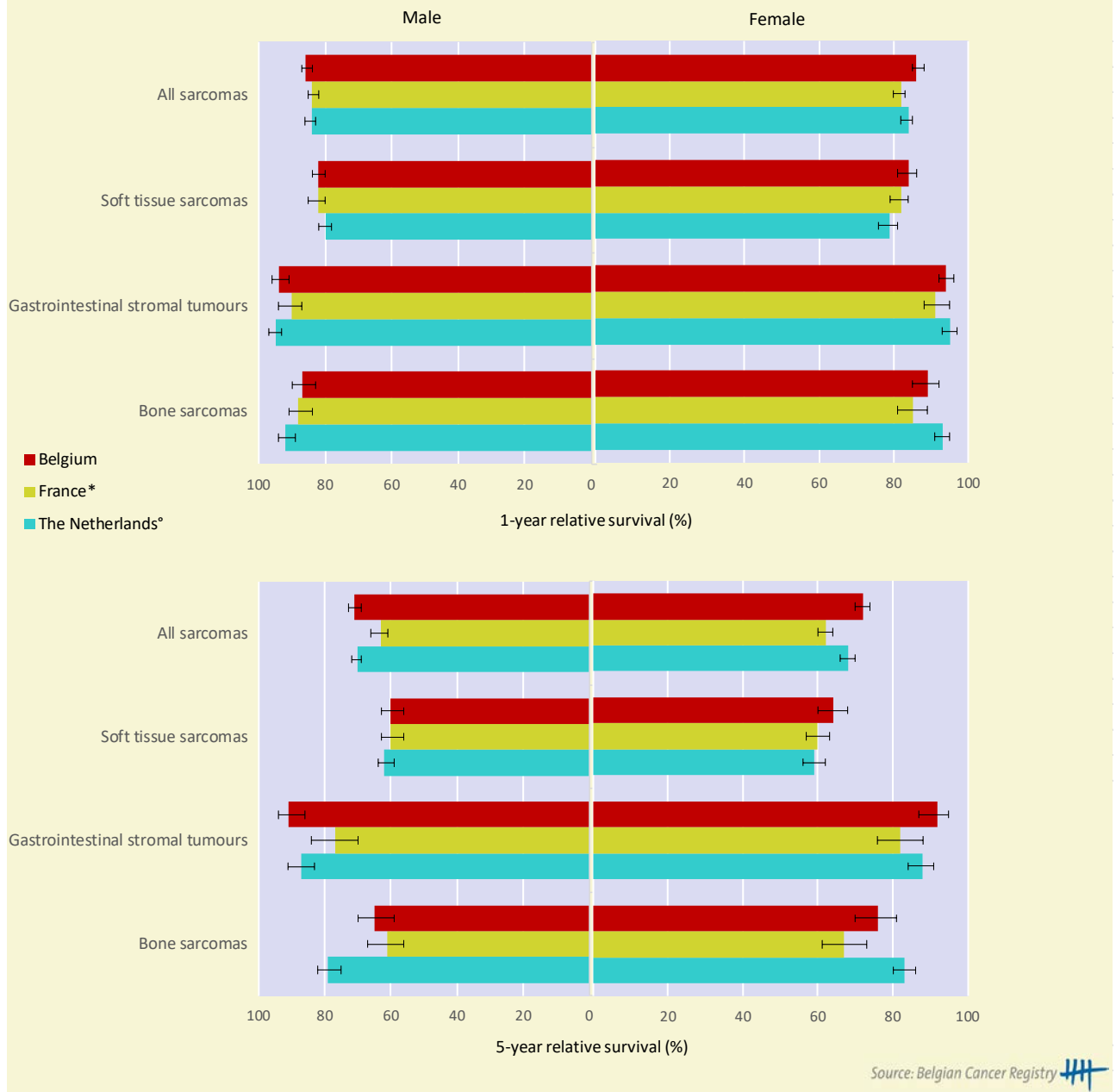
* Data from RARECARENet - On line Analysis (istitutotumori.mi.it)⁽²⁹⁾

Table 3 European comparison of sarcoma 5-year relative survival (Ederer II method), 2000-2007									
5-y relative survival [95% CI]									
	Belgium (Flanders)	Europe*	Central Europe*	Northern Europe*	Ireland and UK*	Southern Europe*	Eastern Europe*		
Soft tissue sarcoma (STS)	67 [65;69]	57 [56;57]	58 [57;59]	55 [53;57]	55 [55;56]	60 [59;61]	53 [52;54]		
Gastrointestinal stromal tumour	91 [86;95]	72 [70;74]	74 [71;76]	73 [68;80]	-	69 [65;73]	71 [64;80]		
STS of limbs	75 [70;79]	68 [67;69]	70 [68;72]	70 [66;74]	67 [65;68]	70 [68;72]	63 [60;65]		
STS of head and neck	57 [47;69]	60 [58;62]	59 [55;62]	53 [44;64]	61 [57;65]	62 [57;67]	60 [55;66]		
STS of brain and other parts of the nervous system	50 [39;61]	55 [52;57]	58 [54;62]	59 [52;68]	51 [47;55]	55 [50;61]	51 [45;57]		
STS of uterus	63 [57;69]	52 [51;53]	54 [52;56]	53 [48;58]	47 [44;49]	54 [51;57]	55 [52;58]		
STS of superficial trunk	57 [50;64]	48 [47;50]	47 [45;50]	45 [40;51]	46 [44;49]	50 [47;53]	53 [49;57]		
Other STS of genitourinary tract	49 [38;59]	50 [48;53]	51 [47;55]	42 [35;52]	46 [42;50]	62 [57;67]	49 [44;54]		
STS of pelvis	58 [47;69]	47 [45;50]	53 [49;57]	41 [30;56]	45 [41;48]	50 [45;55]	41 [36;47]		
STS of viscera	40 [32;47]	42 [41;44]	38 [36;41]	38 [31;46]	40 [38;43]	50 [47;54]	43 [39;47]		
STS of retroperitoneum and peritoneum	47 [39;55]	39 [37;41]	47 [44;50]	35 [27;45]	34 [31;37]	40 [37;44]	34 [31;38]		
Bone sarcoma	68 [64;72]	59 [58;60]	67 [65;68]	68 [64;73]	55 [53;57]	58 [56;60]	47 [45;50]		
Chondrogenic sarcoma	79 [72;85]	70 [68;72]	80 [77;82]	77 [70;84]	69 [65;72]	66 [61;70]	51 [46;56]		
Ewing sarcoma of bone	60 [48;70]	53 [50;55]	60 [56;64]	70 [58;84]	52 [48;56]	49 [44;55]	43 [37;49]		
Osteogenic sarcoma	57 [47;65]	51 [50;53]	59 [56;63]	63 [55;72]	45 [42;48]	56 [52;61]	44 [39;49]		

Source: Belgian Cancer Registry 

* Data from RARECARENet - On line Analysis (istitutotumori.mi.it)⁽²⁹⁾

Figure 9 Comparison of sarcoma relative survival (Pohar-Perme method) with neighbouring countries, 2010-2015



Only patients aged ≥15 years old at the time of diagnosis were included

The relative survival values are represented with 95% Confidence Intervals

* Data from Institut national du Cancer, March 2021⁽³⁰⁻³²⁾

° Data from Dutch Cancer Registry administrated by IKNL (Integraal Kankercentrum Nederland)

Table 4 Comparison of 1-year and 5-year sarcoma relative survival (Pohar-Perme method) with neighbouring countries, 2010-2015

1-y relative survival [95% CI]	Belgium			France*			The Netherlands°		
	Male	Female	Both sexes	Male	Female	Both sexes	Male	Female	Both sexes
All sarcomas	86% [84;87]	86% [85;88]	86% [85;87]	84% [82;85]	82% [80;83]	83% [82;84]	84% [83;86]	84% [82;85]	84% [83;85]
Soft tissue sarcoma	82% [80;84]	84% [81;86]	83% [81;85]	82% [80;85]	82% [79;84]	82% [81;84]	80% [78;82]	79% [76;81]	80% [78;81]
Gastrointestinal stromal tumour	94% [91;96]	94% [92;96]	94% [92;95]	90% [87;94]	91% [88;95]	91% [88;93]	95% [93;97]	95% [93;97]	95% [94;96]
Bone sarcoma	87% [83;90]	89% [85;92]	88% [85;90]	88% [84;91]	85% [81;89]	86% [84;89]	92% [89;94]	93% [91;95]	92% [91;94]
5-y relative survival [95% CI]									
All sarcomas	71% [69;73]	72% [70;74]	72% [70;73]	63% [61;66]	62% [60;64]	63% [61;64]	70% [69;72]	68% [66;70]	69% [68;70]
Soft tissue sarcoma	60% [56;63]	64% [60;68]	62% [59;64]	60% [56;63]	60% [57;63]	60% [58;62]	62% [59;64]	59% [56;62]	61% [59;63]
Gastrointestinal stromal tumour	91% [86;94]	92% [87;95]	91% [88;94]	77% [70;84]	82% [76;88]	79% [75;84]	87% [83;91]	88% [84;91]	88% [85;90]
Bone sarcoma	65% [59;70]	76% [70;81]	70% [66;74]	61% [56;67]	67% [61;73]	64% [60;68]	79% [75;82]	83% [80;86]	81% [79;83]

