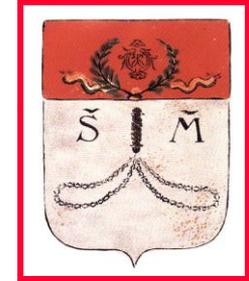


A List of Rare Cancer



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Why a List of Rare Cancers?

- Recognition of tumor types is the key to proper treatment
- Classification is based on pathology and genetics
- Need for closer relationships between tumor type and treatment



Why a List of Rare Cancers?

- Rarity represents a major challenge
- Diagnosis
- Clinical decision making
- Clinical studies
- Health care organization



How we Got to the Rare Cancer List

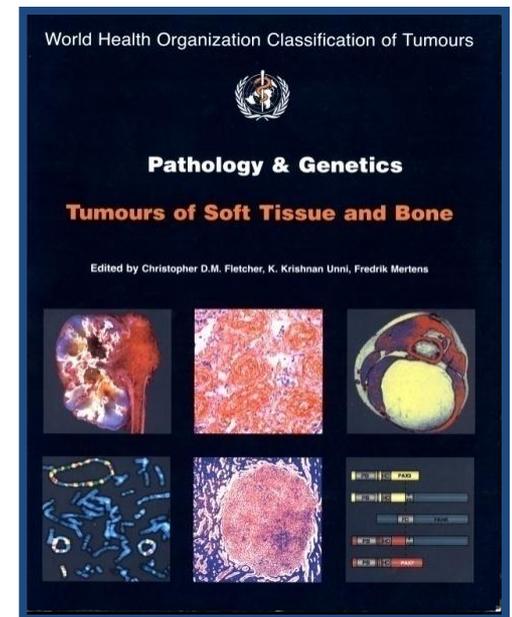
The RARECARE project

- Multiprofessional effort
- Pathologists
- Medical Oncologists
- Haematologists
- Epidemiologists
- Consensus conferences



Classification of Tumors

- Based on Pathology
- WHO
 - Pathology and Genetics
- Need for clinical meaning
- Rationale grouping of WHO entities



How we Got to the Rare Cancer List

The RARECARE project

- Three layers
- 1st layer: families of tumors
- 2nd layer: tumours perceived by clinicians as a single disease)
- Third layer: WHO entities

Rare Cancer List

The RARECARE project

- 1st layer: families of tumors
 - Consensus-based clinical perspective
 - Patient referral purposes
 - Relevant for Health Care organization



Surveillance of Rare Cancers in Europe

Rare Cancer List

The RARECARE project

- 2nd layer: : tumours defined in a clinically sound way
 - perceived by clinicians as a single disease
 - Relevant for decision making purposes
 - Clinical studies



Surveillance of Rare Cancers in Europe

Rare Cancer List

The RARECARE project

- 3rd layer: WHO entities
 - Based on pathology with integration of molecular genetics
 - List is not a replacement of current (evolving) classification schemes

Rare Cancer List

The RARECARE project

- Crude incidence
- Incidence rate calculated on the basis of 70 population-based cancer registries



Surveillance of Rare Cancers in Europe



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journal homepage: www.ejconline.com



Rare cancers are not so rare: The rare cancer burden in Europe

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Table 2 – RARECARE estimates of incidence, survival and prevalence of cancers for EU27, together with expected number of new cases per year and prevalent cases in EU27.

