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Networking in rare cancers: What was done, what's next

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ABSTRACT

Rare cancers represent approximately one fourth of all cancers. Despite being a heterogeneous group of diseases, they share similar problems including lack of expertise, issues in quality of care, discrepancies in outcome and limitations in research. Traditionally, centralization of rare cancer patients to dedicated reference centres has been recommended to ensure expertise, multidisciplinarity and access to innovation. However, centralization entails health migration, rationing of resources and a potential failure in routine care. By ensuring appropriate care to all patients regardless the point of access, networking seems the most appropriate answer to the problem of rare cancers. The launch of the Joint Action on Rare Cancers as well as the recent establishment of the European Reference Networks represent for the first time a concrete opportunity to make networking a reality and ultimately reduce disparities and improve outcome in these diseases.

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Introduction

Despite the rarity of each of them, with 198 nosographic entities identified, collectively rare cancers represent 24% of all new cancer cases diagnosed in the EU27 every year [1]. On average, their outcome is slightly worse compared to common neoplasms. In the face of that, discrepancies in survival are visible across European regions, a high rate of health migration does exist, issues in quality of care are reported, research is problematic [2]. The RARECAREnet study on quality of care in two distinct families of rare cancers (extremity soft tissue sarcomas and head and neck cancers) showed that diagnosis and treatment might lie below optimal standards in some EU countries. This was more likely when care is delivered by institutions with limited expertise in the field and/or low case volumes, and unfortunately this is often the case with both cancers (*Trama A., unpublished results*).

Traditionally, there has been a general consensus to recommend centralize rare cancer patients to dedicated reference centres with multidisciplinary expertise. Pathologic diagnosis is one of the most crucial items, among others. There is a price to pay to centralization, however, in terms of health migration, implicit rationing of resources, and so forth. Health migration has a dramatic impact on patient's quality of life and finances, giving rise to huge social costs for patients and society. On the other side, improper referral also causes direct health costs, due to the large number of inappropriate interventions, which, at best, will be subsequently corrected in reference centres, thus worsening quality of life, if not survival, and costs. Collaborative health networks are the most obvious alternate option to centralizing all patients for their entire treatment. Networking would be crucial to research, as well, allowing collaborative studies, virtual tissue banking, and the like. However, networking is challenging. We are in acute need to address its challenges.

Networking and rare cancers

One of the main aims of a rare cancer networks is to ensure that appropriate care is guaranteed to all patients regardless of where they live, thus limiting health migration. The most logical model for networks on rare cancers is the “hub-and-spoke” model.

The “hub-and-spoke” model makes available to collaborating centres (“spokes”) the expertise of reference centres (“hubs”). While a “hub” provides a full range of services, a “spoke” works in close connection with a hub, offers a more limited service arrays

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but ensures geographical coverage. The "hub-and-spoke" model maximises efficiency and effectiveness in those settings marked by a medium-low number of cases and a need for highly specialized expertise and access to innovation.

"Hubs" in rare cancers

Rare cancers, as all cancers, need timely and appropriate diagnosis and optimal treatment strategies. However, because of the low numbers, professional expertise and multidisciplinarity are rarely found in the community. To overcome this limitation, hubs should concentrate the best expertise available and ensure an accurate clinical, pathologic and biological assessment of the disease, as well as an expert clinical decision provided by a dedicated multidisciplinary team. In addition, in rare cancers hubs have the task of promoting research on new treatments, setting up clinical registries and biobanks and referring patients to open clinical studies.

"Spokes" in rare cancers

Spokes are meant to be centres with good expertise in managing cancer patients, providing a limited range of high-quality services, being located as close as possible to patient's home. Medical treatments as well as surveillance are interventions that can be foreseen at the level of a spoke. The number of spokes required at a national level should be tailored according to the entire population and the need of ensuring full geographical coverage. This is essential to reach one of the main goals of an effective network in rare cancers, which is to improve patient's quality of life by limiting efforts and costs due to migration and avoiding possible diagnostic delays.

"Hub and spokes": how should they work together?

A continuous dialogue and a close link between the hub and the spokes are vital to make the network effective. A multidisciplinary assessment of the case should be part of every rare cancer patient management from the very beginning. This process should not be put in place only for defining the initial treatment strategy, but should be deployed whenever the status of the disease changes (i.e. diagnosis, recurrence, progression). Still, in a limited number of situations, the patient will need to travel to the hub to access services that cannot be provided by the spoke, including surgery in some circumstances, high-tech treatments and access to clinical studies.

The challenges of networking

Ensuring quality in a "hub-and-spoke" network

There are critical points in the establishment of a "hub and spoke" networks, such as ensuring the quality of both hubs and spokes, ensuring the quality of patient's pathway and generating data on the effectiveness and cost/effectiveness of the effort.

