RARECARENet country report

Ireland

23 May 2016

EUROPEAN CANCER PATIENT COALITION

nccp National Cancer Control Programme

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1. Introduction

In 2011 the RARECARE (surveillance of rare cancers in Europe) project proposed a new definition for rare cancers and quantified the burden of rare cancers in Europe. RARECARE showed that rare cancers pose an important challenge in Europe. They are thought to represent nearly one quarter of all new diagnoses of cancer in Europe (22%) and have lower survival rates than common cancers. Thus obtaining reliable estimates of the number and type of rare cancers, and identifying ways to improve the quality of care for patients should be recognised as a public health priority.[1]

A second project, **Information Network on Rare Cancers (RARECARENet)** was established in 2012 with the aim **to build an information network that may provide comprehensive information on rare cancers to the community at large.**

The RARECARENet project had 3 main components:

- **European and country-specific epidemiological indicators** were estimated on the basis of population-based cancer registries (94 cancer registries from 24 European countries corresponding to 46% of the population of the European Union).
- A '**Pilot Study on hospital volume**' examined the extent of rare cancer treatment centralisation, and looked at the association between hospital volume and outcomes for two types of rare cancers – sarcomas and head and neck cancers.
- A wide consensus process together with a '**High resolution study on Centres of Expertise for rare cancers'** identified quality criteria for centres of expertise in the management of rare cancers.

*(For a fuller description of the RARECARENet project, see **Appendix 1**).*

In addition, multi-stakeholder meetings were organised in four countries (Belgium, Bulgaria, Slovenia and Ireland) to discuss the emerging public health issue of rare cancers in each country. These meetings allowed the RARECARENet team to present country-specific findings to participating local experts, and discuss how the management of rare cancers may be improved locally for the benefit of patients.

The purpose of this report is to summarise the main discussions and recommendations from the RARECARENet meeting that took place in Ireland at the offices of the National Cancer Control Programme (NCCP_ on 2nd September 2015. The meeting gathered European and local experts on rare cancers (epidemiologists, oncologists, surgeons, and pathologists), cancer registry leads, patient organisations and representatives of the NCCP and was co-hosted by the European Cancer Patients Coalition and the National Cancer Control Program. (*The meeting agenda and the list of participants is provided in* **Appendix 2***).*

Discussions focused on four key priorities which correspond to the objectives of the RARECARENet project:

- Describing the epidemiology of rare cancers in Ireland
- Discussing the most appropriate quality indicators for centres of expertise for specific rare cancers
- Discussing the availability of centres of treatment for rare cancers in Ireland
- Identifying existing challenges in the management of rare cancers in Ireland.

2. The epidemiology of rare cancers in Ireland

Rare cancers account for 17% of all cancer diagnoses in Ireland

The incidence of rare cancers in Ireland, grouped by different crude incidence cut-off rates, can be seen in **Appendix 3**.

Ireland has 5,270 new cases of rare cancers per year, accounting for 17% of the total cancer diagnoses. This much smaller than the average across Europe (22%), and among the lowest proportion across the countries studied by RARECARENet. The small numbers of cancers involved will have important implications for planning and management of these diseases.

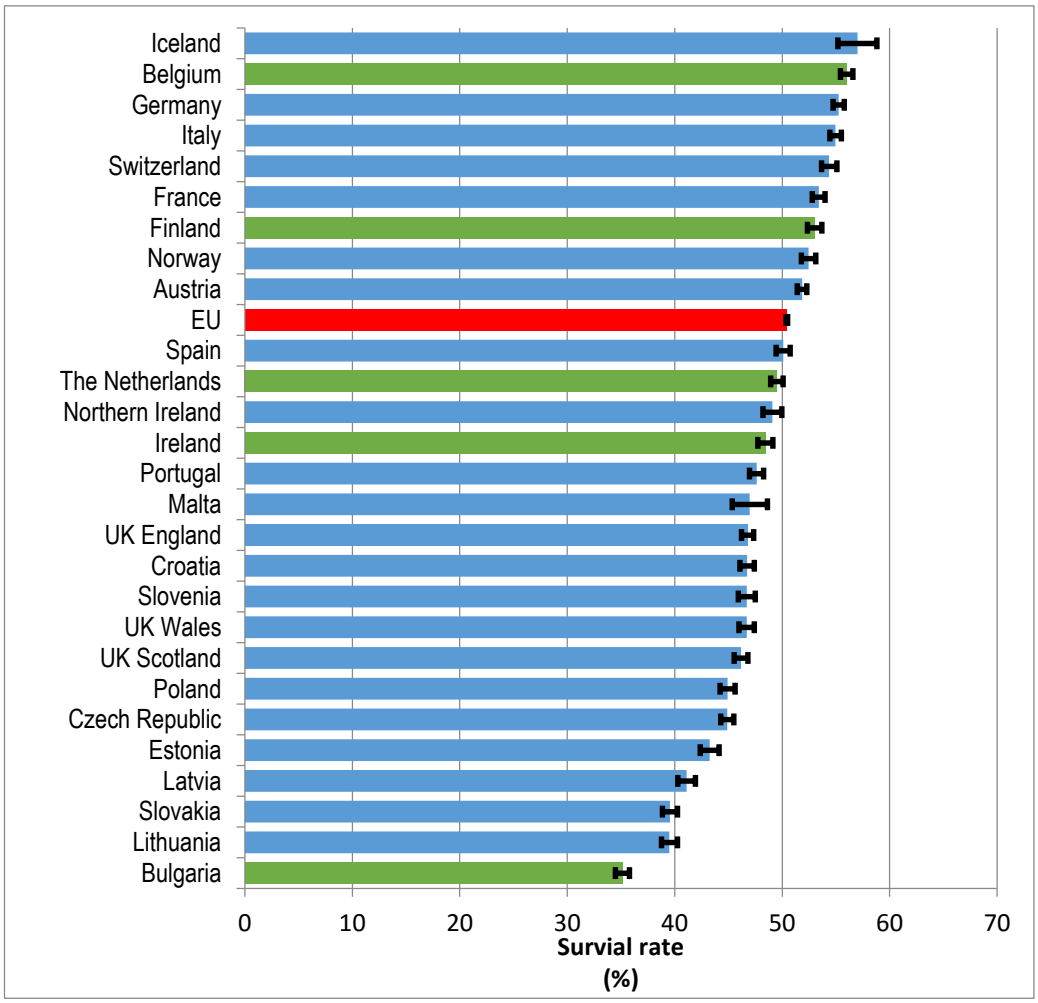
Utilising the RARECARE threshold of <6 per 100,000 incidence per year, all rare cancers in Europe are rare also in Ireland.

