# CONSENSUS MEETING AND CONFERENCE IMPROVING THE METHODOLOGY OF CLINICAL RESEARCH ON RARE CANCERS BRUSSELS, 10 FEBRUARY 2012

#### The epidemiology of rare cancers in Europe

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

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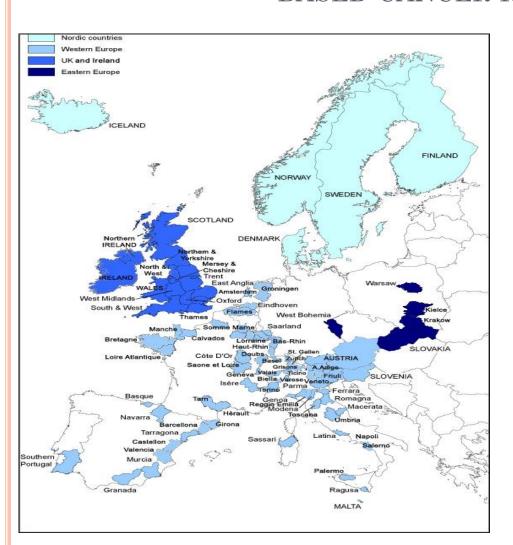
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### Numbers in Europe



Surveillance of Rare Cancers in Europe

### EPIDEMIOLOGY OF RARE CANCERS = NETWORKS OF POPULATION BASED CANCER REGISTRIES



Europe: RARECARE/EUROCARE

RESEARCH ARTICLES

### The Occurrence of Rare Cancers in U.S. Adults, 1995–2004

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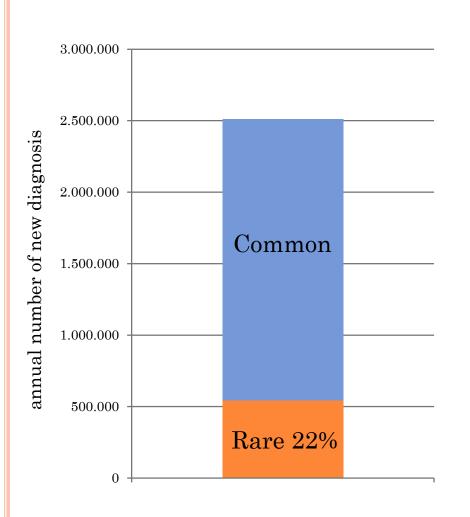
#### **SYNOPSIS**

**Objective.** Rare cancers have been traditionally understudied, reducing the progress of research and hindering decisions for patients, physicians, and policy makers. We evaluated the descriptive epidemiology of rare cancers using a large, representative, population-based dataset from cancer registries in the United States.

Methods. We analyzed more than 9 million adult cancers diagnosed from 1995 to 2004 in 39 states and two metropolitan areas using the Cancer in North America (CINA) dataset, which covers approximately 80% of the U.S. population. We applied an accepted cancer classification scheme and a published definition of rare (i.e., fewer than 15 cases per 100,000 per year). We calculated age-adjusted incidence rates and rare/non-rare incidence rate ratios using SEER\*Stat software, with analyses stratified by gender, age, race/ethnicity, and histology.

USA: SEER or the Centers for Disease Control and Prevention's National Program of Cancer Registries

