



FONDAZIONE IRCCS  
ISTITUTO NAZIONALE  
DEI TUMORI



Sotto l'alto patrocinio  
  
*Presidenza del Consiglio dei Ministri*



# RARECARE and RARECAREnet Projects on rare cancers in Europe

Clustering and communication meeting on rare diseases  
EAHC 25-26 October, Luxembourg



- n Surveillance of rare cancers in Europe – RARECARE (2007-2010)
- n Information network on rare cancers – RARECAREnet (negotiation)

**Rare diseases : all rare malignant tumours**



n Surveillance of rare cancers in Europe – RARECARE (2007-2010)

n Information network on rare cancers – RARECAREnet (negotiation)

**Source of data = population-based cancer registries**

# RARECARE

- n Definition and list
- n Burden in Europe
- n Improve data quality
- n Dissemination

# RARECARE, coordinating center

## Role in the national healthcare system



## The oldest Italian cancer Institute Involved in care and research

Project coordinated by the Evaluative Epidemiology Unit



# RARECARE, definition and list

- n Consensus process with group of experts
- n 220 clinical entities
- n 186 rare entities

**Comprehensive list of rare entities in [www.rarecare.eu](http://www.rarecare.eu)**

# RARECARE, burden in Europe

## how many cases?

- n About 500,000 new cases per year in the EU27
- n They are 22% of all malignancies

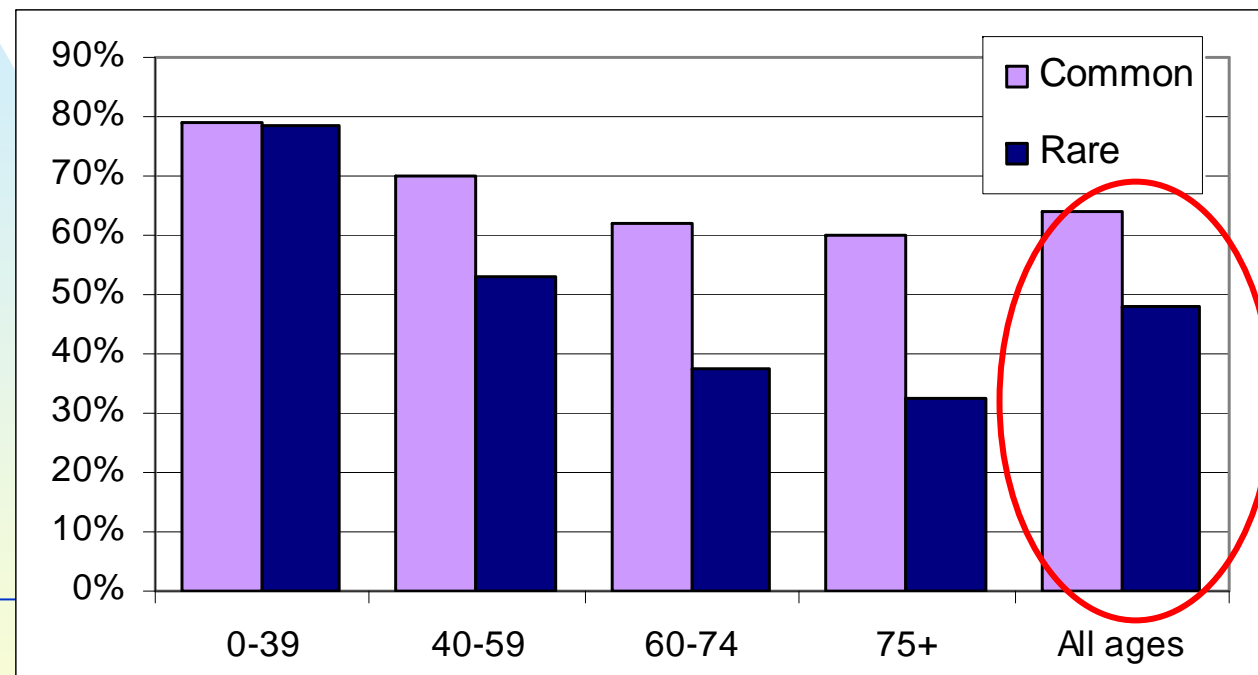
**RARECARE, burden in Europe**

**how many people is living with a  
rare cancer in the EU27?**

**About 4 million**

# RARECARE, burden in Europe

## rare cancer survival in the EU27



**5-year survival:  
48% rare and 64% common cancers**

# RARECARE, improve data quality

- n 8% of all cancer cases in the study was without a clear morphological diagnosis
- n after the revision of pathological reports, the % slightly reduced

