# Defining Soft Tissue Sarcomas

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# Overview of Soft Tissue Sarcomas

- What do we already know about soft tissue sarcoma?
  - Very rare cancer – arises in connective tissue
  - Accounts for approximately 1% of all malignant tumours
- Most common sites
  - Limbs
  - Retroperitoneum
  - Gynaecological sarcomas
- The West Midlands Cancer Intelligence Unit is the lead registry in England for bone and soft tissue sarcoma
- Conduct analysis in collaboration with the sarcoma Site Specific Clinical Reference Group (SSCRG)

## The Challenge

How many soft tissue sarcomas are diagnosed in England each year?



## Simple Solution 1

- Just count by ICD-10 site code C49
  - "Malignant neoplasm of other connective and soft tissue"
  - Only 40-50% of cases captured by site code!
- Not site specific
  - Arise anywhere within the body
  - C49 subsites not detailed enough!
    - Sarcomas of the breast will get coded to C50
- Therefore count by morphology code

# Incidence by morphology

- What are we including or counting as a soft tissue morphology?
  - Kaposi's sarcomas?
    - Result of Human Herpes virus
  - Skin tumours – Dermatofibrosarcomas?
    - Non-melanoma skin cancer excluded from cancer stats
  - Tumours of uncertain or unknown behaviour?
  - Mixed tumour types
    - Carcinosarcomas or Mixed Mullerian?



# Simple Solution 2

- Be consistent with published research
  - No existing research for England or UK
- Base on international research!

Simple Solution 2

## International (In)consistency

- Wibmer et al (2009) – Epidemiologic study from Austria
  - Included Kaposi's sarcoma
  - Excluded Dermatofibrosarcoma
  - Further examination also shows that spindle cell sarcomas are not present



Simple Solution 2

International (In)consistency

- Toro et al (2006) – study from USA
  - Include Dermatofibrosarcoma
  - Excluded Kaposi's sarcoma
  - Also excluded
    - Chordoma
    - Mesenchymomas
    - Neuroblastomas
    - Endometrial stromal sarcomas

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Simple Solution 2

## International (In)consistency

- Gustafson (1994) – study from Sweden
  - Omitted patients <16 years of age
    - Effectively removing Rhabdomyosarcomas and other childhood cancers
  - Only soft tissue sarcomas of extremeties and trunk wall
- International variances in classification of soft tissue sarcoma!
- Requirement for comprehensive list of soft tissue sarcomas!

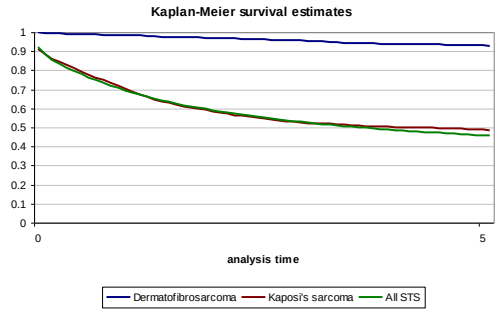


## The Solution – Steps undertaken with the SSCRG

- The WMCIU compiled an initial list of “potential” sarcomas (with the help of a pathologist)
  - This initial compilation included:
    - Dermatofibrosarcoma
    - Kaposi’s sarcoma
    - Carcinosarcomas – and other “mixed” sarcoma types
    - Neuroblastoma/gliosarcoma – and other CNS tumour types
    - Tumours of uncertain behaviour
      - **Neurofibromatosis, Atypical Fibrous Histiocytoma**
    - Cases where the site code and morphology seem unlikely
      - **Chordoma of soft tissue**
    - Excluded bone sarcomas (as they are not soft tissue)

# Differing survival

- In other cancer statistics skin “C44” cases are normally excluded
  - Non-melanoma skin cancer rarely aggressive
- WMCIU calculated survival rates – demonstrate differences in characteristics between Kaposi’s and dermatofibrosarcoma






# Key Decisions

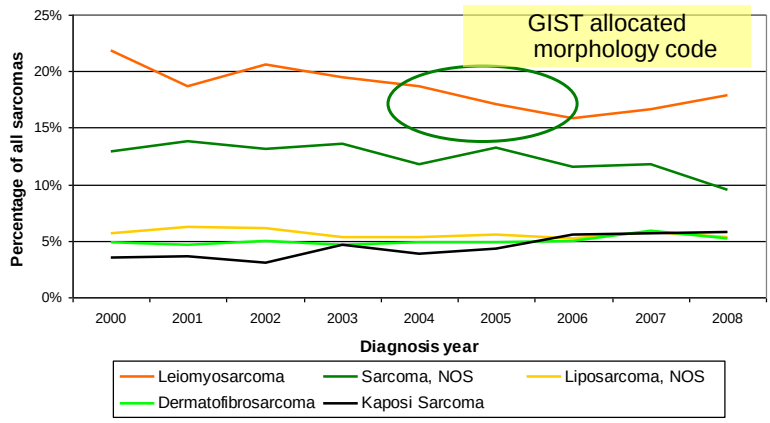
- Dermatofibrosarcomas should be included as they are “bona fide” sarcomas ✓
- Kaposi’s sarcoma should be included ✓
- Carcinosarcomas (and other mixed tumours)
  - Carcinoma with divergent differentiation ✗
- Neuroblastoma/Gliosarcoma
  - Not in WHO classification of soft tissue sarcomas ✗
- Tumours of uncertain behaviour
  - Some are not registerable conditions
  - Better survival – dilute statistics ✗
- Incompatible site code and morphology
  - Include – use as a basis for data quality ✓

# The Results

- The agreed list of soft tissue sarcomas consists of 125 morphology codes which could affect soft tissue
- This list was applied to the latest version of the Merged Cancer Registry dataset (MCR – diagnosis years 1990 – 2008)
  - 45,175 tumours identified
  - 23,700 between 2000 and 2008
  - Only 33 sarcoma types had an average annual incidence >=10 cases



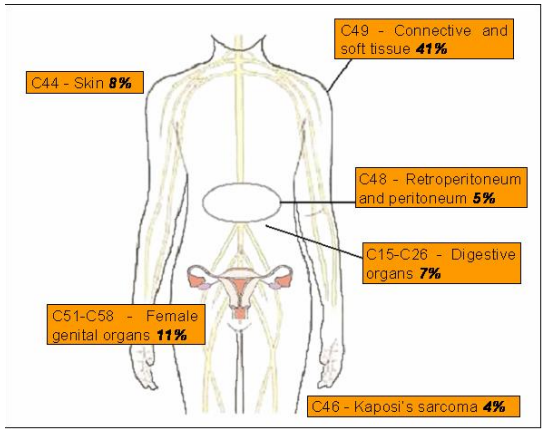
# Most common soft tissue sarcomas 2000-2008




- Leiomyosarcoma most common soft tissue morphology

## Cancer Sub-site Incidence: 2000 - 2008

- Percentage diagnosed by most common cancer site




- Counting C49's only would capture 41% of cases!



## Summary

- Incidence of soft tissue sarcomas cannot be derived from ICD-10 site codes alone
  - They can arise anywhere in the body
  - Must be derived from morphology type alone
- International variation of inclusions and exclusions of soft tissue sarcoma
  - Make international comparisons of incidence impossible
  - Inconsistency of soft tissue sarcoma classification
- A comprehensive list of soft tissue sarcomas compiled by the West Midlands Cancer Intelligence Unit in collaboration with the sarcoma SSCRG
- Utilisation of the list of soft tissue sarcomas should enable comparable soft tissue sarcoma research
  - Available on request

# Any questions?