The survival of people who are diagnosed with a rare cancer is lower than that of people diagnosed with a common cancer.

Using the RARECARENet database, survival analysis for rare cancers was conducted per country. Data from Ireland follow a similar trend observed across all countries: survival rates for rare cancers are lower than those for common cancers.

Figure 1 shows the 5-year survival rates for rare cancers across different European countries. Importantly, rare cancers in Ireland is lower than the average for EU countries, and lower than many other countries studied in detail by RARECARENet (**Figure 1**).

Figure 1 - 5-year survival rates for rare cancers shown in European countries. Survival is adjusted by age and case mix. Countries studied in detail by RARECARENet are shown in green.



3. Quality indicators for centres of expertise for rare cancers

Criteria for centres of expertise in the management of rare cancers

As mentioned previously, one of the main outputs of the RARECARENet project was the specification of general quality criteria for centres of expertise in the management of rare cancers. These criteria were then used to develop specific quality indicators for different cancer types (see **Appendix 4**). The general quality indicators were developed at a European level, based on a consensus approach, utilising both evidence from previous experience at the European level (the European Commission committee of experts on rare diseases) and country level.

According to the general quality criteria, centres of expertise should:

1. Ensure appropriate **clinical management** according to evidence-based guidelines and based on **a patient-centered multidisciplinary approach**
2. Develop/support **patient pathways** nationally and across borders

3. Develop **clinical guidelines** and **good service delivery guidelines**
4. Guarantee a learning environment by **promoting training** and continuing education activities
5. Foster clinical, translational and epidemiological **research for rare cancers**
6. **Empower patients** by providing information adapted to their specific needs, culture and ethnic group
7. Possess an **e-health platform** to process and share information, biomedical images or clinical samples supported by enough human and structural resources as well as equipment
8. Have a transparent quality assurance system including **monitoring and evaluation** of the service provided
9. Consistently **report the volume** of patients treated for rare cancers.

These general quality criteria for centres of expertise aided the development of **specific quality criteria for the clinical management of rare cancers**. Key aspects of clinical management relevant to all rare cancers are presented in **Table 1**. These aspects are important to consider in identifying and evaluating centres of expertise. Specific indicators, based on these key aspects, relevant to two particular rare cancers (soft tissue sarcomas and head and neck cancers) are presented in **Appendix 4**.

Table 1: Main areas of the clinical management from which rare cancer specific quality criteria for centres of expertise have been proposed.

Critical areas	**Why is this important for rare cancers?**
Appropriate and timely diagnosis and staging procedures	Due to the rarity of these cancers, many doctors are unable to make a prompt diagnosis, and pathologists may not have enough experience to identify the cancer. A high level of experience is also necessary for performing all correct staging procedures. The lack of experience due to the rarity of these tumours leads to incorrect or delayed diagnosis and poor staging procedures. This may be compounded by the lack of clear referral pathways, since centres dedicated to rare cancers may not exist or may not be known to general practitioners and other healthcare professionals.
Quality of care	Lack of knowledge and clinical expertise among health care professionals treating rare cancers may result in suboptimal care.[1] Because of the rarity, it is difficult to perform clinical trials and therefore develop evidence-based clinical guidelines. When available, the adherence to clinical guidelines is critical to ensure patients receive the most appropriate, evidence-based care. Independent of clinical guideline availability, it is important to have clinical management agreed upon by a multidisciplinary team, as it ensures a higher quality of care for rare cancer patients.
Quality of pathological report after surgery	Pathological reports are essential for planning appropriate treatment. Ideally, pathology reports should contain a full set of the core data defined by the relevant scientific society guidelines, however a complete set of these standardized data are often missing in reports, challenging the definition of the appropriate treatment plans.
Quality surgery and radiotherapy	Due to the rarity of these cancers, it is difficult to find experienced surgeons and/or radiotherapists. Poor quality surgery and radiotherapy

	can lead to re-operation or recurrences, which are avoidable and may have a high impact on the quality of life for patients.
Availability of formalised multidisciplinary decision-making	Multidisciplinary teams are important for the management of rare cancers,[2] as they allow health care professionals from a range of disciplines to work together to provide a comprehensive treatment plan for patients. This is of particular importance for rare cancers due to the limited availability of scientific evidence. Additionally, multidisciplinary teams in high volume centres have been associated with better outcomes, shorter delays between diagnosis and treatment and a greater availability and range of therapeutic options.[2]
Participation in clinical and translational research	Given the small number of new rare cancer cases each year, it is often difficult to obtain a strong evidence base on which to build clinical management guidelines and protocols. Therefore centres of expertise should network with other centres across Europe to participate in clinical trials, with the aim to improve accuracy and standardisation of diagnosis and treatment for rare cancers. [1]