**Difficulty in clinical diagnosis**

# Dissemination

---



Home

Project Aims

Work Packages /  
Partners

Rare Cancers List

Rare Cancers Indicators

Rare Cancers  
Data Quality **NEW**

Meetings

Publications /  
Presentations at  
Scientific Congresses

Useful Links

Contact Us

## Technical report with basic indicators for rare cancers and health care related macro indicators



The attached documents are the technical report with basic indicators for rare cancers and health care related macro indicators

**Technical Report**

Download PDF



**Technical Report Annex**

Download PDF



**Please Note:** Adobe Reader is required to view PDF's, it can be downloaded from [here](#)

## Rare Cancer Search



Enter a % symbol to return the full list

Search for a Rare Cancer to be given a link to the relevant page of the Technical Report Annex

**Please Note:** The search tool will be improved in the future to include combined searches.

# Dissemination Papers in peer review journals

ARTICLE IN PRESS

EUROPEAN JOURNAL OF CANCER XXX (2011) XXX-XXX

---


EJC 8105  
15 October 2011

ARTICLE IN PRESS

EUROPEAN JOURNAL OF CANCER XXX (2011) XXX-XXX

No. of Pages 12, Model 7

---




**ELSEVIER**

Available at [www.sciencedirect.com](http://www.sciencedirect.com)

SciVerse ScienceDirect

journal homepage: [www.ejconline.com](http://www.ejconline.com)



---

**2 Burden of testicular, paratesticular and extragonadal germ cell**

**3 tumours in Europe**

**4 A. Trama <sup>a,\*</sup>, S. Mallone <sup>b</sup>, N. Nicolai <sup>c</sup>, A. Necchi <sup>c</sup>, M. Schaapveld <sup>d,e</sup>, J Gietema <sup>f</sup>,**