The selection of hub centres for rare cancers must be based on their expertise. The identification of criteria to endorse an institution as a hub can vary according to the disease, although there are principles that should be consistent across all rare cancers families. A hallmark of a hub centre is a well-established multi-disciplinary team, having at least weekly meetings for clinical case discussion. For each rare cancer, the composition of a "core multi-disciplinary team" should be discussed, i.e. those key professional figures that need to regularly attend the meeting and join case discussion. An "extended multi-disciplinary team" can also be foreseen, which will include specialists not needed on a routine basis but only in selected circumstances. Besides having a strong multidisciplinary expertise, hubs need to manage a critical number of patients. The minimum number of cases to manage can vary widely across diseases, with some thresholds established on a national basis or by scientific societies or disease-based communities. As an example, the guidelines for cancer services on sarcoma from the National Institute for Health and Clinical Excellence (NICE) in the UK state that a sarcoma multidisciplinary team should manage at least 100 new soft tissue sarcoma patients and 50 new bone sarcoma patents per year [3]. Similarly, the general threshold identified from the European Neuroendocrine Tumour Society (ENETS) for the accreditation of an institution as a centre of excellence is 80 new neuroendocrine tumour patients to be managed per year [4]. Finally, active engagement of the hub in research activities, as documented by its scientific output, and open clinical trials are criteria, as educational commitment.

Endorsement of spokes may be less demanding. Besides providing good-quality cancer services, the key point for spokes is the commitment of the institution to working within the network. Similarly, it is important that physicians from the spokes regularly attend educational programmes, to build up and maintain the expertise needed to interact with the hub's experts on single clinical cases.

Assuring quality of hubs and spokes in the network is not enough, since there is no guarantee in principle that then they will properly share any single patient. On the contrary, any single patient belonging to a network should be routed via a care pathway complying with quality standards. In other words, it is the single rare cancer patient care pathway that should be quality-assured. For example, one needs to ensure that the single patient's pathologic diagnosis has been centrally reviewed, as well as the case shared over the network with a hub at all crucial clinical steps (e.g., when treatments are changed, etc.). Of course, the patient needs to be informed that he/she has been routed along such a care pathway over the network. Then, multidisciplinary interactions will often end up into deviations from standard care pathways, but they should be properly justified.

Is networking cost/effective?

Funding is one of the main constraints of networks. Although often neglected, networking entails costs. Today, the IT tools and the network infrastructures are probably not the problem. Much more, it is the administrative burden, the logistics and, above all, the extra medical workload that need to be reimbursed. Otherwise, networking will never fly, all the more at a time of economic crisis. Clearly, effectiveness should improve, although this is not always easy to prove objectively. Social costs are diminished in principles by effective networks, but they are not covered by health systems. However, often it may be easy to demonstrate that some direct health costs are decreased through a decrease of costs from inappropriate treatments. For example, if surgery has to be done twice, of course costs will be rising, even leaving behind all shortcomings in terms of patient's prognosis and quality of life. Thus, networks should make an effort to objectify such costs, thus proving obvious economies achieved through networking as such. On the long run, networks should annotate their outcomes, in order to provide reasonable demonstrations of the high quality of care they are able to guarantee. Likewise, under a society perspective, one should factor economies made through the decrease of social costs due to health migration, and the like. However, providing evidence in support of the hypothesis that networking is cost/effective still remains a challenge, and prospective collection of all relevant data should definitely be encouraged.

Rare cancers: solutions in Europe

In conclusion, proper referral of patients and effective clinical networking are crucial in rare cancers. In the EU, a big opportunity is provided at the moment by the European Reference Networks (ERNs). They are foreseen by the Cross-border Health-care" Directive of the EU, setting the conditions under which EU citizens have the right to travel to another EU country to receive medical assistance and to be reimbursed for care abroad by their home country [5]. These conditions are not ideal, indeed. However, this Directive provides the legal basis for the institution of the European Reference Networks (ERNs). The ERNs are virtual networks of selected institutions targeting rare conditions that require highly specialized treatments and a concentration of knowledge and resources. The 24 ERNs currently active have been announced in March 2017 and cover a wide range of rare conditions, including rare cancers. Three ERNs are dedicated to rare cancers: EuroBloodNet (for rare haematological diseases; https://www.eurobloodnet.eu), PaedCan (for paediatric cancers; http://paedcan.ern-net.eu) and EURACAN (for adult solid rare cancers; http://euracan.ern-net.eu). The core business of ERNs is tele-consultation, which means providing multidisciplinary second opinions for the management of complex cases. A common platform allowing data sharing among ERNs healthcare providers is currently under development. Besides teleconsultations, ERNs are also meant to generate and disseminate knowledge on rare cancers, promote medical education and patient's information, produce clinical practise guidelines and foster research as well as epidemiological surveillance. Among problems faced during ERN development one finds limited fundings and the lack of any legal status. In order to guide, support and harmonize the work of the three ERNs focused on rare cancers, the European Commission has launched in 2016 a Joint Action on Rare Cancers (JARC; www.jointactionrarecancers.eu), acknowledging the peculiar challenges of rare cancers as compared to rare non-neoplastic diseases. The JARC is a multi-stakeholder collaboration between 18 EU Countries and the European Commission, coordinated by Fondazione IRCCS Istituto Nazionale dei Tumori of Milan, which will be pursued in a 3-year span (2016–2019). The effort of the JARC has been totally shaped around ERN development. There are today 34 partners involved in the JARC. The contribution of patient associations to this project is felt to be essential and their presence is meant as cross-cutting all JARC activities.

The final aim of ERNs and JARC is to bring innovation, knowledge and expertise from centres of excellence to the patient, independent of the point of access. We strongly believe that this goal can only be achieved by progressively transforming ERNs into "network of networks", i.e., networks of hub-and-spoke networks. This will need close collaboration and motivation of all EU member states. National networks on rare cancers need to be established and develop. This is the challenge. However, it is the first time this may really happen.

Competing interest statement

The authors have no competing interests to declare.

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