4. Treatment centres for rare cancers in Ireland

Centralisation of care is viewed as critical for the management of rare cancers to allow specialists and health professionals to diagnose and treat a larger number of these rare cases. Greater centralisation of the treatment of cancers into centres of expertise is one of the explicit priorities stated in Irish Cancer Service strategy for 2006-2015. Hospital services were to be reorganised allowing centralisation to 8 cancer services, and multidisciplinary teams forming the cornerstone for the treatment of cancer.

Rare cancers differ in their degree of centralisation in Ireland, as is illustrated in Figures 2 and 3. Comparing between the main treatment hospital volume, **Figures 2 and 3**, head and neck cancer treatment is more centralised than soft tissue sarcoma. Surgeries for soft tissue sarcomas were performed in more than 60 hospitals across the country. The majority of these hospitals were preforming surgery on less than 2 patients per year and even the hospital treating the highest volume of sarcoma patients was still a comparatively low volume, with only 12 patients per year
The data produced by the RARECARENet pilot study refers to the period between 2000 and 2007. This was before the Irish Cancer Service strategy for 2006-2015 which supported centralisation to eight cancer services across Ireland. However, it did not address the issue of rare cancer centralisation in detail. The Irish experts present during the meeting, **(appendix 2)**, advocated for formal recognition of centres of expertise for rare cancers.

Figure 2 - Hospital volumes for main treatment, radiotherapy and surgery for head and neck cancers in Ireland. Hospitals were ranked by decreasing volume after blind coding. The main treatment is defined as preforming the surgery for tumours of the oral cavity and salivary gland; radiotherapy for tumours of larynx, oropharynx, and hypopharynx.

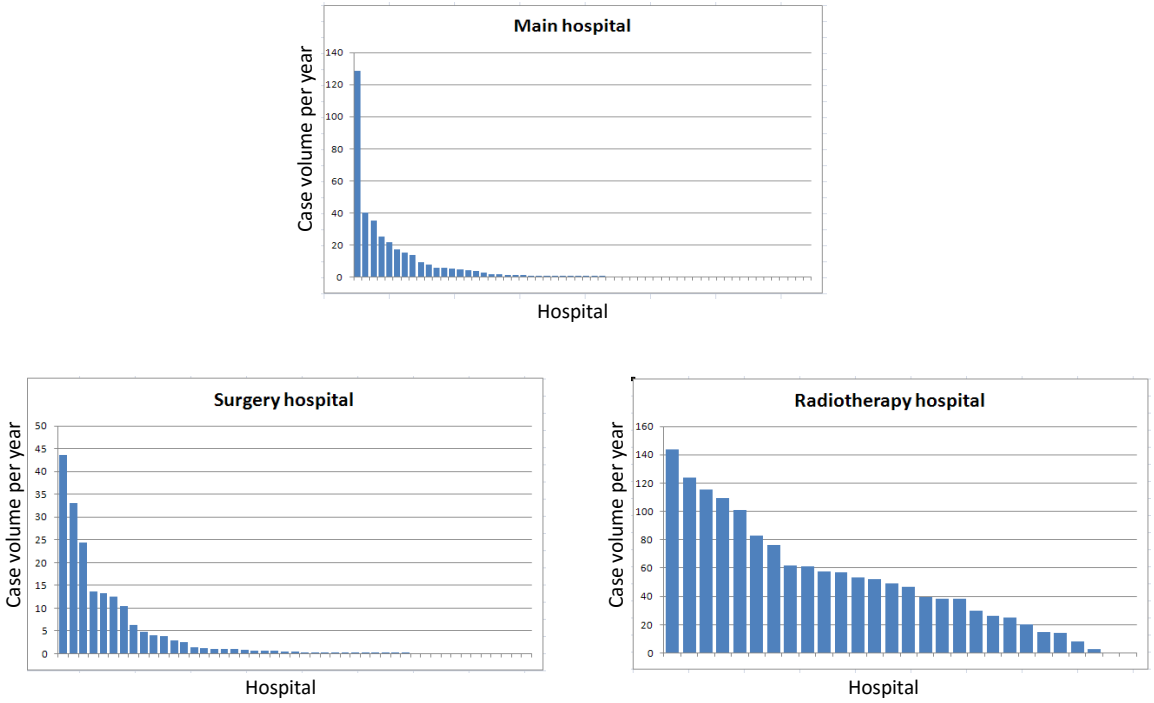
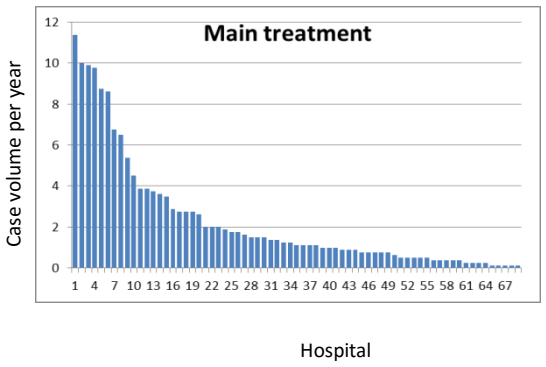


Figure 3 - Hospital volume for main treatment of soft tissue sarcoma in Ireland. Hospitals were ranked by decreasing volume after blind coding. The main treatment is surgery for soft tissue sarcoma.



