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Centers of excellence or excellence networks: The surgical challenge and quality issues in rare cancers

S. Sandrucci [a, *], P. Naredi [b], S. Bonvalot [c]

[a] Visceral Sarcoma Unit, City of Health and Science, University of Turin, Italy
[b] Department of Surgery, Institute of Clinical Sciences, Sahlgrenska Academy, University of Gothenburg, Gothenburg, Sweden
[c] Department of Surgery, Institut Curie, PSL Research University, Paris, France

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ABSTRACT

There are several suggestions that centralization of care improves outcome for rare cancers, particularly when optimal treatment requires complex surgery or high-technology radiotherapy equipment. Diagnosis and treatment in reference centers are expected to be more accurate because they benefit from large numbers of cases discussed in a multidisciplinary tumor board with a well-run pathway. However, centralization is sometimes moderately perceived by oncologists as a solution to be endorsed for rare cancer patients; disadvantages of centralization are the need for patients to move and the risk of a longer waiting list, with discomfort and possible negative effects on outcome. It is difficult to find single experts on rare cancers: all the more it will be difficult to find a multidisciplinary panel of experts, and the role of the surgeon is to be a functional part of it. On the other side, from a surgical point of view, the quality of the initial management of many rare cancers directly impacts the final outcome; surgery of rare cancers may not necessarily be more demanding than the average from a technical point of view, but the lack of cultural knowledge about the disease can well lead to inappropriateness even in the lack of major technical challenges. Care for rare cancer patients must be organized in pathways that cover the patient's journey from their point of view rather than that of the healthcare system, and pathways must follow the best evidence on diagnosis, treatment and follow-up.

Rare cancers have an incidence of less than 6/100,000 per year, corresponding roughly to 640,000 new cases per year in Europe [1]. Despite the rarity of each rare cancer, they constitute all together 24% of all neoplasms diagnosed in the European Union each year. The European project RARECARENet [2], has identified, through a consensus process among several experts, about 198 different rare cancers. According to RARECAREnet, survival differences at 5 years between rare and common cancers are marked (up to 49% vs. 63%), and the disadvantage persists even excluding cancers with good prognosis (skin, prostate or breast). This fact may be due to the limited number of reference centers, both in individual countries and in Europe, and "GP Referral of patients" which implies GP and initial practitioner adequate training [3]. The consequences are delayed treatments, diagnostic uncertainty due to the lack of appropriate tools for the diagnosis and limited access to appropriate therapies and/or clinical expertise. There is a need of driving up quality in cancer organizations and services, given that there is agreement that high quality of care is not comprehensively accessible, and not well coordinated as the current evidence would justify.

An assessment of the quality of cancer care in Europe, according to the commitment of the European Commission, was first made by the European Partnership for Action Against Cancer (EPAAC) [4], which reported in 2014 important variations in service delivery between and within countries, observing that national cancer plans may be responsible for a quarter of the survival differences. EPAAC further focused on the establishment of networks of expertise in regions where it is not possible to establish comprehensive centers [4]. The EU Joint Action on Cancer Control (CANCON) which replaced EPAAC from 2014, published in 2017 the European Guide on Quality Improvement in Comprehensive Cancer Control [5], and Rare Cancers Europe, a multi-stakeholder initiative, is working to put identification and networking of reference centers for rare cancers on the European policy agenda [6].

* Corresponding author.
E-mail address: sergio.sandrucci@unito.it (S. Sandrucci).

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Currently, the main opportunity to improve quality of care has to do with the EU Reference Networks (ERN), an European Community call for the definition of networks for cancer care also in the field of rare cancers, by promoting international collaboration between centers of reference from different countries. The ERN is foreseen by the EU Cross-border Health Care Directive, i.e., the EU instrument by which the right of EU patients to be treated across the EU countries is sanctioned.

The recently established Joint Action on Rare Cancers (JARC) is a framework for involved stakeholders (18 EU state members, 8 ministry of health, 27 universities, public health institutions, cancer registries, oncological institutes, patients advocacies, scientific societies) aimed to prioritize rare cancer in national cancer plans and develop solutions to be implemented through the future ERNs on rare cancers, in the areas of Epidemiology, Quality of Care, Clinical practice guidelines, Innovation and access to innovation, Medical education, Childhood Cancers and Rare Cancer Policy [7].

