Incidence, prevalence and survival of patients with rare epithelial digestive cancers diagnosed in Europe in 1995–2002

J. Faivre a,*, A. Trama b, R. De Angelis c, M. Elferink d, S. Siesling d, R. Audisio e, J.F. Bosset f, A. Cervantes g, C. Lepage a, The RARECARE Working Group

a Burgundy Cancer Registry, INSERM U866, Burgundy University, Dijon University Hospital, BP 87900 21079 Dijon, France
b Fondazione IRCCS Istituto Nazionale dei Tumori, Via Venezian 3, 20133 Milano, Italy
c Centro Nazionale di Epidemiologia, Istituto Superiore Sanita, Viale Regina Elena 299, Rome, Italy
d Comprehensive Cancer Centre, The Netherlands, Hoedemakerplein 2, Enschede, The Netherlands
e University of Liverpool, St. Helens Hospital, Marshalls Cross Roads, St. Helens WA9 5DA, UK
f Service de Radiothérapie CHU Jean Minjoz 2, boulevard Fleming, F-25030 BESANCON, Cedex, France
g Hospital Clínico Universitario, Valencia, Spain

ARTICLE INFO

Article history:
Available online 9 December 2011

Keywords:
Rare diseases
Digestive cancer
Small intestine
Anal canal
Gallbladder and extra-hepatic bile duct cancer
Cancer registries
Incidence
Prevalence
Survival

ABSTRACT

Background and aims: Little is known about the epidemiology of rare epithelial digestive cancers. The aim of this study was to report on their incidence, prevalence and survival across Europe.

Methods: The analysis was carried out on 50,646 cases diagnosed from 1995 to 2002 within a population of 162,000,000 in 21 European countries. Age-standardised incidence rates were computed using the European standard population. Prevalence rates, relative survival and period survival indicators for the years 2000–2002 were calculated. The expected number of new cases per year and of prevalent cases in Europe was estimated.

Results: There were large variations in gallbladder epithelial cancer incidence rates: the incidence in Eastern Europe was 7 times higher than in the UK & Ireland. Differences between incidence rates were smaller for the other sites. The estimated number of new epithelial cancers arising in the EU each year was estimated to be 11,050 for extrahepatic bile duct cancer, 10,713 for gallbladder cancer, 5427 for anal cancer and 3595 for small intestine cancer. The corresponding estimated number of total prevalent cases was 18,483, 15,620, 40,589 and 13,276. There was also a large variation in the 5-year relative survival rate. For epithelial cancer of the anal canal, this varied between 66% (Central Europe) and 44% (Eastern Europe). The corresponding rates for small intestine cancers were 33% and 20%, for extrahepatic bile duct cancers, 17% and 12% and for gallbladder cancer 13% and 10%.

Conclusion: There are large variations within Europe in the incidence and survival of rare digestive cancers according to geographic area.

© 2011 Published by Elsevier Ltd.

1. Introduction

Digestive cancers, which are mostly epithelial cancers, are common in Europe. They accounted for approximately 25% of all cancers in 2008. Among digestive cancers, colorectal cancer was particularly frequent, corresponding to nearly 15% of all cancers. However, some sites were rare (crude incidence rates less than 6/100,000), including small intestine,