INTRODUCTION
INTRODUZIONE

This new monograph of the Italian Association of Cancer Registries (AIRTUM) deals with "rare" cancers, defined as those diagnosed in less than 6 every 100,000 citizens per year in the European population.
Rare cancers are a red-hot issue for all oncology stakeholders: patients, clinicians, the pharmaceutical industry, and policy-makers. The reason is that their rareness makes diagnosis more difficult and causes a lack of specific drugs and widespread expertise in treatment. Moreover, the continuous developments in cancer research have provided new insights into cancer biology, resulting in better identification of specific oncological entities. For example, we are now able – using specific biomarkers – to identify two or several tumours, previously considered as a single type, which differ from each other in biochemical footprint, treatment, and prognosis, as well.
This is what prompted AIRTUM to address this issue, in collaboration with the Italian National Cancer Institute of Milan, the "Istituto Superiore di Sanità", and the involvement of the Italian Association of Medical Oncology (AIOM), the Italian Federation of Volunteer-Based Cancer Organisations (FAVO), the Italian Society of Haematology (SIE), and the Italian Society of Surgical oncology (SICO).
AIRTUM's huge database proved effective in providing high-quality, reliable information even on rare cancers, with more than 330,000 cases used to compute incidence (years: 2000s) and 280,000 used to estimate survival. The study shows that about 89,000 (or 25%) of new annual cancer diagnoses in Italy are of rare cancers. The huge overall number does not solve the peculiarities of rarity: among the 198 analysed rare cancer types, more than two thirds have an incidence below 0.5 cases per 100,000 per year.
Some tumour types that are rare according to the European definition (diffuse large B-cell lymphoma, squamous cell carcinoma of larynx, multiple myeloma, hepatocellular carcinoma, and thyroid carcinoma) have an incidence in Italy that exceeds the European cut-off, introducing the need for harmonization in definition and classification to obtain reliable international comparisons.
The monograph provides an impressive amount of information on detailed types of rare tumours. To cite a few data, 5-year relative survival appears to be lower on average for rare tumours than for frequent types. The number of Italian citizens living with a rare tumour diagnosis has been estimated to be 900,000. Prevalence differs according to the different combination of incidence and survival in each of the rare tumours.
This monograph provides an invaluable amount of previously unpublished information on the epidemiology of rare tumours in Italy, both as an overall group and for each of the 198 types, which have been analysed in detail for the first time.
As is the case for all other AIRTUM reports, the data in this monograph are available to all interested stakeholders, and they focus on national peculiarities, quantifying individual burdens and outcomes, and supporting health care planners in the laying out of case-specific health care services.
Rare cancers are not only relevant today, but their relevance is expected to grow in the future, pending further discoveries and major progress in ongoing cancer research.
The quality and reliability of AIRTUM data, recently confirmed even by the Ministry of Health, support the dissemination of reliable information regarding the epidemiology of cancer in Italy, now including rare tumours.

Steering Committee
Italian Association of Cancer Registries (AIRTUM)