Due to the comparatively small population in Ireland and the small number of soft tissue sarcoma patients managed in each centre, challenges are posed for healthcare professionals to develop a wide experience base and provide the highest quality of care.

Results from the pilot study in Ireland for head and neck cancers, which assessed the relationship between hospital volume and survival, demonstrated that hospital volume was positively associated with outcome for patients with advanced cancer. Patients with advanced stage head and neck cancer in low volume hospitals (<5 cases/year) had a double risk of death compared to those treated in high volume hospitals (>100 cases/year).

The EU Joint Action "Comprehensive Cancer Control" (CanCon) was established to reduce cancer mortality and improve survival, through identifying quality standards and guidelines for care, improving quality of life for patients and facilitating cooperation between member countries. Following the establishment of CanCon, the European Cancer Patients Coalition (ECPC) conducted a survey of 23 CanCon EU Member-State representatives to further understand the referral for rare cancers in each country represented in CanCon and whether specific treatment centres had been identified. However, it was beyond the scope of the RARECARENet project to determine specific centres of expertise for rare cancers in Ireland, therefore it focused on developing quality criteria to support their selection in the future.

Mrs. Conroy, from the Cancer, Blood and Organs Policy Unit, provided a list of centres which provided treatment for rare cancers in Ireland, featured in **Appendix 5.** For several rare cancers, discussions are still taking place to reach a consensus on location of Irish services in the future and how to strike the balance between centralisation to centres of expertise and proximity to home.

5. Challenges in the clinical management of rare cancers in Ireland

A. General challenges in the management of rare cancers in Ireland

The management of rare cancers generally poses challenges, due to their small numbers and lack of available specialists. Some of the most important challenges raised in Ireland for the management of rare cancers confirm the relevance of the quality criteria chosen and the importance of centralised care. They included:

- **Delays in diagnosis,** due to limited experience from pathologists, lack of centralisation and the complexity associated with diagnosing certain rare cancers.
- **Poor access and availability to radiotherapy and other equipment vital to the treatment of cancer**, including limited access to key facilities such as magnetic resonance imaging (MRI), Computerised tomography (CT) and positron emission tomography (PET), delaying the initiation of treatment
- **Poor access to a formalised multidisciplinary team**
- **Lack of experienced surgeons for the often highly complex rare cancer surgeries,** as there is limited centralisation of treatment
- **Low levels of centralisation of treatment**, challenging the provision of continuous, high quality care for every patient
- **Low level of involvement in the translation of clinical research** into day-to-day management of rare cancers.

B. Specific challenges in the management of sarcomas and head and neck cancers in Ireland

The following challenges were highlighted during the meeting for the management of sarcoma, head and neck, neuroendocrine and testicular cancers, however the High Resolution Study only covered sarcoma and head and neck cancers in Ireland. Additionally, the High Resolution Study looked at the time period 2000-2007, whereas the challenges discussed during the presentations (Appendix 2) covers the current state of play for rare cancers in Ireland.

*(Details from the High Resolution Study on sarcomas and head and neck cancers can be found in **Appendix 4**).*

Table 2: Main challenges in the management of soft tissue sarcoma, head and neck, testicular and neuroendocrine cancers in Ireland.

i) Soft tissue sarcomas

Area for improvement	**Identified challenges**
Diagnostic management	Limited awareness of the importance of adequate imaging for diagnosis of rare cancers. In 2009-2010, 39% of sarcoma patients did not have a biopsy carried out before treatment decisions were made, however since then it is hoped that this proportion has decreased.
Radiology	Limited access to MRI, CT scan and PET facilities within the country challenges appropriate staging and treatment.
Treatment	RARECARENet studies found highly variable use of adjuvant therapies. There were no defined treatment pathways for sarcomas, since centres officially designated as national centres for sarcoma do not exist. More recently despite improvement, there are still patients being treated at multiple units, which may not be cancer centres, and there is still highly variable use of adjuvant therapy.
Pathology diagnosis	Very poor centralisation of pathology diagnosis. Fresh tissue is rarely collected. No sarcoma cytogenetics/molecular diagnostics is available outside Our Lady's Children's Hospital thus tissue must be frequently sent overseas.
Centralisation	Treatment poorly centralised (**Figure 3**). However, recently the Irish sarcoma group has found the levels of centralisation have been increasing, with 2-3 hospitals performing the majority of sarcoma surgeries for sarcoma of extremities, gastro intestinal stromal tumours and bone. However, for other types of sarcomas surgeries are still much less centralised.
Multidisciplinary teams	Only 2 hospitals currently have regular sarcoma-specific multidisciplinary teams, however they are sometimes unable to manage all patients referred with bone or soft tissue sarcomas.