Rare (R) or common (C) (middle tier only)	Tier	Top tier (upper case) and middle tier (lower case) tumour categories	Crude incidence per 100,000 per year	Standard error incidence	Expected new cases per year	Observed 5-year survival (%)	Relative 5-year survival (%)	Standard error relative survival (%)	Complete prevalence per 100,000	Standard error complete prevalence	Prevalent Cases
R	1	EPITHELIAL TUMOURS OF NASAL CAVITY AND SINUSES	0.44	0.01	2198	39.3	48.3	1.3	2.92	0.08	14,492
	2	Squamous cell carcinoma with variants of nasal cavity and sinuses	0.31	0.01	1545	40.2	49.2	1.5	2.10	0.07	10,416
R	2	Lymphoepithelial carcinoma of nasal cavity and sinuses	0.00	0.00	12	28.6	31.0	13.1	0.01	0.01	72
R	2	Undifferentiated carcinoma of nasal cavity and sinuses	0.02	0.00	86	27.5	32.4	6.0	0.13	0.02	665
R	2	Intestinal type adenocarcinoma of nasal cavity and sinuses	0.00	0.00	12	43.0	50.1	14.6	0.02	0.01	123
R	1	EPITHELIAL TUMOURS OF NASOPHARYNX	0.44	0.01	2205	44.1	49.1	1.1	2.94	0.09	14,637
	2	Squamous cell carcinoma with variants of nasopharynx	0.33	0.01	1626	44.4	49.2	1.3	2.20	0.07	10,966
R	2	Papillary adenocarcinoma of nasopharynx	0.00	0.00	4	57.1	58.8	23.8	0.01	0.00	29
R	1	EPITHELIAL TUMOURS OF MAJOR SALIVARY GLANDS AND SALIVARY-GLAND TYPE TUMOURS	1.31	0.01	6501	54.2	64.8	0.7	13.08	0.18	65,063
	2	Epithelial tumours of major salivary glands	0.73	0.01	3624	53.7	64.6	1.0	7.90	0.14	39,290
R	2	Salivary gland type tumours of head and neck	0.43	0.01	2134	60.3	69.1	1.2	4.53	0.11	22,553
R	1	EPITHELIAL TUMOURS OF HYPOPHARYNX AND LARYNX	6.26	0.03	31,138	46.9	54.8	0.3	39.98	0.33	198,863
	2	Squamous cell carcinoma with variants of hypopharynx	1.19	0.01	5905	21.6	24.6	0.6	3.47	0.09	17,293
R	2	Squamous cell carcinoma with variants of larynx	4.64	0.02	23,082	54.5	63.7	0.4	34.39	0.28	171,098
R	1	EPITHELIAL TUMOURS OF OROPHARYNX	2.75	0.02	13,667	33.1	37.1	0.4	13.04	0.18	64,877
	2	Squamous cell carcinoma with variants of oropharynx	2.58	0.02	12,858	33.3	37.2	0.5	12.52	0.18	62,254
R	1	EPITHELIAL TUMOURS OF ORAL CAVITY AND LIP	4.79	0.02	23,828	49.0	59.1	0.4	34.07	0.35	169,507
	2	Squamous cell carcinoma with variants of oral cavity	3.28	0.02	16,337	41.3	48.2	0.4	19.34	0.25	96,196
R	2	Squamous cell carcinoma with variants of lip	1.22	0.01	6093	70.1	91.7	0.7	12.79	0.18	63,621
R	1	EPITHELIAL TUMOURS OF OESOPHAGUS	7.51	0.03	37,379	8.4	10.6	0.2	12.11	0.16	60,221