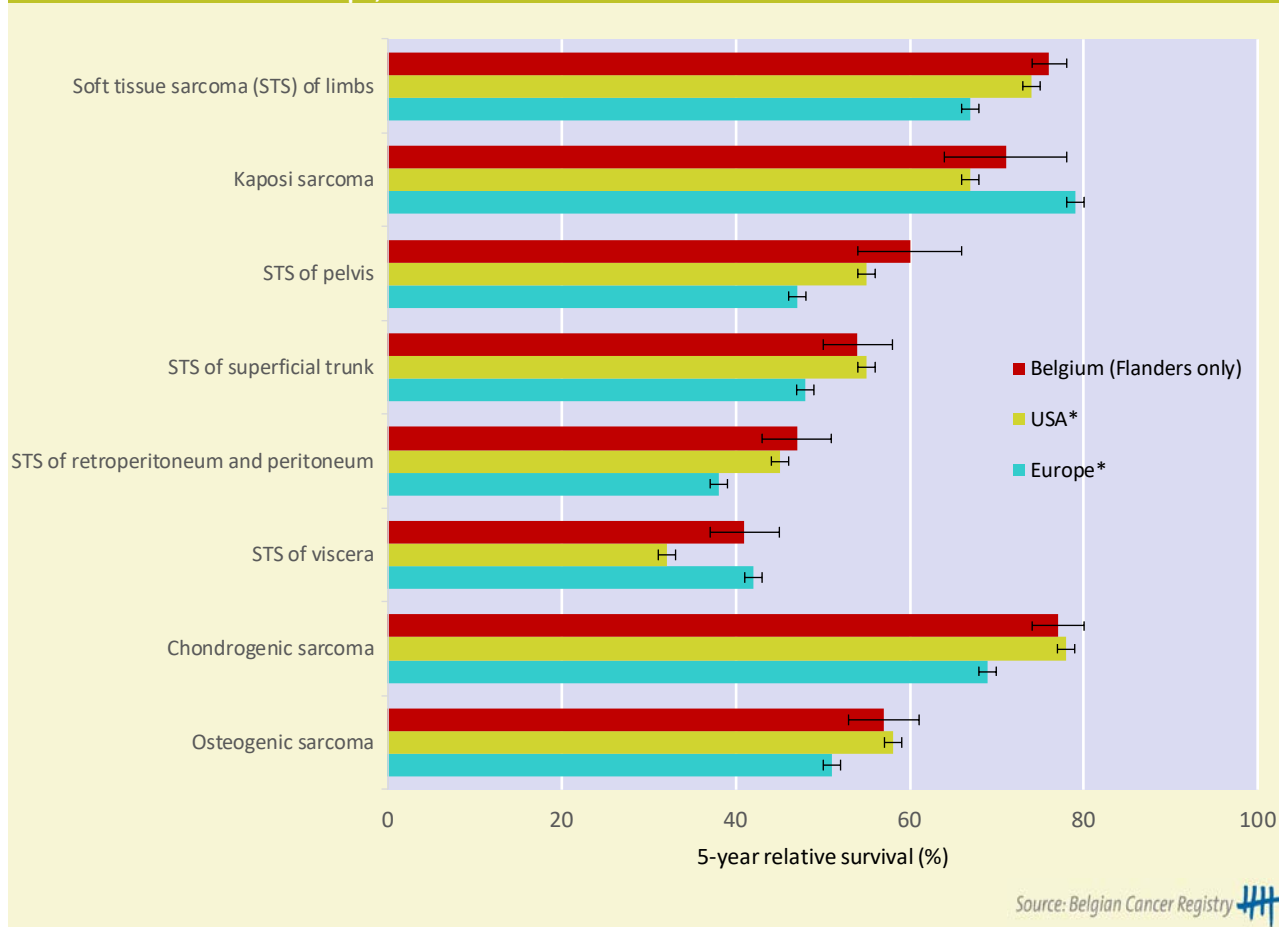
Source: Belgian Cancer Registry

Only patients aged ≥15 years old at the time of diagnosis were included in data

* Data from Institut national du Cancer, March 2021⁽³⁰⁻³²⁾

° Data from Dutch Cancer Registry administrated by IKNL (Integraal Kankercentrum Nederland)

Figure 10 Comparison of sarcoma 5-year relative survival (Pohar-Perme method) with The United States of America and Europe, 2000-2007



The relative survival values are represented with standard error

* Data from Botta et al., 2020⁽²⁸⁾

Table 5 Comparison of sarcoma 5-year relative survival (Pohar-Perme method) with the United States of America (USA) and Europe, 2000-2007

5-year relative survival (standard error)			
	Belgium (Flanders)	USA*	Europe*
Soft tissue sarcoma (STS)			
STS of limbs	77% (2%)	74% (1%)	67% (1%)
Kaposi sarcoma	71% (7%)	67% (1%)	79% (1%)
STS of pelvis	60% (6%)	55% (1%)	47% (1%)
STS of superficial trunk	54% (4%)	55% (1%)	48% (1%)
STS of retroperitoneum and peritoneum	47% (4%)	45% (1%)	38% (1%)
STS of viscera	41% (4%)	32% (1%)	42% (1%)
Bone sarcoma			
Chondrogenic sarcoma	77% (3%)	78% (1%)	69% (1%)
Osteogenic sarcoma	57% (4%)	58% (1%)	51% (1%)

Source: Belgian Cancer Registry

* Data from Botta et al., 2020⁽²⁸⁾

5 LESSONS LEARNED AND RECOMMENDATIONS

Cancer registration in Belgium

- Sustained work to improve quality of cancer registration is important to achieve the most specifically encoded data as possible. Methods such as the (online) trainings by the BCR for data managers in Belgian hospitals and continuous monitoring of registration quality and feedback to data sources should limit the number of incorrectly or imprecisely registered tumours. This in combination with continued data cleaning efforts by the BCR should limit the number of cases registered with e.g. a non-specific histological tumour entity or primary tumour location, missing staging information, coding errors etc. The table in appendix I will also help data managers with the coding of future registrations in combination with the online training from December 2021 that is available through our website. Also, the data managers in the hospitals should be able to rely on the help of the physicians in the hospitals, specifically for this diverse and rare tumour types.
- Although significant improvements were made over time, continued efforts are needed to improve exhaustivity of cancer registration. **Section 2.2.1** of this publication clearly illustrates in **figure 1** and **table 2** that not all soft tissue and bone tumours eligible for cancer registration are actually registered by the clinical network and that for certain tumour types the BCR is still depending for significant percentages on the pathological network only. A registration from both networks is vital to ensure both completeness and to get a detailed view on all cases as datasets from both networks are not fully identical. A specific attention in the hospitals should go to identify or flag all sarcomas to include all these tumours in the cancer registration. Cancer registration in hospitals should (if not already) be centralized to capture all cases, also those treated outside of the typical oncology-related departments.
- It is currently not obliged to register soft tissue and bone tumours of intermediate biologic potential in Belgium. As it concerns locally aggressive tumours or tumours with low metastatic potential that can have a serious impact on the patients' lives and prognosis, we recommend adding them to the inclusion criteria for cancer registration. Certain non-malignant bone and soft tissue tumours require aggressive treatment with severe functional sequelae for the patient (e.g. giant cell tumours, desmoid fibromatosis). In addition, this would allow for a continuous and consistent follow-up of the incidence trends over time for those tumour entities where the classification of the tumour behaviour (malignant or of intermediate biologic potential) might change over time as we have seen in the past for several entities included in this publication.
- Availability of multidisciplinary team meeting (MOC/COM) (textual) reports would improve the possibilities of the BCR to perform data cleaning by providing a more complete overview of the patient's his or her dossier. Currently, the available information to further specify or correct data, or to link and combine data from several sources is limited to the structured datasets registered and the pathology reports without any information on imaging, clinical examinations, molecular analyses etc.

Availability of molecular data

An inherent weakness of the current report is the lack of molecular data. Therefore, the availability of such data to the BCR should be improved. Multiple complementary methods could be used and the ongoing initiatives should be further exploited:

- Standardising pathology reports or more structured delivery of pathology data to the BCR would allow for more data being available in a structured and exploitable way. Especially the availability of molecular information would benefit from this as molecular data are not included in the current (legally defined) registration form.
- Additional efforts to extract molecular data from unstructured pathology reports using text mining, natural language processing and related techniques.
- Extending and updating the current cancer registration form used by the oncological care programs to structurally register such data.
- Coupling the cancer registration database with other databases such as the PITTER registry (predictive tests for a therapeutic response).

Availability of international data

Availability of international data on soft tissue and bone tumours is limited and hindered by changes in classification and inclusion criteria between time periods, countries, publication etc. A more standardized reporting and/or clear specification of inclusion criteria on a detailed level could render the comparison and combination of data between countries and regions easier. International guidelines on how to report specific tumour entities, such as the various types of bone and soft tissue tumours, should improve uniform reporting. Such guidelines should include uniform inclusion criteria for reporting that provide ways to deal with cancer registration-specific issues such as the reporting of historical data registered using continuously evolving tumour classifications. Especially for soft tissue and bone tumours this is relevant due to their rare and diverse nature and the inherent complexity of the classification and the evolution thereof. Compiling an up-to-date classification that resembles the current WHO classification⁽¹⁾ as closely as possible that could also be used retrospectively and cope with registrations coded with limited specificity proved to be one of the biggest challenges when preparing this report.

Multidisciplinary team meetings

Although soft tissue and bone tumours are rare, only 77% (see **chapter 2.2.1, table 2**) is discussed in a MOC/COM (detailed information in **chapter 2.2.1, table 2** and **figure 2**). Both the percentage of cases discussed during a MOC/COM discussion and the percentage of registrations at the BCR by the clinical network drastically drops after the age of 15 years (see **chapter 2.2.1, figure 3-4**). This difference in clinical approach and exhaustivity is something that must be considered by the hospitals. Increasing the percentage of cases discussed on a MOC/COM for tumours of bone and soft tissue might be beneficial for patient care and prognosis. It might also aid in improving the exhaustivity of cancer registration. Although the obligation to register specific cases is not dependent on the case being discussed at a MOC/COM, due to the close relation between registration and the MOC/COM setting, cancers not discussed at a MOC/COM are more prone to be missed by the clinical network when registering.

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Classification of Soft Tissue and Bone Tumours based on ICD-O-3.2 (to be used for new registrations from 2020)

WHO classification of soft tissue tumours	ICD-O-3.2	Comments	Topography
Adipocytic tumours			
<i>Intermediate (locally aggressive)</i>			
Atypical lipomatous tumour	8850/1	If you have the information about "MDM2+", note it in comment field	It's important to have the initial topo to distinguish between 8851/3 and 8850/1
<i>Malignant</i>			
Well-differentiated liposarcoma, NOS	8851/3	If you have the information about "MDM2+", note it in comment field	It's important to have the initial topo to distinguish between 8851/3 and 8850/1
Lipoma-like liposarcoma	8851/3	If you have the information about "MDM2+", note it in comment field	It's important to have the initial topo to distinguish between 8851/3 and 8850/1
Inflammatory liposarcoma	8851/3	If you have the information about "MDM2+", note it in comment field	It's important to have the initial topo to distinguish between 8851/3 and 8850/1
Sclerosing liposarcoma	8851/3	If you have the information about "MDM2+", note it in comment field	It's important to have the initial topo to distinguish between 8851/3 and 8850/1
De differentiated liposarcoma	8858/3	If you have the information about "MDM2+", note it in comment field	
Myxoid liposarcoma	8852/3	If you have the information about "DDIT3+", note it in comment field	Extremities
Pleomorphic liposarcoma	8854/3	If you have the information about "MDM2-" and "DDIT3-", note it in comment field	Extremities
Epithelioid liposarcoma	8854/3	If you have the information about "MDM2-" and "DDIT3-", note it in comment field	
Myxoid pleomorphic liposarcoma	8850/3	"8859/3" in comment, code does not yet exist in ICD-O-3.2 if you have the information about "TP53", note it in comment field Needs to be justified in comment	It's important to have the initial topo to distinguish between 8851/3 and 8850/1
<i>Liposarcoma, NOS</i>			
Fibroblastic and myofibroblastic tumours			
<i>Intermediate (locally aggressive)</i>			
Solitary fibrous tumour, benign	8815/0		
Palmar fibromatosis and plantar fibromatosis	8813/1		
Desmoid-type fibromatosis	8821/1		
Extra-abdominal desmoid	8821/1		
Abdominal fibromatosis	8822/1		
Lipofibromatosis	8851/1		
Giant cell fibroblastoma	8834/1		
<i>Intermediate (rarely metastasizing)</i>			
Dermatofibrosarcoma protuberans	8832/1		
Pigmented dermatofibrosarcoma protuberans (Bednar tumour)	8833/1		
Dermatofibrosarcoma protuberans, fibrosarcomatous	8832/3		
Solitary fibrous tumour, NOS	8815/1		(C44_) exceptionnally C49 (C44_) exceptionnally C49 (C44_) exceptionnally C49
Inflammatory myofibroblastic tumour	8825/1		
(Low-grade) myofibroblastic sarcoma	8825/3		
Superficial CD34-positive fibroblastic tumour	8810/1	"Superficial CD34-positive fibroblastic tumour" in comment, new in WHO 2020 (futur ICD-O-4)	
Myxoinflammatory fibroblastic sarcoma	8811/1	"Myxoinflammatory fibroblastic sarcoma" in comment	
Infantile fibrosarcoma	8814/3		
<i>Malignant</i>			
Solitary fibrous tumour, malignant	8815/3		
Fibrosarcoma, NOS	8810/3		
Myxofibrosarcoma	8811/3		
Low-grade fibromyxoid sarcoma	8840/3		
So-called fibrohistiocytic tumours			
<i>Intermediate (rarely metastasizing)</i>			
Plexiform fibrohistiocytic tumour	8835/1		
Giant cell tumour of soft parts	9251/1		
<i>Malignant</i>			
Malignant tenosynovial giant cell tumour	9252/3		(C49_)