ii) Head and neck cancers

Area for improvement	**Identified challenges**
Timely start of treatment	Delays are common for both the initiation of curative treatment and adjuvant treatment. The delay seemed more frequent if the main conventional treatment is radiotherapy.
Centralisation	Low level of centralisation for this type of cancer, as illustrated in **Figure 2.** Apart from one single hospital to which radiotherapy is strongly centralised.
Pathology reporting	There is often poor quality pathology reporting, challenging post-surgery treatment planning.
Multidisciplinary teams	Not all patients see the full spectrum of health care professionals required for these complex cancers, although a multidisciplinary

	approach is vital due to the unusual nature of these pathologies and complicated surgeries.
Quality of care	There is often a long delay in starting treatment, associated with poor access to radiotherapy and chemotherapy.

iii) Testicular cancers

Area for improvement	Identified challenges
Centralisation	There is poor centralisation of treatment, with approximately 20 hospitals providing care. Particularly, retroperitoneal lymph node dissection (RPLND) needs to be better centralised.

iv) Neuroendocrine tumours

Area for improvement	Identified challenges
Poor referral practices	Lack of national acceptance of centralisation, many physicians do not refer cases to centres of expertise.
Resource availability	There is a lack of junior staff, which may hamper the development of future experts and this might prove to be a significant barrier in the future. Previously there had been a lack of gallium for PET and CT scans, resulting in long waiting times or inadequate diagnostic procedures. However this problem is now hopefully resolved.
Research and clinical trials	Due to the rarity of these cancers, clinical trials are difficult to perform creating challenges in the development or implementation of innovative treatment and health system resource planning.

6. Recommendations for the future

Discussions during the meeting identified a number of key recommendations for the improved management of rare cancers in Ireland. These overarching recommendations for rare cancers can be found below:

1. Improve the standardisation of care for rare cancers to reduce treatment inequalities across centres and improve patient quality of care

    - **Centres of expertise for rare cancers must be identified** and this process should be supported by the Ministry of Health
    - **Centralise care to high volume hospitals,** particularly for complex cases and management of rare cancers should be discouraged in non-specialist centres
    - **Ensure that primary treatment is planned at a multidisciplinary meeting** to improve collaboration among different specialists, quality of pathology reporting and timely start to treatment
    - **Implement national and international pathways for rare cancers, setting minimum standards of quality for services** particularly for pathology and radiology, following the examples set by sarcomas and NETs

2. Empower patients to take a stronger role in their care

    - **Improve communication between clinical experts and patient representatives** to create a stronger local advocacy base for the better management and centralisation of rare cancers.

3. Awareness campaigns

    - **Organise rare cancer awareness and prevention campaigns for the general public**
    - **Awareness and education campaigns may empower general practitioners to improve follow up and better refer to expert centres,** increasing timely and appropriate diagnosis

4. Increase the research base and collaboration in rare cancer care

    - **Use a population-based cancer registry** as the data source to identify and monitor centres of expertise
    - **Involve scientific societies in the discussion on quality indicators**
    - **Utilise European Reference Networks** to enable cross-border collaboration for clinical management, second opinions and clinical research
    - **Improve translational research** to drive innovation and improve the dissemination of best practice amongst centres of expertise
    - **Include rare cancers as a priority within the cancer control strategy.**

Appendices

Appendix 1: About the RARECARENet Project

RARECARE (Surveillance of rare cancers in Europe) data provided a first understanding of the burden posed by rare cancers. RARECARENet project aims at create an information network to provide and disseminate comprehensive information on rare cancers to oncologists, general practitioners, researchers, health authorities, patients and the general public. Additionally, RARECARENet aim to further develop a comprehensive list of patient associations which are dedicated to rare cancers. These objectives are carried out with the eventual aim to improve the timeliness and accuracy of diagnosis, facilitate access to high quality treatment for patients with rare cancers, to identify centres of expertise for rare cancers in Europe and standardise practice across member states. The data facilitating these improvements has been found through the following studies:

RARECARENet EUROCARE-5 is an adult database which was created to update the epidemiological indicators for rare cancers. It covers 94 cancer registries (89 of which were in RARECARE) in 24 countries (19 of which participate in RARECARE). The database covers 48% of the population of the countries participating in RARECARENet, and 46% of the population in the European Union (excluding Norway, Switzerland and Iceland). This database was used to calculate incidence, prevalence and survival of rare cancers.

RARECARENet 'Pilot Study on hospital volume' investigated the extent of centralisation of rare cancer treatment in selected European countries on the basis of population based cancer registries with national coverage. Countries with national coverage were Belgium, Bulgaria, Finland, Navarra (a region of Spain), Ireland, Slovenia and the Netherlands. Objectives of this study were to estimate indicators of the degree of centralisation, map the hospitals where rare cancers are most frequently treated, and analysing the association between hospital volume and outcome for select rare cancers.

RARECARENet 'High resolution study on Centres of Expertise for rare cancers' looked to identify quality criteria for centres of expertise for the management of rare cancers. Through discussion with clinicians, experts, epidemiologists, patient representatives, and cancer registries, general criteria for centres of expertise and specific indicators for selected rare cancers (sarcomas, testicular and head and neck tumours) were identified. The cancer-specific indicators were analysed to test their appropriateness on a retrospective analysis of studying hospital patient files and pathology reports in selected participating cancer registries from Belgium, Bulgaria, Finland, Ireland, Italy, Slovenia and the Netherlands.