R	1	EPITHELIAL TUMOUR OF TRACHEA	0.13	0.00	670	10.1	12.1	1.4	0.28	0.02	1396
R	2	Squamous cell carcinoma with variants of trachea	0.08	0.00	408	7.2	8.5	1.4	0.12	0.01	602
R	2	Adenocarcinoma with variants of trachea	0.01	0.00	67	6.6	7.6	3.3	0.02	0.01	119
R	2	Salivary gland type tumours of trachea	0.01	0.00	48	50.9	55.2	7.7	0.11	0.02	523
C	1	EPITHELIAL TUMOUR OF LUNG	55.93	0.08	278,226	8.5	10.6	0.1	85.00	0.44	422,831
C	2	Squamous cell carcinoma with variants of lung	13.49	0.04	67,125	10.9	13.4	0.1	25.35	0.23	126,097
C	2	Adenocarcinoma with variants of lung	10.29	0.04	51,193	11.8	13.9	0.2	22.14	0.22	110,140
R	2	Large cell carcinoma of lung	4.01	0.02	19,936	10.2	12.3	0.3	6.83	0.12	33,969
R	2	Well differentiated endocrine carcinoma of lung	0.63	0.01	3148	53.0	58.7	1.0	6.96	0.18	34,627
C	2	Poorly differentiated endocrine carcinoma of lung	7.68	0.03	38,221	3.9	4.6	0.1	8.43	0.13	41,925
R	2	Bronchiolo-alveolar carcinoma of lung	0.68	0.01	3383	26.5	31.1	0.9	2.42	0.07	12,066
R	2	Salivary gland type tumours of lung	0.04	0.00	220	38.5	43.4	3.6	0.30	0.03	1505
R	2	Sarcomatoid carcinoma of lung	0.14	0.00	697	13.4	15.9	1.5	0.32	0.02	1621
R	2	Undifferentiated carcinoma of lung	0.98	0.01	4887	5.6	6.6	0.4	1.27	0.05	6328
R	1	EPITHELIAL TUMOURS OF THYMUS	0.17	0.00	829	52.6	57.7	1.9	1.40	0.06	6962
R	2	Malignant thymoma	0.14	0.00	680	55.7	60.9	2.0	1.22	0.06	6055
R	2	Squamous cell carcinoma of thymus	0.00	0.00	23	40.0	44.6	10.9	0.02	0.01	119
R	2	Undifferentiated carcinoma of thymus	0.00	0.00	12	16.7	18.2	11.8	0.00	0.00	16

Rare Cancer List

Endorsers

- Childhood Cancer Research Group, <http://www.ccrq.ox.ac.uk>
- CML Advocates Network, <http://www.cmladvocates.net>
- EUROPA DONNA - The European Breast Cancer Coalition:
www.europadonna.org
- European Association for Cancer Research (EACR),
<http://www.eacr.org>
- European Association of Neuro-Oncology (EANO),
<http://www.eano.eu>
- European Association of Nuclear Medicine (EANM),
<http://www.eanm.org>

Rare Cancer List

Endorsers

- European Cancer Patient Coalition (ECPC),
<http://www.ecpc-online.org>
- European Group for Blood and Marrow Transplantation: www.ebmt.org
- European Oncology Nursing Society,
<http://www.cancernurse.eu>
- European Prostate Cancer Coalition, Europa Uomo,
<http://www.europa-uomo.org>

Rare Cancer List

Endorsers

- European Society of Gynaecological Oncology (ESGO),
<http://www.esgo.org>
- European Society for Medical Oncology (ESMO),
<http://www.esmo.org>
- European Society for Paediatric Oncology (SIOP),
<http://www.siope.eu>
- European Society of Surgical Oncology (ESSO),
<http://www.essoweb.org>

Rare Cancer List

Endorsers

- European Society for Therapeutic Radiology and Oncology (ESTRO), <http://www.astro.org>
- International Brain Tumour Alliance (IBTA), <http://www.theibta.org>
- International Union Against Cancer (UICC), <http://www.uicc.org>
- MYELOMA EURONET A.i.s.b.l.(ME) Leukaemiehilfe RHEIN-MAIN e.V. (LHRM), <http://www.myeloma-euronet.org>

Rare Cancer List

Prevalence vs. Incidence

- Should rare cancers be defined the same way as rare diseases?
 - Prevalence < 50/100,000
- Life expectancy is variable across rare cancers
- Prevalence not the best indicator of rare cancer frequency
- Incidence more useful

Rare Cancer List

What threshold for rarity?

- Any threshold is artificial
- Any threshold should be used with flexibility
- 6/100,000/year is the results of the consensus among clinicians

Rare Cancer List

Future Perspectives

- WHO classifications evolve
- New entities
- Increasing role of molecular partitioning
- Breast
 - Luminal A, B, HER2, triple negative...
- Lung
 - EGFR, KRAS; HER2, ALK...