Classification of Soft Tissue and Bone Tumours based on ICD-O-3.2 (to be used for new registrations from 2020)

WHO classification of soft tissue tumours (continued)	ICD-O-3.2	Comments	Topography
Vascular tumours			
<i>Intermediate (locally aggressive)</i>			
Kaposiform haemangiioendothelioma	9130/1		
<i>Intermediate (rarely metastasizing)</i>			
Retiform haemangiioendothelioma	9136/1		
Composite haemangiioendothelioma	9136/1		
Neuroendocrine composite haemangiioendothelioma	9136/1		
Papillary intralymphatic angioendothelioma	9135/1		
Pseudomyogenic (epithelioid sarcoma-like) haemangiioendothelioma	9138/1		
Kaposi sarcoma	9140/3		
Classic indolent Kaposi sarcoma	9140/3		
Endemic African Kaposi sarcoma	9140/3		
AIDS-associated Kaposi sarcoma	9140/3		
Iatrogenic Kaposi sarcoma	9140/3		
<i>Malignant</i>			
Epithelioid haemangiioendothelioma NOS	9133/3		
Epithelioid haemangiioendothelioma with WWTR1-CAMTA1 fusion	9133/3		
Epithelioid haemangiioendothelioma with YAP1-TFE3 fusion	9133/3		
Angiosarcoma	9120/3		
Pericytic (perivascular) tumours			
<i>Intermediate</i>			
Glomangiomas	8711/1		
Glomus tumour of uncertain malignant potential	8711/1		
Myofibromatosis	8824/1		
Infantile myofibromatosis	8824/1		
<i>Malignant</i>			
Glomus tumour, malignant	8711/3		
Smooth muscle tumours			
<i>Intermediate</i>			
EBV-associated smooth muscle tumour, smooth muscle tumour NOS	8897/1	"EBV-associated smooth muscle tumour" in comment, new in WHO 2020 (futura ICD-O-4)	
<i>Malignant</i>			
Leiomyosarcoma, NOS	8890/3		
Skeletal muscle tumours			
<i>Malignant</i>			
Embryonal rhabdomyosarcoma, NOS	8910/3		
Embryonal rhabdomyosarcoma, pleomorphic	8910/3		
Alveolar rhabdomyosarcoma	8920/3		
Pleomorphic rhabdomyosarcoma, NOS	8901/3		
Spindle cell rhabdomyosarcoma	8912/3		
Congenital spindle cell rhabdomyosarcoma (with VGLL2/NCOA2/CITD2 rearrangements)	8912/3		
MYOD1-mutant spindle cell / sclerosing rhabdomyosarcoma	8912/3		
Intraosseous spindle cell rhabdomyosarcoma (with TFCP2/NCOA2 rearrangements)	8912/3		
Ectomesenchymoma	8921/3		
Rhabdomyosarcoma, NOS	8900/3		
Chondro-osseous tumours			
<i>Malignant</i>			
Osteosarcoma, extraskeletal	9180/3		

Classification of Soft Tissue and Bone Tumours based on ICD-O-3.2 (to be used for new registrations from 2020)

WHO classification of soft tissue tumours (continued)	ICD-O-3.2	Comments	Topography
Peripheral nerve sheath tumours			
<i>Malignant</i>			
Malignant peripheral nerve sheath tumour (MPNST), NOS	9540/3		
Malignant melanotic nerve sheath tumour	9540/3	"Malignant melanotic nerve sheath tumour" in comment, new in WHO 2020 (future ICD-O-4)	
Malignant peripheral nerve sheath tumour (MPNST) with skeletal muscle differentiation	9561/3		
Malignant Triton tumour	9561/3		
Malignant peripheral nerve sheath tumour (MPNST), epithelioid	9542/3		
Granular cell tumour, malignant	9580/3		
Perineurioma, malignant	9571/3		
Tumours of uncertain differentiation			
<i>Intermediate (locally aggressive)</i>			
Haemosiderotic fibroplomatous tumour	8811/1		
Angiomyolipoma, epithelioid	8860/1	"Haemosiderotic fibroplomatous tumour" in comment	
<i>Intermediate (rarely metastasizing)</i>			
Atypical fibroxanthoma	8830/1		
Angiomatoid fibrous histiocytoma	8836/1		
Ossifying fibromyxoid tumour, NOS	8842/0		
Mixed tumour, NOS	8940/0		
Mixed tumour, malignant	8940/3		
Myoepithelioma, NOS	8982/0		
<i>Malignant</i>			
Phosphatric mesenchymal tumour, malignant;	8990/3		
Mesenchymoma, malignant	8990/3		
NTRK-rearranged spindle cell neoplasm (emerging)	8800/3	"NTRK-rearranged spindle cell neoplasm (emerging), malignant" in comment	
Synovial sarcoma, NOS	9040/3		
Synovial sarcoma, poorly differentiated	9040/3		
Synovial sarcoma, spindle cell	9041/3		
Synovial sarcoma, biphasic	9043/3		
Epithelioid sarcoma	8804/3		
Proximal or large cell epithelioid sarcoma	8804/3		
Classic epithelioid sarcoma	8804/3		
Alveolar soft part sarcoma	9581/3		
Clear cell sarcoma, NOS	9044/3		
Clear cell sarcoma of kidney	8964/3		(C64_)
Extraskeletal myxoid chondrosarcoma	9231/3		
Desmoplastic small round cell tumour	8806/3		
Rhabdoid tumour, NOS	8963/3		
Atypical teratoid/rhabdoid tumour	9508/3		
Pervascular epithelioid tumor (PEComa), malignant	8714/3		
Infantal sarcoma	9137/3		
Ossifying fibromyxoid tumour, malignant	8842/3		
Myoepithelial carcinoma	8982/3		
Gastrointestinal stromal tumours			
Gastrointestinal stromal tumour	8936/3	Mitotic rate should be added in comment	
Endometrial stromal sarcoma			
<i>Malignant</i>			
Endometrial stromal sarcoma, high grade, NOS	8990/3		(C54.1)
Endometrial stromal sarcoma, low grade	8991/3		(C54.1)
			Outside the central nervous system (renal or extrarenal) (C71_)

Classification of Soft Tissue and Bone Tumours based on ICD-O-3.2 (to be used for new registrations from 2020)

WHO classification of soft tissue tumours (continued)		ICD-O-3.2	Comments	Topography
Undefined and poorly characterised tumours				
<i>Malignant</i>				
Stromal sarcoma, NOS		8935/3	Needs to be justified in comment	
Stromal tumour of uncertain malignant potential, NOS		8935/1	Needs to be justified in comment	
Sarcoma, NOS		8800/3	Needs to be justified in comment	
WHO classification of undifferentiated small round cell sarcomas of bone and soft tissue				
Ewing sarcoma		9364/3		
Round cell sarcoma with EWSR1-non-ETS fusions		8803/3	9366/3 in comment. Code does not yet exist in ICD-O-3.2	
CIC-rearranged sarcoma		8803/3	9367/3 in comment. Code does not yet exist in ICD-O-3.2	
Sarcoma with BCOR genetic alterations		8803/3	9368/3 in comment. Code does not yet exist in ICD-O-3.2	
WHO classification of bone tumours				
Chondrogenic tumours				
<i>Intermediate (locally aggressive)</i>				
Synovial chondromatosis		9220/1		(C40_ C41_) exceptionally soft tissue
Central atypical cartilaginous tumour		9222/1		(C40_ C41_) exceptionally soft tissue, use the initial topo to distinguish between 9222/3 & 9222/1
Secondary peripheral atypical cartilaginous tumour		9222/1		(C40_ C41_) exceptionally soft tissue, use the initial topo to distinguish between 9222/3 & 9222/1
<i>Malignant</i>				
Central chondrosarcoma, grade 1		9222/3		(C40_ C41_) exceptionally soft tissue, use the initial topo to distinguish between 9222/3 & 9222/1
Secondary chondrosarcoma, grade 1		9222/3		(C40_ C41_) exceptionally soft tissue, use the initial topo to distinguish between 9222/3 & 9222/1
Central chondrosarcoma, grade 2		9220/3		(C40_ C41_) exceptionally soft tissue
Secondary peripheral chondrosarcoma, grade 2		9220/3		(C40_ C41_) exceptionally soft tissue
Central chondrosarcoma, grade 3		9220/3		(C40_ C41_) exceptionally soft tissue
Secondary peripheral chondrosarcoma, grade 3		9220/3		(C40_ C41_) exceptionally soft tissue
Periosteal chondrosarcoma		9221/3		(C40_ C41_) exceptionally soft tissue
Clear cell chondrosarcoma		9242/3		(C40_ C41_) exceptionally soft tissue
Mesenchymal chondrosarcoma		9240/3		(C40_ C41_) also possible in soft tissue, and intracranial sites
Dedifferentiated chondrosarcoma		9243/3		(C40_ C41_) exceptionally soft tissue
Osteogenic tumours				
<i>Intermediate (locally aggressive)</i>				
Osteoblastoma, NOS		9200/1		(C40_ C41_) exceptionally soft tissue
<i>Malignant</i>				
Low-grade central osteosarcoma		9187/3		(C40_ C41_) exceptionally soft tissue
Osteosarcoma, NOS		9180/3		(C40_ C41_) exceptionally soft tissue
Conventional osteosarcoma		9180/3		(C40_ C41_) exceptionally soft tissue
Telangiectatic osteosarcoma		9183/3		(C40_ C41_) exceptionally soft tissue
Small cell osteosarcoma		9185/3		(C40_ C41_) exceptionally soft tissue
Parosteal osteosarcoma		9192/3		(C40_ C41_) exceptionally soft tissue
Periosteal osteosarcoma		9193/3		(C40_ C41_) exceptionally soft tissue
High grade surface osteosarcoma		9194/3		(C40_ C41_) exceptionally soft tissue
Secondary osteosarcoma		9184/3		(C40_ C41_) exceptionally soft tissue
Fibrogenic tumours				
<i>Intermediate (locally aggressive)</i>				
Desmoplastic fibroma		8823/1		
<i>Malignant</i>				
Fibrosarcoma, NOS		8810/3		
Vascular tumours				
<i>Intermediate (locally aggressive)</i>				
Epithelioid haemangioma		9125/0		
<i>Malignant</i>				
Epithelioid haemangioidheilioma NOS		9133/3		
Angiosarcoma		9120/3		