#### RARE CANCERS: INCIDENCE

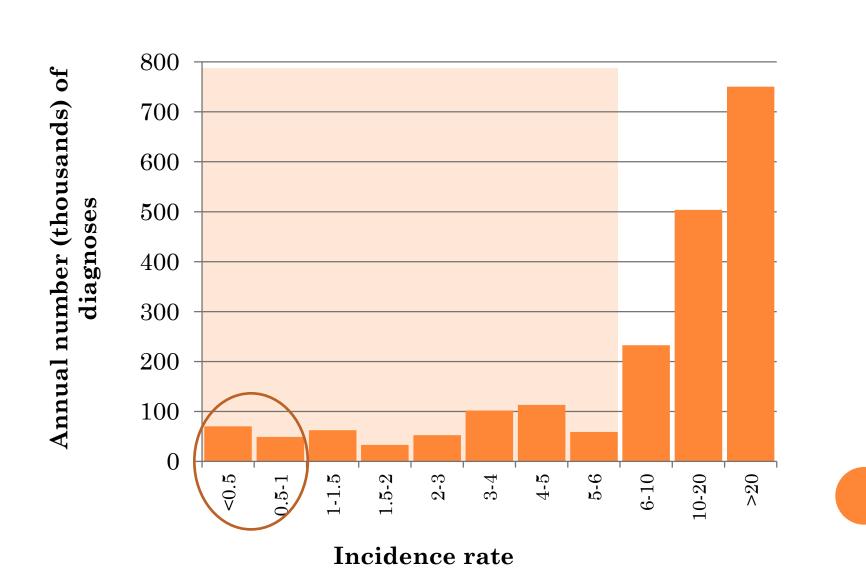


186 rare cancers

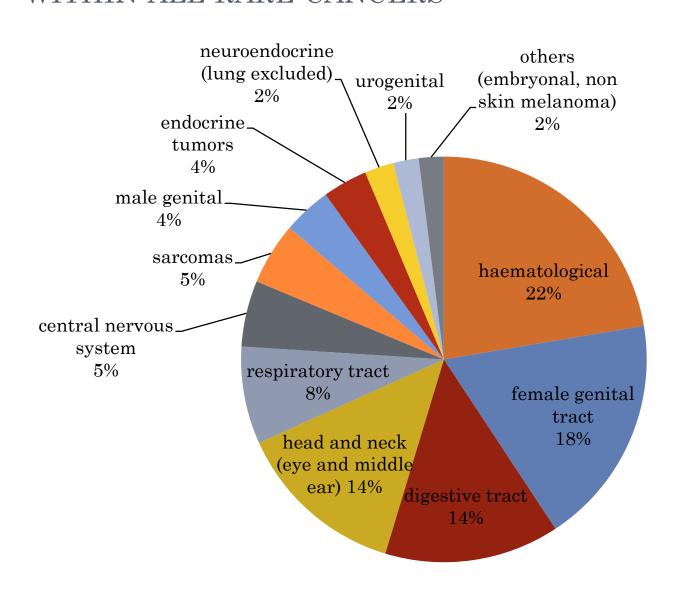
About 500,000 new cases/year in EU27

22% of all cancer diagnosed/year

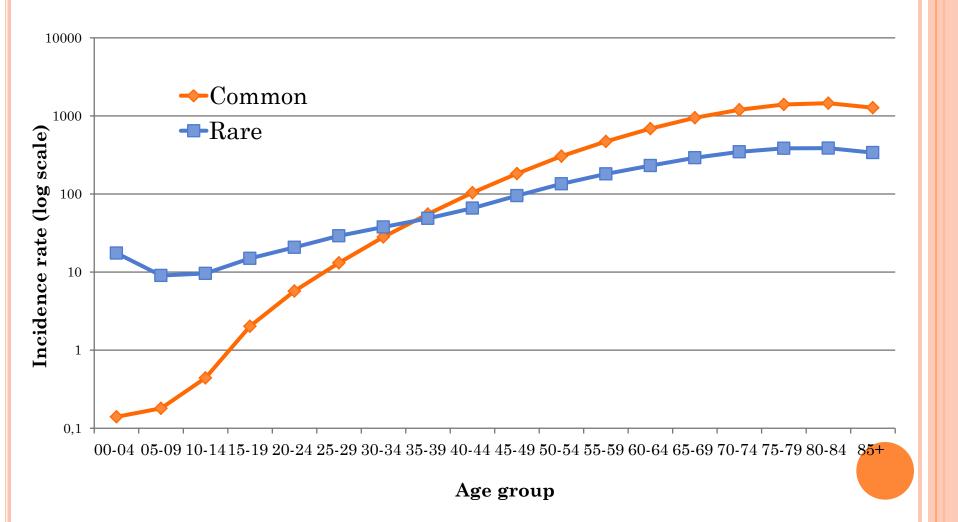
### DISTRIBUTION OF ANNUAL NUMBER OF DIAGNOSES IN EU27 ACCORDING TO CATEGORIES OF INCIDENCE RATE



