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
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

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

Gemma Gatta a,*, Jan Maarten van der Zwan b, Paolo G. Casali c, Sabine Siesling b, Angelo Paolo Dei Tos d, Ian Kunkler e, Renée Otter b, Lisa Licitra f, Sandra Mallone g, Andrea Tavilla g, Annalisa Trama a, Riccardo Capocaccia g, The RARECARE working group
<table>
<thead>
<tr>
<th>Rare (R) or common (C) (middle tier only)</th>
<th>Tier</th>
<th>Top tier (upper case) and middle tier (lower case) tumour categories</th>
<th>Crude incidence per 100,000 per year</th>
<th>Standard error incidence</th>
<th>Expected new cases per year</th>
<th>Observed 5-year survival (%)</th>
<th>Relative 5-year survival (%)</th>
<th>Standard error relative survival (%)</th>
<th>Complete prevalence per 100,000</th>
<th>Standard error complete prevalence</th>
<th>Prevalent Cases</th>
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Rare Cancer List

Endorsers

- Childhood Cancer Research Group, http://www.ccrg.ox.ac.uk
- 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
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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
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Endorsers

- European Society for Therapeutic Radiology and Oncology (ESTRO), http://www.estro.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...
THE END