**5 A. Znaor <sup>g,h</sup>, E. Ardanaz <sup>i,j</sup>, F. Berrino <sup>a</sup>, The RARECARE working group**

**burden in Europe**

**<sup>c</sup>, Sabine Siesling <sup>b</sup>,**

**<sup>f</sup>, Sandra Mallone <sup>g</sup>,**

**RARECARE working group**

ori, Via Venezian 1, 20133 Milan, Italy

9700 AH Groningen, The Netherlands

, 20133 Milan, Italy

XU, UK

Via Venezian 1, 20133 Milan, Italy

taly

| Topics                      | Epidemiological paper authors  | paper progress                                | Clinical papers authors  |
|-----------------------------|--|---|--|
| Rare cancers of HEAD & NECK | Boukje Van Dijk (Comprehensive Cancer Centre)<br>Gemma Gatta (Ist. Naz. Tumori, )<br>Riccardo Capocaccia (Istituto )<br>Daniela Pierannunzio (Istituto Superiore Sanità, Italy)<br>and the RARECARE WG   | Sent to the EJC<br>(approved for publication) | Lisa Licitra (Ist. Naz. Tumori, )<br>Primož Strojjan (Inst. of Oncology ) - ESTRO  |
| Rare DIGESTIVE cancers      | Jean Faivre (Cote D'Or digestive CR)<br>Roberta De Angelis (Istituto )<br>Annalisa Trama (Ist. Naz. Tumori, )<br>Marloes Elferink (CCC North East)<br>(CCC North East), the RARECARE WG  | Sent to the EJC<br>(approved for publication) | A. Cervantes (Hospital Clínico Universitario, Valencia, )<br>Riccardo Audisio (ESSO)<br>Jean-François Bosset (Oncologie-Radiothérapie, , Besançon)-<br>ESTRO                   |
| Rare MALE GENITAL cancers   | Annalisa Trama (Ist. Naz. Tumori, Milano, Italy)<br>Michael S. Berrino (Ist. Naz. Tumori, Milano, Italy)<br>Ariana Znaor (Ist. Naz. Tumori, Milano, Italy)<br>Sandra Mallone (Ist. Naz. Tumori, Milano, Italy)<br>Eva Ardanaz (Navarra Cancer Registry) and<br>the RARECARE WG                 | <b>Approved for publication</b> (NL)          |  |
| Rare UROGENITAL cancers     | Otto Visser (CCC Amsterdam)<br>Jan Adolfson ( CR)<br>Silvia Rossi (Istituto Superiore Sanità, Italy)<br>Julia Verne (South West Public Health Observatory)<br>Gemma Gatta (Ist. Naz. Tumori, Milano, Italy) and<br>the RARECARE WG   | Sent to the EJC<br>(approved for publication) | J.P. Droz (Centre Léon-Bérard, )<br>Massimo Maffezzini and Fabio Campodonico<br>Dr Kevin N Franks ( St James's . Leeds Teaching Hospitals NHS<br>Trust)-ESTRO (penile cancers) |
| Embryonal tumours           | Gemma Gatta (Ist. Naz. Tumori, Milano, Italy)<br>Charles Stiller (Childhood cancer research group)<br>Guido Pastore (Childhood Cancer Registry, )<br>Annalisa Trama (Ist. Naz. Tumori, Milano, Italy)<br>Riccardo Capocaccia (Istituto Superiore Sanità, Italy)<br>and the RARECARE WG         | Sent to the EJC<br>(approved for publication) | Andrea Ferrari (Ist. Naz. Tumori, Milano, Italy)<br>Gianni Bisogno (SIOPE)   |
| Rare SKIN cancers           | Sandra Mallone (Istituto Superiore Sanità, Italy)<br>Julia Verne (South West Public Health Observatory)<br>Ruth Martinez (Basque Country CR)<br>Carmen Navarro, Maria Dolores Chirlaque (Murcia CR)<br>Eva Ardanaz (Navarra Cancer Registry)<br>Jan Willem, Esther de Vries<br>the RARECARE WG | Sent to the EJC<br>(approved for publication) | C.J. Punt ( , )  |