Classification of Soft Tissue and Bone Tumours based on ICD-O-3.2 (to be used for new registrations from 2020)

WHO classification of soft tissue tumours (continued)	ICD-O-3.2	Comments	Topography
Osteoclastic giant cell-rich tumours			
<i>Intermediate (locally aggressive, rarely metastasizing)</i>			
Giant cell tumour of bone	9250/1		
<i>Malignant</i>			
Giant cell tumour of bone, malignant	9250/3		
Notochordal tumours			
<i>Malignant</i>			
Conventional chordoma	9370/3		
Poorly differentiated chordoma	9370/3		
Chondroid chordoma	9370/3		
Decidifferentiated chordoma	9372/3		
Other mesenchymal bone tumours			
<i>Intermediate (locally aggressive)</i>			
Osteofibrous dysplasia-like adamantinoma	9261/1		
Fibrocartilaginous mesenchymoma	8990/1		
<i>Malignant</i>			
Adamantinoma of long bones	9261/3		(C40._)
Decidifferentiated adamantinoma	9261/3		(C40._)
Leiomyosarcoma NOS	8890/3		(C40._, C41._)
Pleomorphic sarcoma, undifferentiated	8802/3		(C40._, C41._)

WHO classification of bone tumours and soft tissue tumours of uncertain differentiation

Tumours of uncertain differentiation			
<i>Malignant</i>			
Epithelioid sarcoma, NOS, undifferentiated	8804/3	After exclusion	
Undifferentiated sarcoma	8805/3	After exclusion	
Spindle cell sarcoma, undifferentiated	8801/3	After exclusion	
Pleomorphic cell sarcoma, undifferentiated	8802/3	After exclusion	
Round cell sarcoma, undifferentiated	8803/3	After exclusion	
Sarcoma, NOS	8800/3	Needs to be justified in comment	(C44._) for the pleomorphic dermal sarcoma

In grey: codes and / or names of entities which should be avoided in favour of a more specific code

APPENDIX II

Belgium: Number of new diagnoses (N), age-specific and age-standardised incidence (N/100,000) of bone and soft tissue tumours in males in 2010-2019 by histological subtype																	
	Number of new diagnoses (N)					Age specific incidence (N/100,000)					CR	ESR	WSR	CRI			
	Total	0-14y	15-29y	30-44y	45-59y	60-74y	75+	0-14y	15-29y	30-44y					45-59y	60-74y	75+
Sarcomas classified by primary tumour location																	
Soft tissue and visceral sarcoma	5,143	123	189	548	1,177	1,782	1,324	1.3	1.8	4.9	10.0	21.9	34.6	9.4	7.7	5.8	0.61
Bone sarcoma	606	68	154	88	96	121	79	0.7	1.5	0.8	0.8	1.5	2.1	1.1	1.1	1.0	0.08
Sarcomas classified by histological type																	
Liposarcoma	842	3	11	84	208	350	186	0.0	0.1	0.8	1.8	4.3	4.9	1.5	1.2	0.9	0.11
Liposarcoma well differentiated and atypical lipomatous tumours	302	-	1	25	81	130	65	-	0.0	0.2	0.7	1.6	1.7	0.6	0.4	0.3	0.04
Dedifferentiated liposarcoma	342	-	3	15	79	149	96	-	0.0	0.1	0.7	1.8	2.5	0.6	0.5	0.3	0.04
Myxoid liposarcoma	130	3	6	35	34	39	13	0.0	0.1	0.3	0.3	0.5	0.3	0.2	0.2	0.2	0.02
Liposarcoma NOS and other	68	-	1	9	14	32	12	-	0.0	0.1	0.1	0.4	0.3	0.1	0.1	0.1	0.01
Malignant (myo)fibroblastic tumours and so-called fibrohistiocytic tumours	639	10	54	131	156	168	120	0.1	0.5	1.2	1.3	2.1	3.1	1.2	1.0	0.8	0.08
Dermatofibrosarcoma protuberans	289	6	42	100	73	45	23	0.1	0.4	0.9	0.6	0.6	0.6	0.5	0.5	0.4	0.04
Solitary fibrous tumour	83	-	2	6	18	37	20	-	0.0	0.1	0.2	0.5	0.5	0.2	0.1	0.1	0.01
Fibrosarcoma	29	-	1	3	11	8	6	-	0.0	0.0	0.1	0.1	0.2	0.1	0.0	0.0	0.00
Myxofibrosarcoma	211	3	6	15	48	73	66	0.0	0.1	0.1	0.4	0.9	1.7	0.4	0.3	0.2	0.02
Miscellaneous malignant (myo)fibroblastic tumours and so-called fibrohistiocytic tumours	27	1	3	7	6	5	5	0.0	0.0	0.1	0.1	0.1	0.1	0.0	0.0	0.0	0.00
Vascular sarcoma	577	2	18	103	150	168	136	0.0	0.2	0.9	1.3	2.1	3.6	1.1	0.9	0.7	0.07
Kaposi sarcoma	362	-	15	85	112	88	62	-	0.1	0.8	0.9	1.1	1.6	0.7	0.6	0.4	0.04
Angiosarcoma	189	2	1	10	33	72	71	0.0	0.0	0.1	0.3	0.9	1.9	0.3	0.3	0.2	0.02
Leiomyosarcoma	470	2	8	29	96	153	182	0.0	0.1	0.3	0.8	1.9	4.8	0.9	0.7	0.5	0.05
Rhabdomyosarcoma	133	54	24	6	13	18	18	0.6	0.2	0.1	0.1	0.2	0.5	0.2	0.3	0.3	0.02
Peripheral nerve sheath tumours	128	2	7	30	37	30	22	0.0	0.1	0.3	0.3	0.4	0.6	0.2	0.2	0.2	0.02
Other tumours of uncertain differentiation	260	40	32	56	49	56	27	0.4	0.3	0.5	0.4	0.7	0.7	0.5	0.5	0.4	0.03
Synovial sarcoma	96	7	13	26	19	25	6	0.1	0.1	0.2	0.2	0.3	0.2	0.2	0.2	0.1	0.01
Myoepithelioma	37	1	3	4	8	11	10	0.0	0.0	0.0	0.1	0.1	0.3	0.1	0.1	0.0	0.00
Rhabdoid tumours	23	21	1	1	-	-	-	0.2	0.0	0.0	-	-	-	0.0	0.1	0.1	0.00
GIST	1,464	1	9	74	354	644	382	0.0	0.1	0.7	3.0	7.9	10.0	2.7	2.1	1.5	0.18
Ewing sarcoma	139	35	53	26	12	7	6	0.4	0.5	0.2	0.1	0.1	0.2	0.3	0.3	0.3	0.02
Chondrosarcoma	233	1	22	38	64	74	34	0.0	0.2	0.3	0.5	0.9	0.9	0.4	0.4	0.3	0.03
Osteosarcoma	222	37	80	21	27	32	25	0.4	0.8	0.2	0.2	0.4	0.7	0.4	0.4	0.4	0.03
Other bone tumours of uncertain differentiation	76	-	10	8	14	31	13	-	0.1	0.1	0.1	0.4	0.3	0.1	0.1	0.1	0.01
Chordoma	70	-	6	6	14	31	13	-	0.1	0.1	0.1	0.4	0.3	0.1	0.1	0.1	0.01
Unclassified and poorly characterised sarcoma	566	4	15	30	93	172	252	0.0	0.1	0.3	0.8	2.1	6.6	1.0	0.8	0.5	0.05
All sarcomas	5,749	191	343	636	1,273	1,903	1,403	2.0	3.4	5.7	10.8	23.4	36.7	10.5	8.7	6.8	0.69

CR: crude (all ages) incidence rate (N/100,000 pers on years)

ESR and WSR: age-standardised incidence using the European or World Standard Population (N/100,000 person years)

CRI: Cumulative risk 0-74 years (%)

Belgium: Number of new diagnoses (N), age-standardised incidence (N/100,000) of bone and soft tissue tumours in females in 2010-2019 by histological subtype

Sarcomas classified by primary tumour location	Number of new diagnoses (N)						Age specific incidence (N/100,000)						CR	ESR	WSR	CRI		
	Total		0-14y		15-29y		30-44y		45-59y		60-74y						75+	
	0-14y	15-29y	30-44y	45-59y	60-74y	75+	0-14y	15-29y	30-44y	45-59y	60-74y	75+						
Soft tissue and visceral sarcoma	4,891	97	193	509	1,205	1,533	1,354	1.0	1.9	4.6	10.3	17.5	22.3	8.6	6.6	5.1	0.53	
Bone sarcoma	498	69	83	86	111	92	57	0.7	0.8	0.8	1.0	1.1	0.9	0.9	0.9	0.8	0.07	
Sarcomas classified by histological type																		
Liposarcoma	513	1	10	43	163	159	137	0.0	0.1	0.4	1.4	1.8	2.3	0.9	0.7	0.5	0.06	
Liposarcoma well differentiated and atypical lipomatous tumours	192	-	1	14	69	72	36	-	0.0	0.1	0.6	0.8	0.6	0.3	0.3	0.2	0.02	
Dedifferentiated liposarcoma	190	-	1	5	56	56	72	-	0.0	0.0	0.5	0.6	1.2	0.3	0.2	0.2	0.02	
Myxoid liposarcoma	92	1	8	21	29	20	13	0.0	0.1	0.2	0.2	0.2	0.2	0.2	0.1	0.1	0.01	
Liposarcoma NOS and other	39	-	-	3	9	11	16	-	-	0.0	0.1	0.1	0.3	0.1	0.0	0.0	0.00	
Malignant (myo)fibroblastic tumours and so-called fibrohistiocytic tumours	614	13	73	134	158	130	106	0.1	0.7	1.2	1.4	1.5	1.7	1.1	1.0	0.8	0.07	
Dermatofibrosarcoma protuberans	329	6	56	113	100	36	18	0.1	0.6	1.0	0.9	0.4	0.3	0.6	0.6	0.5	0.04	
Solitary fibrous tumour	75	-	3	5	22	25	20	-	0.0	0.0	0.2	0.3	0.3	0.1	0.1	0.1	0.01	
Fibrosarcoma	29	3	5	-	5	6	10	0.0	0.0	-	0.0	0.1	0.2	0.1	0.0	0.0	0.00	
Myxofibrosarcoma	154	3	9	12	24	54	52	0.0	0.1	0.1	0.2	0.6	0.9	0.3	0.2	0.2	0.02	
Miscellaneous malignant (myo)fibroblastic tumours and so-called fibrohistiocytic tumours	27	1	-	4	7	9	6	0.0	-	0.0	0.1	0.1	0.1	0.0	0.0	0.0	0.00	
Vascular sarcoma	417	1	10	36	61	135	174	0.0	0.1	0.3	0.5	1.5	2.9	0.7	0.5	0.4	0.04	
Kaposi sarcoma	96	-	5	16	19	20	36	-	0.0	0.1	0.2	0.2	0.6	0.2	0.1	0.1	0.01	
Angiosarcoma	292	1	3	12	35	107	134	0.0	0.0	0.1	0.3	1.2	2.2	0.5	0.3	0.2	0.03	
Leiomyosarcoma	759	2	9	72	235	244	197	0.0	0.1	0.7	2.0	2.8	3.2	1.3	1.0	0.8	0.08	
Rhabdomyosarcoma	105	33	13	9	11	21	18	0.4	0.1	0.1	0.1	0.2	0.3	0.2	0.2	0.2	0.01	
Peripheral nerve sheath tumours	143	4	13	34	25	40	27	0.0	0.1	0.3	0.2	0.5	0.4	0.3	0.2	0.2	0.02	
Other tumours of uncertain differentiation	255	24	33	38	61	51	48	0.3	0.3	0.3	0.5	0.6	0.8	0.4	0.4	0.4	0.03	
Synovial sarcoma	84	3	16	18	25	14	8	0.0	0.2	0.2	0.2	0.2	0.1	0.1	0.1	0.1	0.01	
Myoepithelioma	45	1	2	4	9	15	14	0.0	0.0	0.0	0.1	0.2	0.2	0.1	0.1	0.0	0.00	
Rhabdoid tumours	15	15	-	-	-	-	-	0.2	-	-	-	-	-	0.0	0.0	0.1	0.00	
GIST	1,362	-	9	81	312	542	418	-	0.1	0.7	2.7	6.2	6.9	2.4	1.7	1.2	0.15	
Endometrial stromal sarcoma	238	-	3	38	96	59	42	-	0.0	0.3	0.8	0.7	0.7	0.4	0.4	0.3	0.03	
Ewing sarcoma	106	38	26	15	13	12	2	0.4	0.3	0.1	0.1	0.1	0.0	0.2	0.2	0.2	0.02	
Chondrosarcoma	221	3	20	60	72	41	25	0.0	0.2	0.5	0.6	0.5	0.4	0.4	0.4	0.3	0.03	
Osteosarcoma	169	32	41	14	25	38	19	0.3	0.4	0.1	0.2	0.4	0.3	0.3	0.3	0.3	0.02	
Other bone tumours of uncertain differentiation	65	7	7	4	18	20	9	0.1	0.1	0.0	0.2	0.2	0.1	0.1	0.1	0.1	0.01	
Chordoma	55	4	2	4	16	20	9	0.0	0.0	0.0	0.1	0.2	0.1	0.1	0.1	0.1	0.01	
Unclassified and poorly characterised sarcoma	422	8	9	17	66	133	189	0.1	0.1	0.2	0.6	1.5	3.1	0.7	0.5	0.3	0.04	
All sarcomas	5,389	166	276	595	1,316	1,625	1,411	1.8	2.7	5.4	11.3	18.6	23.3	9.5	7.5	6.0	0.60	

