

Forth Eastern European Conference for Rare Diseases and Orphan Drugs  
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## **LIST OF RARE CANCERS AND ITS RATIONALE**

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# Surveillance of rare cancers in Europe



Surveillance of Rare Cancers in Europe

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## Aims

- To provide a definition of "rare cancers" and a list of cancers
- To estimate the burden of rare cancers in Europe
- To improve the quality of data in cancer registration
- To develop strategies for the diffusion of information among all the key players

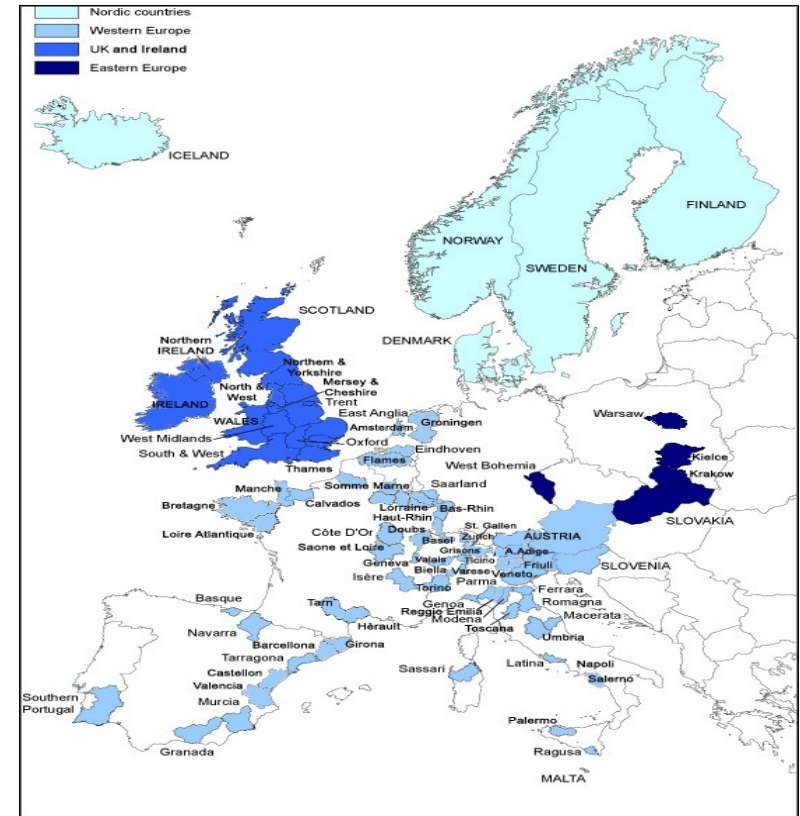
## Actions

- To provide figures on incidence, survival, prevalence and mortality for all rare cancers
- To analyse data quality for a subset of cancers
- To design a web-site on rare cancers to disseminate results

**Duration: 3 years (starting date 1<sup>st</sup> April 2007)**

# European coverage

- 88 European cancer registries (in 22 EU Countries)
- Population covered 146 millions
- Selected diagnosis period: 1995-2002
- 3,180,511 incident cases



# Operational definition of "rare cancer"

## ○ Rationale

- Frequency
  - Rare cancers are those cancers that present specific problem in clinical decision making, health care organization and clinical research because of their low frequency
- Incidence
  - Incidence better indicator for rare cancer
  - Prevalence better indicator for non neoplastic rare diseases
  - Therapy may hopefully impact of the choice of best indicator
  - Awareness of regulatory issues on orphan drugs
- Threshold for rarity = 6/100.000/year  
(any threshold is artificial: flexibility in using it)

# List of rare cancers

- Rarity determined by frequency
- Incidence
- Not a new classification

# The Way to the list

- Proposal from RARECARE WP4
  - Pathologists (few), Oncologists and Epidemiologists
- Consensus workshops
  - Brussels and Treviso
- WEB based discussion
- Consensus with scientific bodies

# Oncologic societies

## Invited

- o Societies under the umbrella of the European CanCer Organisation (ECCO)
- o Patient associations

Association of European Cancer Leagues
Breast International Group
European Association for Cancer Research
European Association of Neuro-Oncology
European Association of Nuclear Medicine
European Association of Urology
European Breast Cancer Coalition, EUROPA DONNA
European Group for Blood and Marrow Transplantation
European Oncology Nursing Society
European Organisation for Research and Treatment of Cancer
European Prostate Cancer Coalition, Europa Uomo
European School of Oncology
European Society for Medical Oncology
European Society for Paediatric Oncology
European Society for Therapeutic Radiology and Oncology
European Society of Breast Cancer Specialists
European Society of Gynaecological Oncology
European Society of Oncology Pharmacy
European Society of Skin Cancer Prevention
European Society of Surgical Oncology
Myeloma Euronet

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## List of Rare Cancers



### Rationale

The attached document provides a definition of rare cancers and explains the criteria used for defining the cancer entities shown in the list.