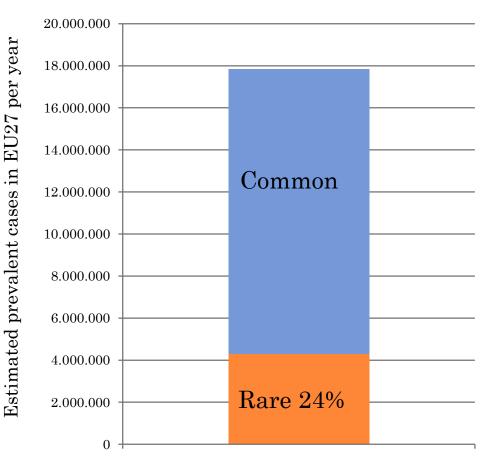
# DISTRIBUTION OF MAJOR FAMILIES OF RARE TUMORS WITHIN ALL RARE CANCERS



## AGE-SPECIFIC INCIDENCE RATES FOR RARE AND COMMON CANCERS IN EU 27



#### RARE CANCERS: PREVALENCE

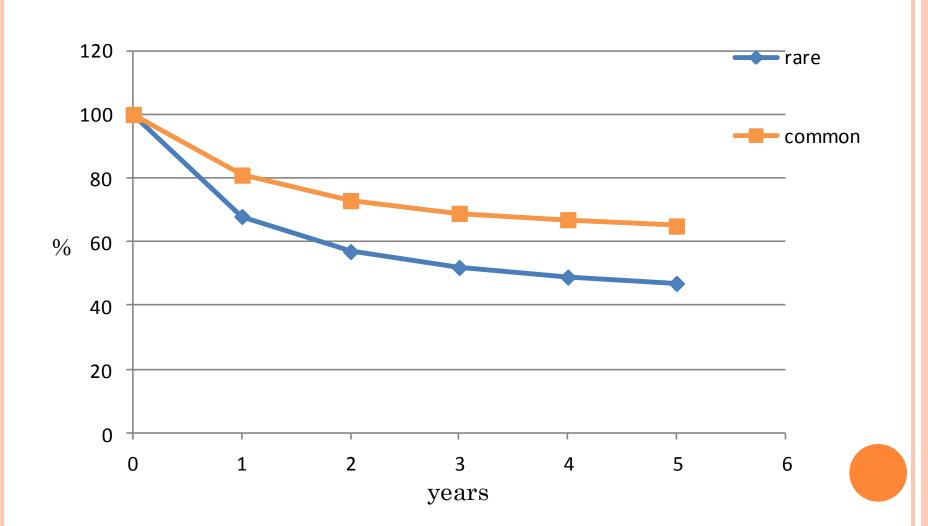


#### 186 rare cancers

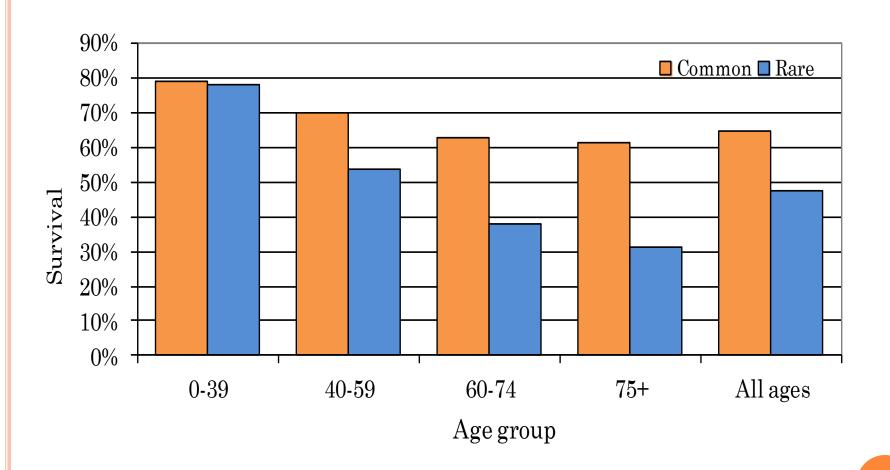
4,300,000 people alive in the EU27 with a previous diagnosis of a rare cancer

24% of the total cancer prevalence

#### RARE CANCERS: 5 YEAR RELATIVE SURVIVAL



### 5 YEAR RELATIVE SURVIVAL FOR RARE AND COMMON CANCERS IN EU27 BY AGE GROUP



#### RARECARE – WHAT'S ADDED

- Put numbers to a problem long known to exist
- Confirmed low outcome of rare cancers
- Made prevalence available for rare cancers
- Identified a new priority for population-based cancer registries