CR: crude (all ages) incidence rate (N/100,000 person years)

ESR and WSR: age-standardised incidence using the European or World Standard Population (N/100,000 person years)

CRI: Cumulative risk 0-74 years (%)

APPENDIX III

Flemish region: Number of new diagnoses (N), age-standardised incidence (N/100,000) of bone and soft tissue tumours in 2010-2019 by histological subtype in males																	
	Number of new diagnoses (N)					Age specific incidence (N/100,000)					CR			WSR	CRI		
	Total	0-14y	15-29y	30-44y	45-59y	60-74y	75+	0-14y	15-29y	30-44y	45-59y	60-74y	75+			CR	ESR
Sarcomas classified by primary tumour location																	
Soft tissue and visceral sarcoma	3,132	71	93	306	694	1,067	901	1.3	1.6	4.9	9.9	21.4	36.9	9.9	7.6	5.7	0.60
Bone sarcoma	352	36	93	46	56	68	53	0.7	1.6	0.7	0.8	1.4	2.2	1.1	1.1	1.0	0.08
Sarcomas classified by histological type																	
Liposarcoma	547	1	8	53	136	219	130	0.0	0.1	0.8	1.9	4.4	5.3	1.7	1.3	1.0	0.11
Liposarcoma well differentiated and atypical lipomatous tumours	210	-	1	16	62	89	42	-	0.0	0.3	0.9	1.8	1.7	0.7	0.5	0.4	0.04
Dedifferentiated liposarcoma	219	-	1	9	47	90	72	-	0.0	0.1	0.7	1.8	2.9	0.7	0.5	0.3	0.04
Myxoid liposarcoma	81	1	5	24	20	24	7	0.0	0.1	0.4	0.3	0.5	0.3	0.3	0.2	0.2	0.02
Liposarcoma NOS and other	37	-	1	4	7	16	9	-	0.0	0.1	0.1	0.3	0.4	0.1	0.1	0.1	0.01
Malignant (myo)fibroblastic tumours and so-called fibrohistiocytic tumours	394	6	25	78	98	105	82	0.1	0.4	1.2	1.4	2.1	3.4	1.2	1.0	0.8	0.08
Dermatofibrosarcoma protuberans	165	3	17	57	44	29	15	0.1	0.3	0.9	0.6	0.6	0.6	0.5	0.5	0.4	0.04
Solitary fibrous tumour	47	-	1	3	10	23	10	-	0.0	0.0	0.1	0.5	0.4	0.1	0.1	0.1	0.01
Fibrosarcoma	20	-	1	3	7	5	4	-	0.0	0.0	0.1	0.1	0.2	0.1	0.1	0.0	0.00
Myxofibrosarcoma	145	3	5	8	34	46	49	0.1	0.1	0.1	0.5	0.9	2.0	0.5	0.3	0.2	0.03
Miscellaneous malignant (myo)fibroblastic tumours and so-called fibrohistiocytic tumours	17	-	1	7	3	2	4	-	0.0	0.1	0.0	0.0	0.2	0.1	0.0	0.0	0.00
Vascular sarcoma	299	1	8	51	76	80	83	0.0	0.1	0.8	1.1	1.6	3.4	0.9	0.7	0.6	0.06
Kaposi sarcoma	155	-	6	40	51	31	27	-	0.1	0.6	0.7	0.6	1.1	0.5	0.4	0.3	0.03
Angiosarcoma	123	1	-	5	21	43	53	0.0	-	0.1	0.3	0.9	2.2	0.4	0.3	0.2	0.02
Leiomyosarcoma	292	-	4	16	55	99	118	-	0.1	0.3	0.8	2.0	4.8	0.9	0.7	0.4	0.05
Rhabdomyosarcoma	72	32	10	2	4	14	10	0.6	0.2	0.0	0.1	0.3	0.4	0.2	0.2	0.3	0.02
Peripheral nerve sheath tumours	77	2	3	15	23	19	15	0.0	0.1	0.2	0.3	0.4	0.6	0.2	0.2	0.2	0.02
Other tumours of uncertain differentiation	161	21	17	40	29	34	20	0.4	0.3	0.6	0.4	0.7	0.8	0.5	0.5	0.5	0.04
Synovial sarcoma	59	4	6	20	12	14	3	0.1	0.1	0.3	0.2	0.3	0.1	0.2	0.2	0.2	0.01
Myoepithelioma	25	1	1	3	5	7	8	0.0	0.0	0.0	0.1	0.1	0.3	0.1	0.1	0.0	0.00
Rhabdoid tumours	13	11	1	1	-	-	-	0.2	0.0	0.0	-	-	-	0.0	0.1	0.1	0.00
GIST	859	1	6	33	206	358	255	0.0	0.1	0.5	2.9	7.2	10.4	2.7	2.0	1.4	0.17
Endometrial stromal sarcoma	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-
Ewing sarcoma	74	21	28	12	4	5	4	0.4	0.5	0.2	0.1	0.1	0.2	0.2	0.3	0.3	0.02
Chondrosarcoma	138	-	14	22	35	46	21	-	0.2	0.4	0.5	0.9	0.9	0.4	0.4	0.3	0.03
Osteosarcoma	144	19	51	11	19	22	22	0.4	0.9	0.2	0.3	0.4	0.9	0.5	0.4	0.5	0.03
Other bone tumours of uncertain differentiation	42	-	5	4	10	14	9	-	0.1	0.1	0.1	0.3	0.4	0.1	0.1	0.1	0.01
Chordoma	39	-	3	3	10	14	9	-	0.1	0.0	0.1	0.3	0.4	0.1	0.1	0.1	0.01
Unclassified and poorly characterised sarcoma	385	3	7	15	55	120	185	0.1	0.1	0.2	0.8	2.4	7.6	1.2	0.8	0.6	0.06
All sarcomas	3,484	107	186	352	750	1,135	954	2.0	3.3	5.6	10.7	22.8	39.0	11.0	8.7	6.8	0.67

CR: crude (all ages) incidence rate (N/100,000 person years)

ESR and WSR: age-standardised incidence using the European or World Standard Population (N/100,000 person years)

CRI: Cumulative risk 0-74 years (%)

Flemish region: Number of new diagnoses (N), age-specific and age-standardised incidence (N/100,000) of bone and soft tissue tumours in 2010-2019 by histological subtype in females