| Topics                      | Epidemiological paper authors   | paper progress                         | Clinical papers authors  |
|-----------------------------|---|--|--|
| Rare CNS cancers            | Emanuele Crocetti, Adele Caldarella (Toscana CR)<br>Charles Stiller (Childhood cancer research group)<br>Jerzy Slovinsky (Silesia CR)<br>Annalisa Trama (Ist. Naz. Tumori, )<br>and the RARECARE WG   | Reviewers comments received            | Brandes (Ospedale Bellaria-Maggiore Bologna, Riccardo Soffiatti<br>Damien C. Weber (Hôpitaux Universitaires de Genève, Suisse)-ESTRO<br>J. Jaal (, Estonia)<br>Umberto Ricardi (Università di Torino, Italy) |
| Endocrine tumors            | (CCC Centre North East)<br>Sandra Mallone (Istituto Superiore Sanità, Italy)<br>Magdalena Bielska-Lasota (Public Health Inst )<br>Roberto Foschi (Ist. Naz. Tumori, Milano, Italy) and the RARECARE WG  | Reviewers comments received            | E. Baudin (Institut Gustave-Roussy, , France)<br>Thera Links (international endocrine group)   |
| SARCOMAS                    | Charles Stiller (Childhood cancer research group)<br>Diego Serraino ( CR),<br>Annalisa Trama (Ist. Naz. Tumori, )<br>Carmen Navarro and Maria Dolores Chirlaque ( CR) and the RARECARE WG   | Draft ready to be submitted to the EJC | P.G. Casali (Ist. Naz. Tumori, )<br>Lex Eggermont<br>Tom Treasure (UCL, surgeon)<br>Umberto Ricardi ( )-ESTRO  |
| NEUROENDOCRINE cancers      | (CCC North East)<br>Annalisa Trama (Ist. Naz. Tumori, )<br>Nerea Larranaga (Basque Country CR)<br>Andrea Tavilla (Istituto )<br>the RARECARE WG   | Draft ready to be submitted to the EJC | E. Baudin (Institut Gustave-Roussy, )<br>Thera Links (international endocrine group)<br>Graeme Poston (surgeon)  |
| Rare HEMATOLOGICAL cancers  | Roberta De Angelis (Istituto Superiore Sanità, Italy)<br>Charles Stiller (Childhood cancer research group)<br>Annalisa Trama (Ist. Naz. Tumori, )<br>Rafael Marcos-Gragera (Girona Cancer Registry)<br>Otto Visser (CCC) and the RARECARE WG  | Draft ready to be submitted to the EJC | H.C. Schouten ( , )<br>Marc Maynadie ( D'Or Hematol)<br>Umberto Ricardi ( )-ESTRO  |
| Rare THORACIC cancers       | (CCC North East)<br>Isabel Izarzugaza (Basque Country CR)<br>Roberto Foschi (Ist. Naz. Tumori, Milano, Italy),<br>Andrea Tavilla (Istituto Superiore Sanità, Italy)<br>Eva Ardanaz ( Cancer Registry) and the RARECARE WG   | Discussion among co-authors on going   | Tom Treasure (UCL, surgeon)<br>J. Jaal ( , oncologist)-ESTRO<br>Umberto Ricardi ( )  |
| Rare FEMALE GENITAL cancers | Roberta De Angelis (Istituto Superiore Sanità, Italy)<br>Emanuele Crocetti, Adele Caldarella (Toscana CR)<br>Ester Hernandez (Basque Country CR)<br>Daniela Pierannunzio (Istituto Superiore Sanità, Italy)<br>Annalisa Trama (Ist. Naz. Tumori, Milano, Italy)<br>(CCC North East) the RARECARE WG | Discussion among co-authors on going   | I. Ray-Coquard (Centre Léon-Bérard, )  |

# RARECARE, potential impact

- n Put numbers (incidence, prevalence, survival) to a problem long known to exist
- n Awareness of cancer registries
- n Necessity to improve quality of diagnosis

# Further prospectives RARECAREnet

- n To describe **diagnosis and treatment pathway** for across EU countries from registry data.
- n To identify the qualification criteria for **centres of expertise** and list centres of expertise
- n To produce and disseminate **information on diagnosis and management** of rare cancers
- n To develop a **clinical database** on very rare cancers to develop new knowledge on their clinical management

**Thank you**

[gemma.gatta@istitutotumori.mi.it](mailto:gemma.gatta@istitutotumori.mi.it)

---

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

# Dissemination, papers in peer review journals

EJC 8105  
15 October 2011



ELSEVIER

## 2 Burden of testicular 3 tumours in Europe

4 A. Trama <sup>a,\*</sup>, S. Maltoni <sup>a</sup>,  
5 A. Znaor <sup>g,h</sup>, E. Ardano <sup>a</sup>

Jean Michel Lutz, Carmen Martinez, et al.

### Summary

**Background** Rare cancers are a challenge to which rare cancers are usually referred in a large population of several countries.

**Methods** We searched medical literature

### Rare Cancers of **head and neck**

Incidence, prevalence and survival in rare epithelial **digestive cancers**

**Rare thoracic cancers**, including peritoneum mesothelioma

Descriptive Epidemiology of **Sarcomas** in Europe. Report from the RARECARE Project

**Extracutaneous melanomas** (uveal and mucosal) and adnexal skin carcinomas in Europe

Rare cancers of **female genital tract**

Rare tumours of **testis and extragonadal germ cell tumours**

Rare **Urogenital Cancers**

Rare **Central Nervous System Cancers**

**Endocrine Tumours**: results of the RARECARE project

**Neuroendocrine Tumours**: results of the RARECARE project

The **embryonal cancers** in Europe

Rare **Myeloid malignancies**

No. of Pages 12, Model 7



## adrenal germ cell

J Gietema <sup>f</sup>,  
ap

centres  
ers in a  
Published online  
December 13, 2005  
DOI:10.1016/S1470-2045(05)  
70471-X

\*Members listed at end of report