RARECARENet 'Information for patients and professionals' was created to provide meaningful information to the community at large. A list of clinical guidelines on rare cancers was developed based on already available information and on new information collected and created in collaboration with State-of-the-Art Oncology in Europe (START), the European Society for Medical Oncology (ESMO) and epidemiological data from the project, Surveillance of Rare Cancers in Europe. The RARECARENet website also provides a list of information materials on rare cancers for patients on the diagnosis, treatment and follow-up of any type of rare cancer. The information has been collected from rare cancer patient organisations participating in the project, more information can be found on the RARECARENet website - http://www.rarecarenet.eu/rarecarenet/index.php/information-on-rare-cancers

Finally, a list of 144 rare cancer patient organisations in Europe was created with the aim to build a network to support patients with rare cancers. The list is available on the RARECARENet website - http://www.rarecarenet.eu/rarecarenet/index.php/patient-organisations

Appendix 2: Agenda for the Dublin meeting on Rare Cancers

Meeting Agenda

RARECAREnet Meeting on Results of the High Resolution Studies in Ireland and on Quality of Care for Rare Cancers

September 2nd, 2015, 9:00–16:00
Location: National Cancer Control Programme Offices,
King's Inns House, 200 Parnell Street, Dublin

Time	Topic	Responsibility
9:00-09:30	**REGISTRATION AND COFFEE**	
09:30–09:45	**Welcome, introduction round, program and aim of the meeting**	RARECARE team, Dr. Jerome Coffey, NCCP
09:45–10:15	**RARECAREnet project overview**	Gemma Gatta, RARECAREnet
10:15–11:40	**Presentations on the situation in the country on rare cancers:** - **Clinical situation/organisation** - **Rare Cancer Patient story** - **Tumour specific Services**	Mr. Michael Conroy, Dept of Health Dr. Jerome Coffey, NCCP Dr. Charles Gillham (Sarcoma) Dr. Dermot O'Toole (NET) Dr Paul Sweeney (Testes) Mr. Leo Stassen (Head & Neck) Mr. John Langton, Patient
11.40-12.00	**COFFEE BREAK**	
12.00-12.30	**Presentation of Quality Criteria**	Annalisa Trama, RARECAREnet
12.30-13.00	**Discussion on the Quality Criteria proposed**	Annalisa Trama, Charles Gillham
13.00-13.45	**L U N C H**	
13:45-14:15	**Results of the Volume Analyses of the Country**	Riccardo Capocaccia, RARECAREnet
14:15 -14:45	**Discussion on the Results of Volume Analyses**	
14:45-15.00	**Patient Support: Treatment Centres and Patient Organisations in Ireland**	Kalliopi Christoforidis
15.00-16:00	**ROUND TABLE:** **Discussion on the Level of Centralization and Hospital Volume**	Dr. Jerome Coffey, NCCP RARECARENet team
	Conclusions and Way Forward	
16:00	**Close of the Day**	

List of participants:

Dr. Jerome Coffey – Host and Director of NCCP
Dr. Mary Hynes – Deputy Director NCCP
Anna Lisa Trama – RARECARENet Team
Riccardo Capocaccia - RARECARENet Team
Gemma Gatta - RARECARENet Team
Kalliopi Christoforidi - RARECARENet Team
Pat Cafferty - NCCP
Maeve Cusack - NCCP
Marie Cox - NCCP
Dr Marie Laffoy - NCCP
Dr Deirdre Murray - NCCP
Eileen Nolan - NCCP
Prof. A. Hill - NCCP
Mr. Leo Stassen – St James Hospital, Dublin
Dr. Charles Gillham – St Luke's Radiation Oncology Network, Dublin
Prof. Dermot O'Toole – St James Hospital, Dublin
John Thornhill – AMNCH Hospital, Dublin
Catherine Dowling
Dr. Dearbhaile O'Donnell – St James Hospital, Dublin
Dr. John McCaffrey – Mater Hospital, Dublin
Michael Conroy – Department of Health, Dublin
Keith Comiskey - Department of Health, Dublin
Dr. John Kennedy – St James Hospital, Dublin
Dr. Harry Comber – National Cancer Registry, Ireland
Donal Buggy – Irish Cancer Society
Naomi Fitzgibbon – Irish Cancer Society
John Langton – Patient representative
Dr Tadgh O' Dwyer – Mater Hospital, Dublin
Eibhlin Mulore – Irish Clinical Oncology Research Group, Dublin
Paul Sweeney, Mercy University Hospital, Cork

Appendix 3: Rare cancer incidence in Ireland (estimated new cases, 2013)

The number of estimated cases in 2013 is estimated applying crude incidence in Europe 2000-2007 to the Irish population at 1st January 2013 (EU population estimated by EUROSTAT; see 'population EU'). The rare cancer entity are ranked on the basis of the incidence rate in Ireland.