	Number of new diagnoses (N)						Age specific incidence (N/100,000)					CR	ESR	WSR	CRI		
	Total						75+										
	0-14y	15-29y	30-44y	45-59y	60-74y	75+	0-14y	15-29y	30-44y	45-59y	60-74y					75+	
Sarcomas classified by primary tumour location																	
Soft tissue and visceral sarcoma	2,925	54	111	278	719	928	835	1.1	2.0	4.5	10.5	17.8	22.9	9.0	6.7	5.2	0.54
Bone sarcoma	285	47	55	48	56	46	33	0.9	1.0	0.8	0.8	0.9	0.9	0.9	0.9	0.9	0.07
Sarcomas classified by histological type																	
Liposarcoma	320	-	5	30	110	94	81	-	0.1	0.5	1.6	1.8	2.2	1.0	0.7	0.6	0.06
Liposarcoma well differentiated and atypical lipomatous tumours	127	-	1	10	51	44	21	-	0.0	0.2	0.7	0.8	0.6	0.4	0.3	0.2	0.03
Dedifferentiated liposarcoma	118	-	1	5	35	35	42	-	0.0	0.1	0.5	0.7	1.2	0.4	0.2	0.2	0.02
Myxoid liposarcoma	54	-	3	14	18	9	10	-	0.1	0.2	0.3	0.2	0.3	0.2	0.1	0.1	0.01
Liposarcoma NOS and other	21	-	-	1	6	6	8	-	-	0.0	0.1	0.1	0.2	0.1	0.0	0.0	0.00
Malignant (myo)fibroblastic tumours and so-called fibrohistiocytic tumours	380	8	46	75	104	77	70	0.2	0.8	1.2	1.5	1.5	1.9	1.2	1.0	0.9	0.08
Dermatofibrosarcoma protuberans	206	5	37	64	71	18	11	0.1	0.7	1.0	1.0	0.3	0.3	0.6	0.6	0.6	0.05
Solitary fibrous tumour	42	-	2	4	10	15	11	-	0.0	0.1	0.1	0.3	0.3	0.1	0.1	0.1	0.01
Fibrosarcoma	17	2	1	-	3	3	8	0.0	0.0	-	0.0	0.1	0.2	0.1	0.0	0.0	0.00
Myxofibrosarcoma	104	-	6	6	17	38	37	-	0.1	0.1	0.2	0.7	1.0	0.3	0.2	0.2	0.02
Miscellaneous malignant (myo)fibroblastic tumours and so-called fibrohistiocytic tumours	11	1	-	1	3	3	3	0.0	-	0.0	0.0	0.1	0.1	0.0	0.0	0.0	0.00
Vascular sarcoma	236	1	5	21	29	83	97	0.0	0.1	0.3	0.4	1.6	2.7	0.7	0.5	0.3	0.04
Kaposi sarcoma	34	-	2	6	5	7	14	-	0.0	0.1	0.1	0.1	0.4	0.1	0.1	0.1	0.00
Angiosarcoma	181	1	1	8	20	70	81	0.0	0.0	0.1	0.3	1.3	2.2	0.6	0.3	0.2	0.03
Leiomyosarcoma	439	-	4	36	138	146	115	-	0.1	0.6	2.0	2.8	3.2	1.3	1.0	0.7	0.08
Rhabdomyosarcoma	66	20	8	6	8	16	8	0.4	0.1	0.1	0.1	0.3	0.2	0.2	0.2	0.2	0.02
Peripheral nerve sheath tumours	91	2	6	19	18	23	23	0.0	0.1	0.3	0.3	0.4	0.6	0.3	0.2	0.2	0.02
Other tumours of uncertain differentiation	148	14	19	21	38	26	30	0.3	0.3	0.3	0.6	0.5	0.8	0.5	0.4	0.4	0.03
Synovial sarcoma	51	1	9	12	18	5	6	0.0	0.2	0.2	0.3	0.1	0.2	0.2	0.1	0.1	0.01
Myoepithelioma	29	1	1	4	5	10	8	0.0	0.0	0.1	0.1	0.2	0.2	0.1	0.1	0.1	0.01
Rhabdoid tumours	8	8	-	-	-	-	-	0.2	-	-	-	-	-	0.0	0.0	0.1	-
GIST	811	-	6	42	168	327	268	-	0.1	0.7	2.4	6.3	7.4	2.5	1.7	1.2	0.15
Endometrial stromal sarcoma	118	-	-	18	49	31	20	-	-	0.3	0.7	0.6	0.5	0.4	0.3	0.2	0.02
Ewing sarcoma	54	17	17	5	8	6	1	0.3	0.3	0.1	0.1	0.1	0.0	0.2	0.2	0.2	0.01
Chondrosarcoma	133	3	15	37	40	25	13	0.1	0.3	0.6	0.6	0.5	0.4	0.4	0.4	0.3	0.03
Osteosarcoma	105	26	24	7	13	22	13	0.5	0.4	0.1	0.2	0.4	0.4	0.3	0.3	0.3	0.03
Other bone tumours of uncertain differentiation	41	7	5	4	10	9	6	0.1	0.1	0.1	0.1	0.2	0.2	0.1	0.1	0.1	0.01
Chordoma	31	4	-	4	8	9	6	0.1	-	0.1	0.1	0.2	0.2	0.1	0.1	0.1	0.01
Unclassified and poorly characterised sarcoma	268	3	6	5	42	89	123	0.1	0.1	0.1	0.6	1.7	3.4	0.8	0.5	0.4	0.04
All sarcomas	3,210	101	166	326	775	974	868	2.0	3.0	5.3	11.3	18.7	23.8	9.9	7.6	6.1	0.61

CR: crude (all ages) incidence rate (N/100,000 person years)

ESR and WSR: age-standardised incidence using the European or World Standard Population (N/100,000 person years)

CRI: Cumulative risk 0-74 years (%)

Walloon region: Number of new diagnoses (N), age-specific and age-standardised incidence (N/100,000) of bone and soft tissue tumours in 2010-2019 by histological subtype in males

	Number of new diagnoses (N)					Age specific incidence (N/100,000)					CR	ESR	WSR	CRI			
	Total					75+											
	0-14y	15-29y	30-44y	45-59y	60-74y	0-14y	15-29y	30-44y	45-59y	60-74y					75+		
Sarcomas classified by primary tumour location																	
Soft tissue and visceral sarcoma	1,484	39	64	159	363	540	319	1.2	1.9	4.6	9.8	21.1	28.9	8.5	7.3	5.6	0.59
Bone sarcoma	175	26	39	28	28	38	16	0.8	1.2	0.8	0.8	1.5	1.5	1.0	1.0	0.9	0.07
Sarcomas classified by histological type																	
Liposarcoma	220	2	3	23	60	96	36	0.1	0.1	0.7	1.6	3.7	3.3	1.3	1.1	0.8	0.09
Liposarcoma well differentiated and atypical lipomatous tumours	71	-	-	6	18	32	15	-	-	0.2	0.5	1.2	1.4	0.4	0.3	0.2	0.03
Dedifferentiated liposarcoma	84	-	2	3	25	40	14	-	0.1	0.1	0.7	1.6	1.3	0.5	0.4	0.3	0.04
Myxoid liposarcoma	41	2	1	10	11	12	5	0.1	0.0	0.3	0.3	0.5	0.5	0.2	0.2	0.2	0.02
Liposarcoma NOS and other	24	-	-	4	6	12	2	-	-	0.1	0.2	0.5	0.2	0.1	0.1	0.1	0.01
Malignant (myo)fibroblastic tumours and so-called fibrohistiocytic tumours	170	3	14	38	40	42	33	0.1	0.4	1.1	1.1	1.6	3.0	1.0	0.9	0.7	0.06
Dermatofibrosarcoma protuberans	79	2	12	31	17	10	7	0.1	0.4	0.9	0.5	0.4	0.6	0.5	0.4	0.4	0.03
Solitary fibrous tumour	30	-	1	2	6	11	10	-	0.0	0.1	0.2	0.4	0.9	0.2	0.1	0.1	0.01
Fibrosarcoma	8	-	-	-	4	2	2	-	-	-	0.1	0.1	0.2	0.0	0.0	0.0	0.00
Myxofibrosarcoma	46	-	-	5	11	17	13	-	-	0.1	0.3	0.7	1.2	0.3	0.2	0.2	0.02
Miscellaneous malignant (myo)fibroblastic tumours and so-called fibrohistiocytic tumours	7	1	1	-	2	2	1	0.0	0.0	-	0.1	0.1	0.1	0.0	0.0	0.0	0.00
Vascular sarcoma	150	1	6	21	36	56	30	0.0	0.2	0.6	1.0	2.2	2.7	0.9	0.7	0.5	0.06
Kaposi sarcoma	95	-	5	18	24	29	19	-	0.1	0.5	0.6	1.1	1.7	0.5	0.5	0.4	0.04
Angiosarcoma	52	1	1	2	11	26	11	0.0	0.0	0.1	0.3	1.0	1.0	0.3	0.2	0.2	0.02
Leiomyosarcoma	138	1	4	8	31	46	48	0.0	0.1	0.2	0.8	1.8	4.4	0.8	0.6	0.4	0.05
Rhabdomyosarcoma	51	19	12	4	6	3	7	0.6	0.4	0.1	0.2	0.1	0.6	0.3	0.3	0.4	0.02
Periphereal nerve sheath tumours	47	-	3	15	14	8	7	-	0.1	0.4	0.4	0.3	0.6	0.3	0.2	0.2	0.02
Other tumours of uncertain differentiation	76	12	11	11	16	20	6	0.4	0.3	0.3	0.4	0.8	0.5	0.4	0.4	0.4	0.03
Synovial sarcoma	28	2	4	3	6	10	3	0.1	0.1	0.1	0.2	0.4	0.3	0.2	0.1	0.1	0.01
Myoepithelioma	10	-	2	-	3	4	1	-	0.1	-	0.1	0.2	0.1	0.1	0.1	0.0	0.00
Rhabdoid tumours	6	6	-	-	-	-	-	0.2	-	-	-	-	-	0.0	0.0	0.1	0.00
GIST	478	-	1	29	125	222	101	-	0.0	0.8	3.4	8.7	9.2	2.7	2.2	1.6	0.20
Endometrial stromal sarcoma	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-
Ewing sarcoma	48	12	19	8	6	2	1	0.4	0.6	0.2	0.2	0.1	0.1	0.3	0.3	0.3	0.02
Chondrosarcoma	66	1	4	12	21	20	8	0.0	0.1	0.3	0.6	0.8	0.7	0.4	0.3	0.3	0.03
Osteosarcoma	57	14	19	7	7	8	2	0.4	0.6	0.2	0.2	0.3	0.2	0.3	0.3	0.4	0.03
Other bone tumours of uncertain differentiation	23	-	2	3	3	14	1	-	0.1	0.1	0.1	0.5	0.1	0.1	0.1	0.1	0.01
Chordoma	22	-	2	2	3	14	1	-	0.1	0.1	0.1	0.5	0.1	0.1	0.1	0.1	0.01
Unclassified and poorly characterised sarcoma	135	-	5	8	26	41	55	-	0.1	0.2	0.7	1.6	5.0	0.8	0.6	0.4	0.04
All sarcomas	1,659	65	103	187	391	578	335	2.0	3.0	5.4	10.5	22.6	30.4	9.5	8.2	6.5	0.66

CR: crude (all ages) incidence rate (N/100,000 person years)

ESR and WSR: age-standardised incidence using the European or World Standard Population (N/100,000 person years)

CRI: Cumulative risk 0-74 years (%)

Walloon region: Number of new diagnoses (N), age-specific and age-standardised incidence (N/100,000) of bone and soft tissue tumours in 2010-2019 by histological subtype in females