Rationale and Questions for Consensus - download PDF



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### List of Rare Cancers

This is the list of tumour entities from which rare tumours are identified as those with incidence less than 6 per 100,000 persons / year. The list presents the number of cases reported by European cancer registries during the period 1995-2002 and the corresponding incidence rates. Both figures are derived from the data of 70 population-based cancer registries adhering to the RARECARE project.

List of Rare Cancers - download PDF



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# The structure of the list of rare cancers

Layer	Tumour	Rate	Patients
1	<b>EPITHELIAL TUMOURS OF THE NASAL CAVITY and SINUSES</b>	<b>0.471</b>	<b>3060</b>
2	<b>Squamous cell carcinoma and variants of the Nasal Cavity and Sinuses</b>	<b>0.37</b>	<b>2615</b>
3	Squamous carcinoma	0.24	1708
3	Verrucous carcinoma	0.01	35
3	Squamous cell carcinoma spindle cell	0	6
3	Papillary squamous cell carcinoma	0	8
3	Basaloid carcinoma	0	10
3	Adenosquamous carcinoma	0	13
3	Squamous cell carcinoma, adenoid	0	3
3	Basaloid squamous cell carcinoma	0	0
3	Spindle cell carcinoma	0	0
2	<b>Lymphoepithelial carcinoma of the Nasal Cavity and Sinuses</b>	<b>0</b>	<b>12</b>
2	<b>Undifferentiated carcinoma of the Nasal Cavity and Sinuses</b>	<b>0.015</b>	<b>106</b>
2	<b>Intestinal type adenocarcinoma the Nasal Cavity and Sinuses</b>	<b>0.002</b>	<b>16</b>
1	<b>EPITHELIAL TUMOURS OF THE NASOPHARYNX</b>	<b>0.446</b>	<b>3171</b>
2	<b>Squamous cell carcinoma and variants of the Nasopharynx</b>	<b>0.32</b>	<b>2288</b>
3	Squamous carcinoma	0.162	1.156
3	Squamous cell carcinoma nonkeratinizing, NOS	0.02	144
3	Squamous cell carcinoma keratinizing, NOS	0.016	111
3	Papillary squamous cell carcinoma	0.001	7
3	Basaloid squamous cell carcinoma	0	0
3	Squamous cell carcinoma, adenoid	0	1
3	Lymphoepithelial carcinoma	0.057	405
3	Undifferentiated carcinoma	0.064	457
2	<b>Papillary adenocarcinoma of the Nasopharynx</b>	<b>0.001</b>	<b>5</b>
1	<b>EPITHELIAL TUMOURS OF THE MAJOR SALIVARY GLANDS and SALIVARY-GLAND TYPE TUMOURS</b>	<b>1.225</b>	<b>9060</b>
2	<b>Epithelial tumours of major salivary glands</b>	<b>0.848</b>	<b>6061</b>
3	Squamous carcinoma	0.119	851

# The Rationale Behind the List

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- First layer  $\Rightarrow$  Families of tumors (common referral pattern)
    - Organization of health care
  - Second layer  $\Rightarrow$  Tumors perceived by clinicians as a single disease entity
    - Clinical decision making; clinical research
  - Third layer  $\Rightarrow$  all WHO tumor entities

# The Rationale Behind the List

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- A tumor can be rare under the perspective of clinical decision making but not of health care organization (i.e. bronchiolo-alveolar carcinoma)
  - A tumor can be rare under both perspectives (i.e. sarcoma)

