RARECARE
Surveillance of Rare Cancers in Europe

RARECARE project

2nd Consensus meeting on definition and list of rare cancers

Treviso, 26th – 27th May, 2008

Minutes

List of participants: Gemma Gatta, Laura Ciccolallo, Franco Berrino, Lisa Licitra, Paolo Casali, Samba Sowe, Angelo P Dei Tos, Joanne Fleming, Ellen Benhamou, Riccardo Capocaccia, Dirk Schrijvers, Harry Schouten, Patrizia Olmi, Stefano Ferretti, Marine Grossgoupil, Ségolène Aymé, Hildrun Sundseth, Giuliano Buzzetti, Marc Maynadie, Pascale Blaes, Rudiger Hehlmann, Simon Baconnier, Juan Antonio Virizuela-Echaburu, Ricardo Gonzalez-Campora, Sandrine Marreaud, Jan Maarten Van der Zwan, Michael Schaapveld, Carmen Martinez, Annet Sollie, Jean-Michel Lutz, Ian Kunkler
Unable to attend: Törgil Moller, Domenica Taruscio, Bernadette Brennan, Andres Cervantes, Alessandro Gronchi, Alba Brandes, Isabelle Ray Coquard, Angela Rincon, Pancras Hogendoorn, Jean-Pierre Droz, Jan-Willen Van De Loo, James Mackay.

After the approval of the agenda, a brief overview of the project was given by the project leader focusing on the two major deliverables for which the contribution of the consensus group is required. These are:

- Definition and list of rare tumours (to be done by the 18th month of the project)
- Estimation of burden indicators for all the rare tumours included in the list (to be done by the 24th month of the project).

Population-based cancer registries are the source for estimating the indicators, and the International Classification of Diseases for Oncology (ICDO) is utilized for the identification of the tumour entities. Comments received from Torgil Möller, a RARECARE partner who was unable to attend the meeting, were briefly explained. He proposed morphology as the main criterion. He also suggested to use a uniform criterion for the definition of topographic sites.

Prof. Marc Maynadie was invited at the meeting to talk about the HAEMACARE project, within which he is responsible for WP 4 - Integration of Clinical and Cancer Registries Data. This is also a DG Sanco funded project. In his presentation, he showed the haematological entities that have been defined by HAEMACARE, based on cancer registry data. After the discussion, the group agreed to adopt the same guidelines for the identification of rare haematological malignancies.

Dr. Paolo Casali summed up the results of the 1st consensus meeting held in Brussels. He stressed that a lot of work was done, and that we are close to a definition. However, he said that a distinction needs to be made between tumor entities and groups of tumors. Tumour entities are relevant for clinical decision-making. Group of tumours are relevant for patient referral and health care organization (where efficacy from clinical trials must

be translated into effectiveness). He argued that for a group of tumours that is rare as a whole, clinical expertise cannot be widespread, and effectiveness may be problematic (availability of clinical expertise needs to be improved by centralization, networks, etc.). On the contrary, when a disease entity is rare as such, but falls within a frequent group of tumors, the clinical expertise is not a problem, although clinical studies may be difficult. Thus, he suggested to have two hierarchies in the list: groups of tumors and tumors, both clinically relevant, with ICDO tumor entities as a third hierarchy. The threshold for rarity may be the same for both, or differentiated (for example 5 x 100,000/year for groups of tumours and 3 x 100,000/year for single tumours, or 5 x 100,000/year for both).

Criteria to identify groups of tumors and tumors in the first two hierarchies will be always the combination of morphology and topography. In any case, tools will be available to make groupings either by topography and by morphology. The importance to include all the cancer sites and morphologies of the ICD-O classification in the final list was also confirmed. As exception, some unspecific cancer sites or morphologies (such as 'carcinoma NOS' or 'malignant neoplasms') could be removed from the list, since they have been just used by registries to codify uncertain cases, but of course do not correspond to any entity.

Dr Paolo Dei Tos proposed a preliminary list of the first hierarchy (groups of tumours). The participants then revised the classification and the identified rare tumours entities within the epithelial cancers.

The discussion continued on the relevance of incidence and prevalence. The group of experts expressed a consensus on the appropriateness of an incidence-based definition of rare tumours. However, both incidence and prevalence are important. RARECARE will provide prevalence data, along with incidence and survival data. Dr. Ségolène Aymé stressed the importance of the definition of rare diseases based on prevalence for the orphan drugs regulation. She went further to admonish the experts group that any final decision on the definition list shouldn't endanger the existing regulations of orphan drugs. The experts group fully agreed on the importance of orphan drugs regulations and said much attention will be paid to avoid endangering such regulations.

It was concluded that a formalised rationale for the definition of rare tumours with the relative list of rare tumours and their incidence rates will circulate at the end of June among all the experts and the partners of the project. The documents will be also available on the RARECARE web site. Comments are expected for the end of July.

Issues on how to improve the consensus among the cancer societies and the patient groups were discussed from the leader of the project. She said the definitive documents will be circulated to the most important European societies or networks (ESTRO, ESSO, ESMO, SIOP, EORTC, European Leukemia Network (ELN)) and the ECPC for patients.

In conclusion, the discussion centred on strategies for the dissemination of the final results. The experts suggested that they should be provided at relevant congresses of Oncologists, Haematologists, Health planners and Patients, and to publish a scientific paper. The project leader was therefore asked to request for a session at the forthcoming ECCO/ESMO meeting in Berlin next year and to prepare a presentation for the ELN symposium (Feb, 2009) in Mannheim, Germany. Prof. Hehlmann will send an official invitation to the project leader.

It was also suggested to prepare a paper for patients. On this subject, Hildrun of the ECPC and Sandrine of the EORTC both agreed to prepare a communication plan for the dissemination of RARECARE results to patients.