	Number of new diagnoses (N)						Age specific incidence (N/100,000)					CR	ESR	WSR	CRI		
	Total						75+										
	0-14y	15-29y	30-44y	45-59y	60-74y	75+	0-14y	15-29y	30-44y	45-59y	60-74y					75+	
Sarcomas classified by primary tumour location																	
Soft tissue and visceral sarcoma	1,487	32	59	163	359	472	402	1.0	1.8	4.7	9.5	16.5	21.1	8.1	6.3	4.9	0.50
Bone sarcoma	152	15	22	30	32	34	19	0.5	0.7	0.9	0.8	1.2	1.0	0.8	0.8	0.7	0.06
Sarcomas classified by histological type																	
Liposarcoma	146	-	4	9	44	47	42	-	0.1	0.3	1.2	1.6	2.2	0.8	0.6	0.4	0.05
Liposarcoma well differentiated and atypical lipomatous tumours	50	-	-	1	14	22	13	-	-	0.0	0.4	0.8	0.7	0.3	0.2	0.1	0.02
Dedifferentiated liposarcoma	54	-	-	-	18	14	22	-	-	-	0.5	0.5	1.2	0.3	0.2	0.1	0.01
Myxoid liposarcoma	28	-	4	6	9	7	2	-	0.1	0.2	0.2	0.2	0.1	0.2	0.1	0.1	0.01
Liposarcoma NOS and other	14	-	-	2	3	4	5	-	-	0.1	0.1	0.1	0.3	0.1	0.1	0.0	0.00
Malignant (myo)fibroblastic tumours and so-called fibrohistiocytic tumours	172	4	19	39	39	42	29	0.1	0.6	1.1	1.0	1.5	1.5	0.9	0.8	0.7	0.07
Dermatofibrosarcoma protuberans	88	1	14	32	22	14	5	0.0	0.4	0.9	0.6	0.5	0.3	0.5	0.5	0.4	0.04
Solitary fibrous tumour	25	-	1	1	8	8	7	-	0.0	0.0	0.2	0.3	0.4	0.1	0.1	0.1	0.01
Fibrosarcoma	9	-	3	-	2	2	2	-	0.1	-	0.1	0.1	0.1	0.0	0.0	0.0	0.00
Myxofibrosarcoma	40	3	1	5	4	14	13	0.1	0.0	0.1	0.1	0.5	0.7	0.2	0.2	0.1	0.01
Miscellaneous malignant (myo)fibroblastic tumours and so-called fibrohistiocytic tumours	10	-	-	1	3	4	2	-	-	0.0	0.1	0.1	0.1	0.1	0.0	0.0	-
Vascular sarcoma	121	-	3	10	19	34	55	-	0.1	0.3	0.5	1.2	2.9	0.7	0.4	0.3	0.03
Kaposi sarcoma	35	-	1	6	5	9	14	-	0.0	0.2	0.1	0.3	0.7	0.2	0.1	0.1	0.01
Angiosarcoma	79	-	2	3	12	23	39	-	0.1	0.1	0.3	0.8	2.0	0.4	0.3	0.2	0.02
Leiomyosarcoma	251	1	2	23	77	77	71	0.0	0.1	0.7	2.0	2.7	3.7	1.4	1.0	0.8	0.08
Rhabdomyosarcoma	29	9	2	2	2	5	9	0.3	0.1	0.1	0.1	0.2	0.5	0.2	0.1	0.2	0.01
Periphereal nerve sheath tumours	43	1	5	13	4	16	4	0.0	0.2	0.4	0.1	0.6	0.2	0.2	0.2	0.2	0.02
Other tumours of uncertain differentiation	84	9	12	14	17	20	12	0.3	0.4	0.4	0.5	0.7	0.6	0.5	0.4	0.4	0.03
Synovial sarcoma	27	2	5	5	5	8	2	0.1	0.2	0.1	0.1	0.3	0.1	0.1	0.1	0.1	0.01
Myoepithelioma	12	-	1	-	4	4	3	-	0.0	-	0.1	0.1	0.2	0.1	0.0	0.0	0.00
Rhabdoid tumours	6	6	-	-	-	-	-	0.2	-	-	-	-	-	0.0	0.0	0.1	0.00
GIST	414	-	3	29	100	170	112	-	0.1	0.8	2.7	6.0	5.9	2.3	1.7	1.2	0.14
Endometrial stromal sarcoma	97	-	2	15	38	23	19	-	0.1	0.4	1.0	0.8	1.0	0.5	0.4	0.3	0.03
Ewing sarcoma	36	15	7	9	1	4	-	0.5	0.2	0.3	0.0	0.1	-	0.2	0.2	0.3	0.02
Chondrosarcoma	62	-	5	19	15	13	10	-	0.2	0.5	0.4	0.5	0.5	0.3	0.3	0.3	0.02
Osteosarcoma	45	4	12	4	8	13	4	0.1	0.4	0.1	0.2	0.5	0.2	0.2	0.2	0.2	0.02
Other bone tumours of uncertain differentiation	19	-	2	-	7	8	2	-	0.1	-	0.2	0.3	0.1	0.1	0.1	0.1	0.01
Chordoma	19	-	2	-	7	8	2	-	0.1	-	0.2	0.3	0.1	0.1	0.1	0.1	0.01
Unclassified and poorly characterised sarcoma	120	4	3	7	20	34	52	0.1	0.1	0.2	0.5	1.2	2.7	0.7	0.4	0.3	0.03
All sarcomas	1,639	47	81	193	391	506	421	1.5	2.5	5.6	10.4	17.7	22.0	8.9	7.1	5.6	0.56

CR: crude (all ages) incidence rate (N/100,000 person years)

ESR and WSR: age-standardised incidence using the European or World Standard Population (N/100,000 person years)

CRI: Cumulative risk 0-74 years (%)

Belgium: Incidence (2010-2019), 5-year prevalence (31/12/2019) and 5-year relative survival (2004-2019) of bone and soft tissue tumours by histological subtype

	Males and females								
	Incidence (2010-2019)			Prevalence (5 years) 31/12/2019			5-year Relative survival (2004-2019)		
	N	CR	WSR	N	CR	WSR	N at risk	%	95%CI
Sarcomas classified by primary tumour location									
Soft tissue and visceral sarcoma	10,034	9.0	5.4	3,988	34.7	20.3	14,787	72.4	[71.5:73.3]
Bone sarcoma	1,104	1.0	0.9	399	3.5	3.2	1,772	70.2	[67.8:72.6]
Sarcomas classified by histological type									
Liposarcoma	1,355	1.2	0.7	589	5.1	2.8	1,987	77.6	[75.1:80.0]
Liposarcoma well differentiated and atypical lipomatous tumours	494	0.4	0.3	279	2.4	1.3	726	99.1	[95.9:101.8]
Dedifferentiated liposarcoma	532	0.5	0.2	179	1.6	0.8	720	57.0	[52.4:61.5]
Myxoid liposarcoma	222	0.2	0.1	94	0.8	0.6	360	84.3	[79.0:88.7]
Liposarcoma NOS and other	107	0.1	0.1	37	0.3	0.2	182	58.3	[49.0:67.0]
Malignant (myo)fibroblastic tumours and so-called fibrohistiocytic tumours	1,253	1.1	0.8	541	4.7	3.3	1,963	88.2	[86.1:90.1]
Dermatofibrosarcoma protuberans	618	0.6	0.5	289	2.5	2.1	972	98.3	[96.4:99.8]
Solitary fibrous tumour	158	0.1	0.1	64	0.6	0.3	210	71.4	[63.0:78.7]
Fibrosarcoma	58	0.1	0.0	11	0.1	0.1	130	65.4	[54.8:74.8]
Myxofibrosarcoma	365	0.3	0.2	143	1.2	0.6	567	82.2	[77.3:86.6]
Miscellaneous malignant (myo)fibroblastic tumours and so-called fibrohistiocytic tumours	54	0.0	0.0	34	0.3	0.2	85	81.6	[68.3:91.5]
Vascular sarcoma	994	0.9	0.5	300	2.6	1.5	1,451	56.7	[53.5:59.7]
Kaposi sarcoma	458	0.4	0.3	177	1.5	1.0	672	81.1	[76.8:85.1]
Angiosarcoma	481	0.4	0.2	106	0.9	0.4	695	33.4	[29.3:37.6]
Leiomyosarcoma	1,229	1.1	0.6	401	3.5	1.9	2,030	57.3	[54.7:59.9]
Rhabdomyosarcoma	238	0.2	0.3	78	0.7	0.8	362	47.9	[42.2:53.4]
Peripheral nerve sheath tumours	271	0.2	0.2	86	0.7	0.5	418	56.0	[50.4:61.4]
Other tumours of uncertain differentiation	515	0.5	0.4	175	1.5	1.3	815	60.2	[56.3:64.0]
Synovial sarcoma	180	0.2	0.1	60	0.5	0.4	310	63.3	[57.1:68.9]
Myoepithelioma	82	0.1	0.0	22	0.2	0.1	126	67.8	[56.7:77.6]
Rhabdoid tumours	38	0.0	0.1	9	0.1	0.1	<50	-	-
GIST	2,826	2.5	1.3	1,357	11.8	5.9	3,802	91.9	[90.3:93.5]
Endometrial stromal sarcoma	238	0.2	0.1	97	0.8	0.6	374	70.0	[64.5:75.0]
Ewing sarcoma	245	0.2	0.3	83	0.7	0.9	394	59.7	[54.4:64.6]
Chondrosarcoma	454	0.4	0.3	169	1.5	1.1	739	80.6	[77.0:83.9]
Osteosarcoma	391	0.4	0.4	137	1.2	1.2	567	60.5	[56.0:64.8]
Other bone tumours of uncertain differentiation	141	0.1	0.1	67	0.6	0.4	204	82.2	[74.4:88.6]
Chordoma	125	0.1	0.1	60	0.5	0.3	184	80.4	[71.8:87.5]
Unclassified and poorly characterised sarcoma	988	0.9	0.4	310	2.7	1.3	1,501	48.0	[44.7:51.1]
All sarcomas	11,138	10.0	6.4	4,385	38.2	23.4	16,546	72.2	[71.3:73.0]

CR: Crude (all ages) rate (N/100,000 person years)

WSR: age-standardised rate, using the world population (N/100,000 person years)

Relative survival calculated for all cases diagnosed between 2004 and 2019

APPENDIX V

NUMBER OF NEW DIAGNOSES AND AGE-STANDARDISED INCIDENCE OF BONE AND SOFT TISSUE
TUMOURS BY HISTOLOGICAL SUBTYPE, SEX AND INCIDENCE YEAR (2004-2019)

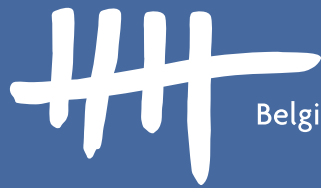
APPENDIX VI

Belgium: 5-year relative survival trends in males and female of bone and soft tissue tumours by cohort and histological subtype