1	<b>EPITHELIAL TUMOUR OF THE LUNG</b>	<b>56.392</b>	<b>403258</b>
2	<b>Squamous cell carcinoma and variants of the Lung</b>	<b>14.071</b>	<b>109034</b>
3	Squamous carcinoma	12.798	91514
3	Papillary squamous cell carcinoma	0.005	37
3	Basaloid squamous cell carcinoma	0.001	9
3	Squamous cell carcinoma small cell nonkeratinizing	0.026	184
3	Squamous cell carcinoma clear cell type	0	1
2	<b>Adenocarcinoma and variants of the Lung</b>	<b>10.33</b>	<b>73890</b>
3	Adenocarcinoma NOS	9.363	66947
3	Mucinous adenocarcinoma	0.108	773
3	Papillary adenocarcinoma, NOS	0.137	980
3	Solid carcinoma, NOS	0.024	174
3	Clear cell adenocarcinoma, NOS	0.053	378
3	Acinic cell adenocarcinoma	0.047	337
3	Signet ring cell carcinoma	0.02	145
3	Adenosquamous carcinoma	0.283	2,022
3	Mucinous cystadenocarcinoma, NOS	0	2
3	Adenocarcinoma with mixed subtypes	0	1
3	Fetal adenocarcinoma	0	0
2	<b>Large cell carcinoma of the Lung</b>	<b>2.35</b>	<b>16806</b>
2	<b>Poorly differentiated endocrine carcinoma of the Lung</b>	<b>9.842</b>	<b>70369</b>
3	Small cell endocrine carcinoma of the Lung	7.485	53513
3	Large cell endocrine carcinoma of the Lung	2.357	16856
2	<b>Bronchiolo-alveolar carcinoma of the Lung</b>	<b>0.698</b>	<b>4984</b>
3	Bronchiolo-alveolar carcinoma, mucinous	0.002	11
3	Bronchiolo-alveolar carcinoma, non mucinous	0.001	9
3	Bronchiolo-alveolar carcinoma, mixed mucinous and non-mucinous	0.001	4
2	<b>Salivary gland type tumours of the Lung</b>	<b>0.04</b>	<b>293</b>
3	Adenoid cystic carcinoma	0.016	117
3	Mucoepidermoid carcinoma	0.024	174

1	<b>SOFT TISSUE SARCOMA</b>	5.558	39741
2	<b>Soft tissue sarcoma of the Head and Neck</b>	0.336	2408
3	Sarcoma NOS	0.03	212
3	Epithelioid sarcoma	0.001	10
3	Desmoplastic small round cell tumour	0	1
3	Fibrosarcoma	0.012	85
3	Fibromyxosarcoma	0.003	21
3	Infantile fibrosarcoma	0	2
3	Myxoinflammatory fibroblastic sarcoma	0	0
3	Malignant fibrous histiocytoma	0.039	280
3	DFSP	0	0
2	<b>Soft tissue sarcoma of the Mediastinum *</b>	0.026	186
2	<b>Soft tissue sarcoma of the Heart *</b>	0.016	110
2	<b>Soft tissue sarcoma of the Breast *</b>	0.202	1444
2	<b>Soft tissue sarcoma of the Uterus *</b>	0.917	6558
2	<b>Soft tissue sarcoma of the Genitourinary (other) *</b>	0.575	4109
2	<b>Soft tissue sarcoma of the other Viscera *</b>	0.582	4155
2	<b>Soft tissue sarcoma of the Paratesticular region *</b>	0.033	233
2	<b>Soft tissue sarcoma of the Retroperitoneum and Peritoneum *</b>	0.284	2028
2	<b>Soft tissue sarcoma of the Pelvis *</b>	0.017	116
2	<b>Soft tissue sarcoma of the Skin *</b>	0.319	2274
2	<b>Soft tissue sarcoma of the Paraorbital region *</b>	0.008	53
2	<b>Soft tissue sarcoma of the Brain and other Nervous System *</b>	0.188	1370
	<i>* includes all the entities listed for the soft tissue of the Head and Neck</i>		
2	<b>Embryonal rhabdomyosarcoma of the Soft Tissue</b>	0.046	334
2	<b>Alveolar rhabdomyosarcoma of the Soft Tissue</b>	0.024	176
2	<b>Ewing's family tumours of the Soft Tissue</b>	0.014	114
1	<b>BONE SARCOMA</b>	0.763	5456
2	<b>Osteogenic sarcoma</b>	0.218	1546
3	Conventional osteosarcoma	0.201	1,434
3	Teleangectatic osteosarcoma	0.005	35
3	Small cell osteosarcoma	0.002	11
3	Paraosteal osteosarcoma	0.009	62

## What to do with this list

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- Produce burden indicators
  - ⇒ Report soon available on the web-site
- Improve data quality in registration
- Identify areas in which special efforts are needed

# What to do with this list

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- Ideas from the last RARECARE meeting (June 10, 2009)
    - Call for
      - centers of expertise into European reference networks
      - timely, appropriate diagnosis and treatment for rare cancers
      - investigation on the off-label use in rare cancers
      - alternative research methodologies for rare cancer
      - a greater involvement of diseases oriented research communities in providing advice to pharmaceutical industry on the development of new drugs

Thank you for your attention

RARECARE web site

<http://www.rarecare.eu/>