Solid rare cancers: keyfacts and challenges

Diagnostics and primary treatment of rare cancers require multidisciplinary approaches [2,8] and very specific expertise. In addition, clinical and translational research calls for a high level of centralization and international collaboration. To what extent appropriate policies for rare cancer patients are implemented at the country level has seldom been studied, so information for policy makers and stakeholders is generally scarce.

A recent paper [2] provides up-to-date incidence and survival estimates based on data collected from 94 population-based cancer registries (CRs), for 198 rare cancers diagnosed in 2000–2007 and for 12 major families of rare cancers. It also presents data on the levels of centralization for rare cancers in selected European countries. The degrees of centralization vary across Europe, and to a large extent is affected by the population size. In countries with a small population it is easier to concentrate patients in a single or few hospitals. High admission volumes are more likely to be achieved in reference centers in larger-population countries. In this paper, the extent of centralization of rare cancer treatment is calculated by the *mean admission volume* (MAV) indicator, considered overall and by country for 38 cancer groups ranked by decreasing incidence. Pooled MAV ranged from a maximum of 83 treatments per year for head and neck tumors to fewer than 0.5 per year for choriocarcinoma of the placenta, some embryonal and endocrine tumors. The higher the incidence, the larger the MAV of treating hospitals. The relationship between cancer incidence and MAV in the pool of countries was very strong, though with several outliers. This was the case for epithelial tumors of the ovary, which had a higher incidence but a lower MAV than CNS tumors, whose patients seemed therefore to be more centralized than ovarian cancer patients. Similarly, soft tissue sarcomas had a ten times higher incidence, but received less centralized treatment than bone sarcomas. In contrast, tumors of the urinary tract, gastro-entero-pancreatic neuroendocrine tumors (GEP-NET), small intestine, non-epithelial ovary cancers, and NET of skin were treated in centers with an even lower MAV than would be expected because of their very low occurrence.

There are several suggestions that centralization of care improves outcome for rare cancers. This is particularly true when optimal treatment requires complex surgery or high-technology radiotherapy equipment. However, It is essential that treatment is organized in reference networks with hubs with a significant annual cases load. These rare cancers should be their main interest or one of their primaries; and spoke units or centers organized as an extended multidisciplinary team (MDT) at an appropriate geographical level (regional, national and supranational) [9].

Diagnosis and treatment in reference centers are expected to be more accurate because they benefit from large numbers of cases, which are discussed in a multidisciplinary tumor board involving expert professionals, with a well-run pathway [10]. Often centralized sites are connected to research centers participating in international debates and research. Disadvantages of centralization are the need for patients to move and the risk of a longer waiting list, with consequent discomfort and possible negative effects on outcome. Sometimes, centralization was only moderately perceived by oncologists as a solution to be endorsed for rare cancer patients.

It is essential that these patients should be addressed at the beginning of their illness at a stage where they are often curable by an adapted management.

Rare cancers and surgery

The RARECARENet project data [11] has shown that 65% of all rare solid tumors receive a surgical treatment, compared to 35 and 28% referred to radiation therapy and chemotherapy. Considering the treatment of the rare solid tumors by stage, about 4/5 of patients with a localized disease benefit of surgery [12].

The main issue concerning surgery of rare cancers is that a “rare cancers surgeon” can seldom exist. Rare cancers are almost ubiquitous, and involve every district of the human body: therefore, there may be some rare cancer surgeons for some of them while many cancer surgeons will deal with some of them, more or less occasionally, depending on the anatomical district they specialize in.

The surgeon role in rare cancers is best played if incorporated within a functioning multidisciplinary team. It is difficult to find single experts on rare cancers: all the more it will be difficult to find a multidisciplinary panel of experts. On the other side, from a surgical point of view, the quality of the initial management of many rare cancers directly impacts the final outcome. In other words, it is the multidisciplinary clinical decision on how to plan treatment at the beginning of patient clinical history and the quality of initial surgery, which will largely determine the patient's outcome [8]. It is worth recalling that surgery of rare cancers may not necessarily be more demanding than the average from a technical point of view, but the lack of cultural knowledge about the disease can well lead to inappropriateness even in the lack of major technical challenges.

