Introduction

The Belgian Cancer Registry (BCR), founded in 2005, progressively achieves more results on population based cancer statistics. Data on cancer incidence are available for Belgium from 2004 on and for the Flemish Region from 1999 onwards. Since 2009, the BCR is authorised to use the national number for social security (NNSS) as the unique patient identifier, to link the data from this BCR database with data on cancer-related diagnostic and therapeutic procedures and pharmaceuticals [1], which are obtained from all seven Belgian health insurance companies via the Intermutualistic Agency. The NNSS can also be used to retrieve the vital status from the "Kruispuntbank van de Sociale Zekerheid/Banque Carrefour de la Sécurité Sociale". Previous publications on cancer incidence (2004-2005 [2], 2008 [3] and 2010 [4]) and survival [5] have mainly provided general results by cancer type (ICD-10 [6]) at both the national and regional level. The current project is the first to provide a detailed inventory of rare cancer incidence, survival and clinical care in the Flemish Region.

RARECARE has been set up as a European initiative to estimate the epidemiology, treatment and survival of rare cancers. Within the RARECARE working group, rare cancers have been defined as groups of rare malignancies with an incidence rate lower than 6/100,000 per year, for both sexes combined [7,8]. Numerous international publications within and outside the RARECARE initiative have shown that clinical management of rare cancers is often poorly organized, resulting in suboptimal treatment outcomes [7-9]. In concrete, a lack of medical expertise in the management of rare cancers, poor referral rates from general practitioners and pathological misdiagnoses may lead to an impaired quality of care for these cancer types. On the other hand, rare cancers are often not prioritized by the public health system due to their relatively low burden at the population level. The same is true for pharmaceutical industry which considers rare cancers to be less profitable than their common counterparts. Therefore, research on diagnosis and treatment of these tumour entities gains less attention.
Several initiatives possibly leading to an improvement of this situation have been formulated such as the creation of evidence based guidelines and the establishment of reference networks or centres of expertise.

To date, little is known on the burden and clinical management of these tumour types in the Flemish Region. The current project aimed therefore to estimate the incidence and survival of an extended list of rare cancers in the Flemish Region, and to provide more detailed insight in the clinical care for a selection of rare malignancies in this region. The selection of rare cancers in the current project was based on the RARECARE cancer list [10]. This list is based on a combination of topography and morphology codes according to ICD-0-3 [11] for incidences between 2002 and 2010. For incidence year 2001, selection was based on ICD-O-2 [12], which might cause a small, negligible bias.

The first part of this issue describes incidences, trends and survival (observed and relative) for an extended list of rare cancers across different organ systems. Analyses were performed at the overall level and for subgroups according to sex, age and stage. For this part, all Belgian patients with residence in the Flemish Region and a first diagnosis of a rare cancer between 2001 and 2010 were included.

The second part presents detailed analyses of clinical care for a selection of 11 rare cancers, namely: Nasopharynx, Salivary Glands, Hypopharynx, Larynx, Oropharynx, Oral Cavity, Lip, Anal Canal, Vulva, Vagina and Mesothelioma. For this part, all Belgian patients living in the Flemish Region, with an incidence of a rare cancer between 2004 and 2007 were considered. Each tumour type within this part is introduced by a review of its epidemiology, aetiology, diagnostic and therapeutic management. Detailed analyses of different patient (age, sex) and tumour (stage, histology) characteristics are provided. In addition, clinical care for these tumour entities in terms of multidisciplinary oncological discussion, diagnosis and treatment is described. Survival analyses are presented for the overall level and for different subgroups, in line with the major differences in patient or tumour characteristics or treatment schemes. Finally, data on how clinical management for these patients is spread amongst different hospitals and the major differences in treatment schemes between low- and high-volume centres are shown.

Hopefully, this project can increase knowledge on rare cancer burden and management in the Flemish Region. These insights could be of use in future reflections on optimization of clinical rare for patients diagnosed with rare malignancies.

References

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