#### RARE CANCER DEFINITION

• Incidence is the best indicator to define rare cancers

	Crude incidence per 100,000/ year	Relative 5-year survival (%)	Complete prevalence per 100,000
Squamous cell carcinoma with variants of lung	13	13	<b>25</b>
Adenocarcinoma with variants of lung	10	14	22
Adenocarcinoma with variants of stomach	15	23	46

#### NOT JUST DESCRIPTIVE EPIDEMIOLOGY

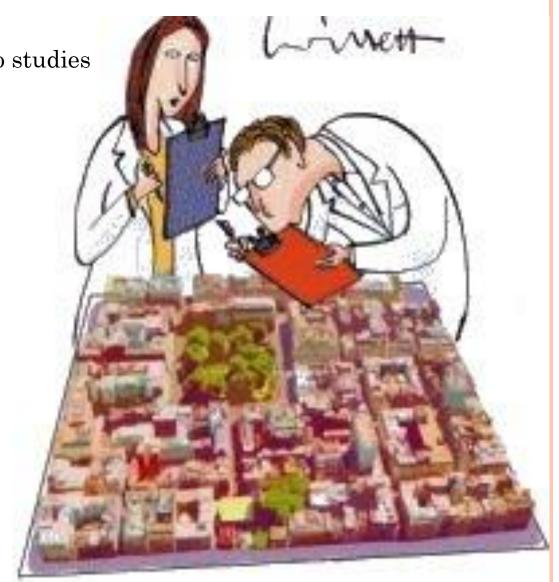
### RARECARENET

#### **AIMS**

- To describe the **health care pathways** for rare cancers and update **epidemiological** indicators.
- To identify and disseminate information on centers of expertise for rare cancers.
- To produce and disseminate information on diagnosis and management of rare cancers.
- To develop and disseminate information for patients and patients' associations dedicated to rare cancers.

### RESEARCH COMMUNITY

Don't forget to include those who studies the disease in the Community



#### Thank you for your attention!!

#### www.rarecare.eu

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#### THE RARE CANCER BURDEN IN EUROPE

- o 22% of all new malignancies
  - About 30% of Europeans have one of the particularly rare forms that affect <1/100,000
- 24% of the total cancer prevalence (4 millions people living with a diagnosis of rare cancers in EU27)

• Rare cancer survival is worse than common cancer 47% vs 65%



- uterine cervix and thyroid carcinoma are rare according to the incidence (RARECARE) criterion and 'common' according to the prevalence criterion.
- Six cancers are common according to the incidence criterion and rare according to the prevalence criterion:
- 1. stomach adenocarcinoma,
- 2. pancreatic adenocarcinoma,
- 3. Lung adenocarcinoma,
- 4. lung squamous cell carcinoma,
- 5. poorly differentiated endocrine carcinomas of lung and
- 6. the group othernon-Hodgkin mature B cell lymphomas.

# ESTIMATES OF PREVALENCE FOR RARE AND COMMON CANCERS BY SITE IN EU27

		Prevalence per	Estimated	Prevalence
		100,000	prevalent cases in	distribution (%)
			EU27 per year	
Rare	Haematopoietic system	90	$450,\!444$	70
Common	Haematopoietic system	32	161,618	25
All	Haematopoietic system	130	647,596	100
Rare	Female genital tract	176	880,922	<b>5</b> 3
Common	Female genital tract	127	633,280	38
All	Female genital tract	332	1,658,891	100
Rare	Respiratory tract	60	300,193	46
Common	Respiratory tract	56	279,942	43
All	Respiratory tract	130	648,321	100
Rare	Male genital tract	93	465,363	23
Common	Male genital tract	279	1,396,883	70
All	Male genital tract	400	1,997,563	100
$\overline{ ext{Rare}}$	Digestive tract	51	254,473	11
Common	Digestive tract	399	1,996,625	84
All	Digestive tract	476	2,380,246	100
$\overline{ ext{Rare}}$	Breast	60	300,759	9
Common	Breast	520	2,599,432	74
All	Breast	700	3,500,906	100
Rare	Urinary system	18	91,683	8
Common	Urinary system	202	1,010,735	85
All	Urinary system	238	1,188,660	100