There is a wide consensus among rare cancer experts that patients with rare cancers should be treated at centers of reference from the very beginning of their clinical history, possibly from the time of biopsy. It is well established in rare cancers pathology that “expert” opinion is not always absolutely convergent, though often different conceptualizations of a tumor do not necessarily generate different treatment implications. A centralized pathological review, rapid and efficient help with access to molecular biology analysis seem of vital importance in these rare tumors [10]. More efficient information and education of the pathologists also seems essential to ensure accurate diagnosis and grading [13]. Probably, the main criterion for a reference institution, all the more for a surgical facility, is a “volume” high enough so as to guarantee medical expertise, technical appropriateness, and access to clinical trials [8].

Finding centers of reference may be a frustrating experience for rare cancer patients and it may be difficult also for general practitioners. For this reason, one of the object of RARECAREnet project was to make available a list of hospitals where rare cancers are treated by European countries and by rare cancer [14]. However, such a list is difficult to achieve at national level, actually when MSs applied for the ERN, the identification of center of reference, was not an easy task. The alternatives is to individuate national and

supranational networks, i.e., “reference networks”, “networks of excellence”, and such like. As a matter of fact, how rare cancer patients are referred and managed differ substantially across European countries, depending on several factors, which have largely to do with the peculiarities of national or even regional health systems, which include a cancer plan with a task on rare cancers, the size of countries, and probably the GDP and the TNEH [15].

Quality of care

Essential requirements for the organization of solid rare cancers care are: cancer care pathways that cover the entire patient journey, timeliness of care, minimum case volumes, multidisciplinary team working in dedicated centers or units, peer reviewing of diagnostics and pathology reports, education, policies to enrol patients in clinical trials, patient information, audit and quality assurance of outcomes and care processes [10].

Care for rare cancer patients must be organized in pathways that cover the patient's journey from their point of view rather than that of the healthcare system, and pathways must follow the best evidence on diagnosis, treatment and follow-up.

It must be clear to the patient which professional is responsible for each step in the treatment pathways and who is following the patient during the journey (usually called a case manager or patient navigator) [16]. In many countries, case managers during the main stages of treatment are cancer nurses.

Patients must be involved in every step of the decision-making process. Their satisfaction with their care must be assessed throughout the patient care pathway. Details regarding a rare cancer are scarcely available, especially for the public, thus patients must be offered relevant and understandable informations to help them appreciate the process that will be followed with their treatment, from the diagnosis standpoint. They must be supported and encouraged to engage with their health team to ask questions and obtain feed-back on their treatment wherever possible and on new treatment or clinical trial available. Children need to be involved in an age-appropriate manner and their parents/carers should be included in the process as appropriate.

It is also essential that rare cancer patient support organizations are involved whenever relevant throughout the patient pathway. These groups must work to improve patients' knowledge and ability to take decisions, secure access to innovative therapies and improve quality of treatment.

To properly assess quality of rare cancers care, three categories of outcomes must be measured and collected in a database: clinical outcomes, process outcomes and patient-reported outcomes (PROs) [10]. These databases have to produce useful information for all the stakeholders and that are linkable between them and with other international database, since the rarity of the disease make this imperative.

To ensure appropriate, timely and high-quality care, a quality management system (QMS) must be in place. It must involve clinical care, strategic planning, human resource management, training etc. The QMS must be accountable at an institutional management level and be based on written and agreed documentation such as guidelines, protocols, patient pathways, structured referral systems and standard operating procedures (SOPs); unfortunately, guidelines are rarely available for rare cancers because experience is low. The QMS must ensure the continuity of care for patients, the involvement of patients in cancer care pathways, and the reporting of patient outcomes and experience.

As part of a QMS, an effective data management and reporting system, and an internal audit system, are necessities. Transparency on activity of the expert centers should be developed: for patients and health care providers, on-line access of the cases load and treatment received should be available.

Conflict of interest

No Conflict of interest concerning this paper.

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