Rare cancer entity	**Crude incidence rate per 100,000/year**	**Estimated new cases in 2013**
Rare lymphoid diseases	3<incidence<6 ‘Rare cancers’	729
Rare epithelial tumours of oesophagus		260
Rare epithelial tumours of ovary and tubes		279
Tumours of central nervous system		240
Epithelial tumours of cervix uteri		240
Soft tissue sarcoma		192
Epithelial tumours of hypopharynx and larynx		241
Myeloproliferative neoplasms		129
Testicular and paratesticular cancers		150
Neuroendocrine tumours		139
Epithelial tumours of breast		156
Epithelial tumours of gall bladder and EBT		161
Acute myeloid leukaemia and related precursor neoplasms		150
Myelodysplastic syndrome and myelodysplastic/myeloproliferative diseases		92
Epithelial tumours of oral cavity and lip	0.5<incidence<3	190
Carcinomas of thyroid gland		214
Epithelial tumours of lung		173
Epithelial tumours of liver and intrahepatic bile tract		180
Epithelial tumours of oropharynx		137
Epithelial tumours of vulva and vagina		69
Epithelial tumours of major salivary glands and salivary-gland type		56
Malignant melanoma of uvea		28
Bone sarcoma		37
Epithelial tumours of anal canal		44
Epithelial tumours of small intestine		30
Malignant mesothelioma		84
Epithelial tumours of pelvis and ureter		60
Rare epithelial tumours of corpus uteri		25
Epithelial tumours of penis		27
Epithelial tumours of bladder	Incidence<0.5 ‘Very rare cancers’	25
Adnexal carcinoma of skin		11
Epithelial tumours of nasal cavity and sinuses		18
Epithelial tumours of nasopharynx		20
Embryonal tumours of central nervous system		11
Rare epithelial tumours of stomach		13
Non epithelial tumours of ovary		11
Nephroblastoma		8
Malignant melanoma of mucosa (extracutaneous)		5
Neuroblastoma and ganglioneuroblastoma		7

Gastrointestinal stromal sarcoma		12
Epithelial tumours of thymus		7
Rare epithelial tumours of colon		5
Carcinoma of adrenal gland		9
Extragonadal germ cell tumours		6
Kaposi's sarcoma		10
Carcinomas of pituitary gland		1
Rare epithelial tumours of rectum		4
Rare epithelial tumours of prostate		24
Retinoblastoma		4
Rare epithelial tumours of pancreas		3
Epithelial tumour of trachea		4
Hepatoblastoma		1
Epithelial tumours of urethra		5
Epithelial tumours of eye and adnexa		2
Carcinomas of parathyroid gland		1
Histiocytic and dendritic cell neoplasms		2
Rare epithelial tumours of kidney		2
Olfactory neuroblastoma		1
Trophoblastic tumour of placenta		1
Epithelial tumours of middle ear		1
Odontogenic malignant tumours		0
Pleuropulmonary blastoma		0
Pancreatoblastoma		0

Appendix 4: Quality indicators and outcomes for two chosen rare cancers in Ireland.

Soft tissue sarcoma:

Criteria	Quality indicator	Findings from the high resolution study in Ireland
Diagnostic management	Percentage of patients with sarcoma undergoing preoperative scan and biopsy before treatment (MRI and/or CT locally and lung CT)	39% of sarcoma patients received a treatment without a histological verification.
	Diagnosis done by an expert pathologist (or second opinion carried out in an expert centre if diagnosis is not carried out by an expert pathologist)	Data not available at the cancer registry. It was observed that the second opinion was mainly asked by small volume hospitals
Adherence to clinical guidelines	Percentage of patients with low grade and R0 resection margin undergoing surgery alone.	65% of low grade, R0 sarcomas underwent surgery alone. In the other countries included in the RARECARENet study, the proportion was higher ranging from 72% to 100%.
	Percentage of patients with high grade and R0 resection undergoing surgical intervention and radiotherapy or radiotherapy and chemotherapy.	60% of high grade, R0 patients underwent surgery with radiotherapy or with radiotherapy and chemotherapy. In the other countries included in the RARECARENet study, the proportion was lower ranging from 20% to 63%.
	Percentage of patients with R1 or R2 resection margin undergoing surgical re-intervention or, radiotherapy, or chemotherapy and radiotherapy.	76% of R+ patients underwent re-intervention or radiotherapy or radiotherapy and chemotherapy. In the other countries included in the RARECARENet study it ranged from 68% to 73%.
Quality of surgery	Complete tumour resection of definitive surgery	22% of surgeries were R+ (i.e. incomplete surgical resection) and 6% R level was missing or unknown. Of the R+ surgeries, 29% were in low volume hospitals and 24% in high volume hospitals. (In Belgium and in Slovenia, it was very clear that R+ surgeries were mostly from low volume hospitals).
	Reoperation after primary definitive surgery	The reoperation rate after primary definitive surgery was 23% and were mainly performed in small volume hospitals
Quality of pathology report after surgery	Percentage of pathology reports with a full set of core data items recorded according to the ESMO guidelines (site, size and grade)	37% of pathology reports provided all required information. In the other countries included in the RARECARENet study, the proportion ranged from 56% to 72%.
Availability of formalised multidisciplinary decision making and care		No indicators collected by high resolution study because the information was not retrievable in the medical records.