	Males and females								
	N at risk			5-yr RS			95% CI		
	2004-2009	2009-2014	2014-2019	2004-2009	2009-2014	2014-2019	2004-2009	2009-2014	2014-2019
Sarcomas classified by primary tumour location									
Soft tissue and visceral sarcoma	4,890	5,458	6,359	70.1	72.0	74.3	[68.5:71.6]	[70.5:73.4]	[72.7:75.8]
Bone sarcoma	681	648	638	67.4	72.5	72.2	[63.4:71.1]	[68.6:76.1]	[67.7:76.3]
Sarcomas classified by histological type									
Liposarcoma	639	695	904	76.7	75.2	77.8	[72.3:80.8]	[71.1:79.1]	[73.6:81.7]
Liposarcoma well differentiated and atypical lipomatous tumours	233	218	344	95.8	96.5	102.0	[89.5:100.7]	[90.4:101.1]	[96.3:105.8]
Dedifferentiated liposarcoma	193	274	353	56.6	59.8	53.5	[48.1:64.8]	[52.6:66.7]	[46.3:60.6]
Myxoid liposarcoma	138	138	138	82.5	82.7	86.6	[73.6:89.4]	[73.9:89.5]	[77.5:93.1]
Liposarcoma NOS and other	75	65	70	56.6	51.7	63.5	[42.8:69.3]	[37.7:64.6]	[45.4:78.6]
Malignant (myo)fibroblastic tumours and so-called fibrohistiocytic tumours	725	712	748	87.6	88.8	88.3	[84.2:90.7]	[85.4:91.8]	[84.4:91.7]
Dermatofibrosarcoma protuberans	361	357	362	95.7	100.4	100.6	[92.1:98.3]	[97.6:102.3]	[97.6:102.4]
Solitary fibrous tumour	56	77	101	72.7	66.1	68.6	[56.8:85.2]	[52.7:77.6]	[54.8:80.4]
Fibrosarcoma	73	<50	<50	69.5	-	-	[55.2:81.5]	-	-
Myxofibrosarcoma	204	214	218	84.0	82.9	79.4	[76.1:90.6]	[75.3:89.5]	[69.8:87.7]
Miscellaneous malignant (myo)fibroblastic tumours and so-called fibrohistiocytic tumours	<50	<50	<50	-	-	-	-	-	-
Vascular sarcoma	477	576	612	58.0	55.6	58.1	[52.7:63.1]	[50.8:60.3]	[53.0:63.1]
Kaposi sarcoma	223	278	276	77.0	83.8	86.0	[69.4:83.6]	[77.4:89.2]	[78.8:92.1]
Angiosarcoma	224	271	298	39.4	28.0	31.4	[32.1:46.7]	[22.2:34.2]	[24.7:38.6]
Leiomyosarcoma	817	730	738	54.4	58.2	58.2	[50.4:58.3]	[53.9:62.4]	[53.3:62.9]
Rhabdomyosarcoma	124	131	151	49.2	44.0	47.8	[39.8:57.9]	[35.1:52.6]	[37.5:57.6]
Peripheral nerve sheath tumours	149	156	168	54.5	58.3	60.9	[45.4:63.0]	[49.4:66.4]	[51.4:69.4]
Other tumours of uncertain differentiation	304	300	310	59.2	60.8	60.6	[53.0:65.1]	[54.5:66.6]	[53.5:67.1]
Synovial sarcoma	132	121	96	61.2	64.4	68.2	[51.9:69.4]	[54.8:72.6]	[56.2:77.9]
Myoepithelioma	<50	56	<50	-	69.2	-	-	[52.5:83.0]	-
Rhabdoid tumours	<50	<50	<50	-	-	-	-	-	-
GIST	995	1,460	1,854	93.3	90.7	91.7	[90.1:96.1]	[88.1:93.1]	[89.0:94.2]
Endometrial stromal sarcoma	136	146	151	64.3	66.2	75.8	[55.0:72.4]	[57.1:74.1]	[66.5:83.3]
Ewing sarcoma	151	152	138	56.7	59.8	63.8	[48.3:64.3]	[51.4:67.4]	[53.5:72.6]
Chondrosarcoma	286	283	256	77.5	84.1	82.1	[71.5:82.7]	[78.6:88.7]	[75.2:87.7]
Osteosarcoma	186	208	238	58.6	63.6	59.9	[50.8:65.6]	[56.3:70.1]	[52.2:67.0]
Other bone tumours of uncertain differentiation	65	68	86	79.1	83.0	85.9	[64.7:89.9]	[70.3:91.7]	[72.2:95.2]
Chordoma	61	59	76	79.2	80.2	83.2	[64.2:90.5]	[65.9:90.2]	[67.2:94.3]
Unclassified and poorly characterised sarcoma	521	502	654	45.2	45.6	52.4	[40.2:50.2]	[40.4:50.8]	[46.8:58.0]
All sarcomas	5,570	6,104	6,993	69.7	72.1	74.1	[68.3:71.2]	[70.7:73.4]	[72.6:75.5]

Belgium: 5-year relative survival trends of bone and soft tissue tumours by cohort, sex and histological subtype

	Males										Females									
	N at risk		5-yr RS		95% CI		N at risk		5-yr RS		95% CI		N at risk		5-yr RS		95% CI			
	2004-2009	2014-2019	2004-2009	2014-2019	2004-2009	2014-2019	2004-2009	2014-2019	2004-2009	2014-2019	2004-2009	2014-2019	2004-2009	2014-2019	2004-2009	2014-2019	2004-2009	2014-2019		
Sarcomas classified by primary tumour location																				
Soft tissue and visceral sarcoma	2,407	2,738	3,284	72.0	72.3	74.7	[69.6;74.2]	[70.1;74.4]	[72.5;76.9]	2,483	2,720	3,075	68.3	71.7	73.8	[66.1;70.4]	[69.6;73.7]	[71.7;75.9]		
Bone sarcoma	350	338	377	63.1	66.0	68.3	[57.4;68.4]	[60.3;71.3]	[62.2;73.8]	331	310	261	71.9	79.5	77.8	[66.3;76.8]	[74.1;84.0]	[71.0;83.4]		
Sarcomas classified by histological type																				
Liposarcoma	362	418	561	76.8	75.0	77.2	[70.8;82.3]	[69.5;80.0]	[71.4;82.4]	277	277	343	76.5	75.6	78.5	[69.9;82.3]	[69.0;81.4]	[72.1;84.0]		
Liposarcoma well differentiated and atypical lipomatous tumours	128	129	207	97.3	97.5	101.0	[88.1;103.9]	[88.9;103.4]	[91.3;107.0]	105	89	137	94.2	95.0	102.3	[84.5;100.7]	[85.1;101.2]	[95.2;105.9]		
De differentiated liposarcoma	109	171	226	54.7	60.6	53.2	[43.1;65.7]	[51.4;69.2]	[43.7;62.3]	84	103	127	58.8	58.6	54.3	[46.0;70.4]	[46.8;69.5]	[42.7;65.1]		
Myxoid liposarcoma	90	81	86	83.1	76.7	85.8	[71.8;91.5]	[64.1;86.5]	[73.0;94.2]	<50	57	52	-	91.1	87.6	-	[78.1;98.3]	[72.8;96.0]		
Liposarcoma NOS and other	<50	<50	<50	-	-	-	-	-	-	<50	<50	<50	-	-	-	-	-	-		
Malignant (myo)fibroblastic tumours and so-called fibrohistiocytic tumours	345	357	379	84.1	87.6	88.6	[78.5;88.9]	[82.3;92.1]	[82.6;93.6]	380	355	369	90.8	90.1	88.0	[86.4;94.4]	[85.5;93.7]	[82.7;92.4]		
Dermatofibrosarcoma protuberans	168	162	176	92.2	103.1	102.5	[85.5;96.9]	[98.2;105.5]	[97.6;104.8]	193	195	186	98.7	98.3	98.9	[94.6;100.9]	[94.1;100.5]	[94.3;101.2]		
Solitary fibrous tumour	<50	<50	51	-	-	68.7	-	-	[48.6;85.2]	<50	<50	50	-	-	68.8	-	-	[48.9;84.0]		
Fibrosarcoma	<50	<50	<50	-	-	-	-	-	-	<50	<50	<50	-	-	-	-	-	-		
Myxofibrosarcoma	100	119	125	80.0	78.8	79.2	[67.6;90.1]	[67.6;88.2]	[65.6;90.5]	104	95	93	87.8	87.9	80.4	[77.1;95.8]	[76.9;95.8]	[66.0;91.6]		
Miscellaneous malignant (myo)fibroblastic tumours and so-called fibrohistiocytic tumours	<50	<50	<50	-	-	-	-	-	-	<50	<50	<50	-	-	-	-	-	-		
Vascular sarcoma	252	326	353	63.3	61.1	59.1	[56.1;70.0]	[54.7;67.0]	[52.4;65.6]	225	250	259	51.8	48.3	56.7	[44.1;59.4]	[41.0;55.5]	[48.6;64.4]		
Kaposi sarcoma	171	218	216	78.6	84.1	86.5	[70.2;85.6]	[77.1;89.8]	[78.5;93.1]	52	60	60	71.6	82.5	84.2	[53.6;86.4]	[65.7;95.5]	[66.2;97.3]		
Angiosarcoma	68	95	120	30.1	15.1	13.0	[18.7;42.9]	[8.4;24.1]	[6.4;22.3]	156	176	178	43.5	34.9	45.2	[34.6;52.4]	[27.1;43.0]	[35.6;54.7]		
Leiomyosarcoma	301	274	294	59.9	63.6	63.0	[52.9;66.6]	[56.1;70.7]	[54.3;71.1]	516	456	444	51.4	55.1	54.9	[46.5;56.1]	[49.8;60.2]	[49.0;60.6]		
Rhabdomyosarcoma	76	75	86	52.5	42.4	47.3	[40.4;63.4]	[30.9;53.7]	[35.2;58.6]	<50	56	65	-	46.0	45.8	-	[32.2;59.0]	[26.9;63.5]		
Peripheral nerve sheath tumours	87	74	80	64.3	50.5	57.3	[51.9;75.1]	[37.7;62.5]	[44.0;69.0]	62	82	88	41.0	65.0	64.2	[28.2;53.7]	[52.6;75.5]	[50.5;75.7]		
Other tumours of uncertain differentiation	163	154	156	49.9	55.4	59.2	[41.4;58.1]	[46.7;63.4]	[48.8;68.6]	141	146	154	69.8	66.4	61.9	[60.8;77.5]	[57.2;74.6]	[51.9;70.8]		
Synovial sarcoma	72	65	53	49.4	63.9	72.8	[37.1;60.8]	[50.6;74.8]	[56.3;84.7]	60	56	<50	75.5	65.1	-	[61.8;85.4]	[50.5;76.6]	-		
Myoepithelioma	<50	<50	<50	-	-	-	-	-	-	<50	<50	<50	-	-	-	-	-	-		
Rhabdoid tumours	<50	<50	<50	-	-	-	-	-	-	<50	<50	<50	-	-	-	-	-	-		
GIST	534	747	949	92.8	89.0	90.6	[88.2;96.8]	[85.1;92.6]	[86.6;94.2]	461	713	905	93.8	92.5	92.8	[89.4;97.5]	[89.0;95.6]	[89.0;96.0]		
Endometrial stromal sarcoma	-	-	-	-	-	-	-	-	-	136	146	151	64.3	66.2	75.8	[55.0;72.4]	[57.1;74.1]	[66.5;83.3]		
Ewing sarcoma	89	86	84	54.7	58.4	58.5	[43.7;64.5]	[46.9;68.4]	[45.6;69.5]	62	66	54	59.5	61.5	72.8	[46.1;70.7]	[48.6;72.2]	[55.0;85.1]		
Chondrosarcoma	135	136	146	75.7	78.3	82.2	[66.1;83.5]	[69.2;85.6]	[72.5;89.7]	151	147	110	79.1	89.4	81.9	[71.0;85.7]	[82.4;94.3]	[71.4;89.4]		
Osteosarcoma	98	114	141	58.0	58.1	57.0	[47.1;67.6]	[48.0;67.0]	[46.8;66.2]	88	94	97	59.1	70.1	64.2	[47.7;69.1]	[59.2;78.9]	[51.8;74.5]		
Other bone tumours of uncertain differentiation	<50	<50	51	-	-	86.1	-	-	[67.8;97.8]	<50	<50	<50	-	-	-	-	-	-		
Chordoma	<50	<50	<50	-	-	-	-	-	-	<50	<50	<50	-	-	-	-	-	-		
Unclassified and poorly characterised sarcoma	283	286	386	48.9	47.0	54.8	[41.7;56.0]	[39.9;54.0]	[47.2;62.4]	238	216	268	40.9	43.9	49.1	[34.0;47.8]	[36.3;51.5]	[41.1;57.0]		
All sarcomas	2,757	3,074	3,659	70.8	71.6	74.0	[68.7;72.9]	[69.6;73.6]	[71.9;76.1]	2,813	3,030	3,334	68.7	72.5	74.2	[66.7;70.7]	[70.6;74.4]	[72.1;76.1]		



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