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Rare cancers of the head and neck area in Europe

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ABSTRACT

The RARECARE project has proposed a different and more detailed grouping of cancers, based on localisation and histological type, in order to identify rare entities with clinical meaning. RARECARE gathered data on cancer patients diagnosed from 1978 to 2002 and archived in 76 population-based cancer registries, all of which had vital status information available up to at least 31st December 2003. This study provides incidence, prevalence and survival rates for rare head and neck epithelial (H&N) cancers.

Among the rare H&N cancers, those of oral cavity had the highest annual crude incidence rate of 48 per million, followed by oropharynx and 'major salivary glands and salivary gland type tumours' (28 and 13 per million, respectively). Incidence rates of epithelial tumours of nasal cavities, nasopharynx, eye and adnexa and middle ears were all lower than 5 per million. The prevalence for all investigated entities was lower than 35 per 100,000. The 5-year relative survival rates ranged from 40% for epithelial cancer of oropharynx to 85% for epithelial cancer of eye and adnexa. Survival rates were lower for men and for patients aged ≥ 65 years. With few exceptions, the lowest and highest survival figures were observed for Eastern Europe and Northern Europe, respectively.

According to the definition for rare tumours by RARECARE (incidence < 6 per 100,000), as well as according to the definition for rare diseases by the European Commission (prevalence < 50 per 100,000) the H&N cancers described in this paper should be considered rare and diagnosis and treatment of these cancers should therefore be centralised.