Participation in clinical and translational research		No indicators collected by high resolution study because the information was not retrievable in the medical records

Head and neck:

Criteria	Quality indicator	Findings from the high resolution study in Ireland
Timely start of treatment	Time to start treatment (time between definitive diagnosis by a pathologist and beginning of surgery or radiotherapy)	61% of head and neck patients received treatment over one month after diagnosis. Other countries included in the RARECARENet study ranged from 44% to 63%.
		Of patients starting treatment over a month after diagnosis, 68% of this were for radiotherapy, and 32% was surgery.
		Of those who were begun in over a month after diagnosis, 28% were early, 51% were advanced and 1% was metastatic. 20% of the data was missing however.
	Time in starting postoperative radiotherapy or concomitant chemo-radiotherapy (adjuvant treatments)	67% of adjuvant treatment occurred after 8 weeks. The other countries included in the RARECARENet study ranged from21% to 46%.
Stage at diagnosis	Proportion of patients with a definitive stage at diagnosis	For cancer of the larynx - 40% were localised, 35% advanced, 3% metastatic and for 22% the information was missing.
		For all other sites (oral cavity, oropharynx, hypopharynx) – 17% was localised, 55% advanced, 4% metastatic and for 24% of the patients information was missing.
Adherence to clinical guidelines	Percentage of patients with early stage I and II referred for either surgery or radiotherapy	For cancers of the larynx which are localised: 25% had only surgery, 11% had surgery and radiotherapy or surgery and concomitant radio-chemotherapy. 4% had concomitant chemo-radiotherapy, and 6% had only radiotherapy. In other countries included in the RARECARENet study, the percentage of patients with early stage I and II referred for surgery ranged between 12% and 67%. Patients referred for radiotherapy ranged between 12% and 64%.

	Percentage of patients with locally advanced stage III and IV referred for surgery plus postoperative radiotherapy or postoperative chemo-radiotherapy	For cancers of the larynx which are advanced - 12% had only surgery, 14% had surgery and radiotherapy or surgery and concomitant radio-chemotherapy. 30% had concomitant chemo-radiotherapy. 26% had only radiotherapy. In other countries in the RARECARENet study, the percentage of patients with locally advanced stage III and IV referred for surgery plus post-operative radiotherapy or chemo-radiotherapy ranged between 11% and 44%.
Quality of surgery and radiotherapy	Complete tumour resection (histological verification of tumour free margins after surgery)	14% of head and neck cancer patients had a complete tumour resection. However the information was missing for a high proportion of the sample.
	Readmission, reoperation within 30 days from main surgery	Unknown - the indicator was proposed by experts after the data collection.
	grade 3 or more late toxicities (more than 3 months after radiotherapy)	Unknown - the indicator was proposed by experts after the data collection.
	Percentage of patients receiving intensity-modulated radiation therapy vs receiving 3D conformal radiation therapy	Unknown - the indicator was proposed by experts after the data collection.
	Availability of all types of surgery and reconstructive surgery	Unknown - the indicator was proposed by experts after the data collection.
Quality of pathology report after surgery	Percentage of pathology reports with a full set of core data items recorded according to the ESMO guidelines (site and laterality of the carcinoma, maximum diameter of tumour, maximum depth of invasion, histological type of carcinoma, degree of differentiation (grade), pattern of invasion, lymph node involvement)	This indicator was provided on a sample of 50 cases only. Information about site, laterality, histological type, maximum diameter of tumour, margin status were always available. However, maximum depth of invasion and pattern invasion were not included on the reports, and only 88% included the degree of differentiation (grade).
Availability of formalised multidisciplinary decision making and care	No indicators	No indicators collected by the high resolution study because the information was not retrievable in the medical records
Participation in clinical and translational research	No indicators	No indicators collected by the high resolution study because the information was not retrievable in the medical records

Appendix 5: Treatment centres identified for rare cancers in Ireland.

For some of the rare cancers below, designated centres have been established. However, discussions are still taking place with a view to reaching a consensus on where services should be located.

Cancer type	**Treatment centre**
Head and neck	Eight designated centres including: • Beaumont Hospital, Dublin • Cork University Hospital • St James' Hospital, Dublin
CNS	Operating as one national centre: • Beaumont Hospital Dublin • Cork University Hospital
Neuroendocrine tumours	• St Vincent's University Hospital, Dublin
Endocrine tumours	Discussions underway
Sarcomas (including GIST)	Discussions underway, but most likely multiple centres
Haematological tumours	Eight designated centres operating in a hub and spoke model including: • Sligo General Hospital • Letterkenny General Hospital • Tullamore Hospital • Mercy University Hospital, Cork Stem cell transplants only occur in: • St James' Hospital Dublin • University College Hospital Galway
Female genital organs	• Mater Hospital Dublin • St James' Hospital Dublin
Male genital organs	Not yet addressed
Embryonal tumours	Not yet addressed
Childhood cancers	• Mater Hospital, Dublin • St James' Hospital, Dublin
Thymus cancers	Not yet addressed
Digestive cancers	• St Vincent's University Hospital, Dublin • Cork University Hospital (operating as a satellite centre)
Malignant mesothelioma	• St. James's Hospital • Mater Hospital, Dublin • Cork University Hospital • University College Hospital Galway

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2. Singh S, Law C. Multidisciplinary reference centers: the care of neuroendocrine tumors. Journal of oncology practice / American Society of Clinical Oncology 2010;**6**(6):e11-6.

Other key reading:

ESMO / European Sarcoma Network Working Group. (2012). "Soft tissue and visceral sarcomas: ESMO Clinical Practice Guidelines for diagnosis, treatment and follow-up." Ann Oncol 23 Suppl 7: vii92-99.

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For further information, please see:

- RARECARENet - http://www.rarecarenet.eu/rarecarenet/
- European Cancer Patients Coalition (ECPC) - http://www.ecpc.org/
- Joint action on Cancer Control (CanCon) - http://www.cancercontrol.eu/index.php