AREA MAJOR GROUPS	MALIGNANT CASES DIAGNOSED BETWEEN 2000 AND 2010	NOS[$]	DCO	AUTOPSY ONLY CASES	CASES WITH ZERO SURVIVAL TIME	MICROSCOPICALLY CONFIRMED CASES	LOST TO FOLLOW-UP CASES
	No.	%	%	%	%	%	%
AIRTUM POOL							
Epithelial tumours of head and neck	43 163		0.3	0.2	0.2	97.1	0.8
Tumours of the eye	1 530		0.6	0.2	0.2	58.9	0.7
Digestive system tumours	358 109		1.4	0.6	0.2	78.7	0.6
Rare epithelial tumours of the digestive system	57 891		0.7	1.2	0.2	73.5	0.6
Thoracic cavity tumours	157 478		1.6	1.0	0.2	74.4	0.5
Rare epithelial tumours of the thoracic cavity	12 027		0.1	2.8	0.2	97.4	0.5
Female genital system tumours	246 903		0.6	0.1	0.2	95.5	1.0
Rare tumours of the female genital system	41 141		0.1	0.2	0.2	99.3	1.0
Urinary system tumours	104 116		0.6	0.4	0.2	89.0	0.9
Rare epithelial tumours of the urinary system	6 394		0.2	0.5	0.2	94.5	0.6
Male genital system tumours	152 102		0.8	0.3	0.2	92.3	0.8
Rare tumours of the male genital system	9 049		0.1	0.1	0.3	95.4	1.8
Tumours of the Central Nervous System	13 071		0.0	0.5	0.2	91.2	1.0
Haematological diseases	123 307		0.8	0.5	0.3	93.1	0.9
Rare haematological diseases	91 094		0.4	0.5	0.3	94.4	0.9
Skin tumours^	33 823		0.1	0.0	0.3	97.5	5.9
Rare skin tumours and malignant melanoma of mucosa	1 699		0.0	0.0	0.5	99.5	1.5
Embryonal tumours	859		0.1	0.0	0.5	93.2	0.9
Sarcomas	20 019		0.0	0.4	0.3	98.6	1.4
Neuroendocrine tumours	9 196		0.0	0.5	0.3	99.6	0.8
Tumours of the endocrine organs*	32 268		0.2	0.2	0.3	95.4	1.2
NORTH-WEST							
All malignant cancers^	445 918	14.7	1.1	0.1	0.3	85.7	1.0
Rare cancers	111 744	–	0.3	0.0	0.4	92.1	1.3
NORTH-EAST							
All malignant cancer^	470 760	13.4	0.8	1.1	0.1	87.3	0.2
Rare cancers	116 808	–	0.1	0.0	0.1	93.5	2.0
CENTRE							
All malignant cancer^	125 671	14.9	0.7	0.0	0.1	85.1	1.6
Rare cancers	31 005	–	0.3	0.2	0.2	90.0	0.8
SOUTH							
All malignant cancer^	282 706	19.2	2.1	0.0	0.3	82.5	0.9
Rare cancers	79 846	–	0.9	0.5	0.0	91.1	0.0
AIRTUM POOL							
All malignant cancer^	1 325 055	15.2	1.2	0.4	0.2	85.5	0.8
Rare cancers	339 403	–	0.9	0.0	0.0	92.1	0.6

[$] **NOS**: ICD-O-3 morphological code: 8000,8001,9800,9590,9820,9760,9860,9800,9801 / * including carcinomas of thyroid gland / ^ excluding non melanoma skin cancer

Table 3. Number of cases diagnosed in 2000-2010 and data quality indicators for the 14 major groups of rare cancers and corresponding common cancers (when the group included also common cancers), and for all malignant tumours vs. all rare cancers, by geographic area and in the overall AIRTUM Pool. Quality indicators include proportion of not otherwise specified (NOS) morphologies (8000,8001 for solid cancers, and 9590, 9591, 9760, 9800, 9801, 9820, 9860 for haematological diseases), death certificate only (DCO) cases, autopsy only cases, cases with zero survival time, microscopically confirmed cases, and lost to follow-up cases (follow-up time <5 years). Pool of 39 general CR of the AIRTUM database.

All standard indicators of data quality for Italian CRs are satisfactory according to international standards.[15]

The new and most relevant indicator to evaluate the accuracy of diagnosis for rare cancers, with respect to the other routinely applied indicators, is the proportion of cases with a NOS category (ICD-O-3 8000-8001 for solid cancers, and ICD-O-3 9590-9591, 9760, 9800-9801, 9820, 9860 for haematological diseases). For rare cancers, the most likely quality problem is lack of specificity of morphology codes, which make it impossible to assign such cases to a specific (rare) cancer entity, resulting in underestimation of the true incidence and prevalence of such entities. Unspecified morphology can be due to genuine difficulty in assigning a specific morphological category or because inadequate documentation was supplied to the CR when the case was registered. The latter problem is registration bias and results in incidence and prevalence underestimation. To assess the extent of registration bias at European level, RARECARE reviewed the original data (mainly pathologic reports) of a selected sample (about 18,000 cases) of eight rare cancers (for details see RARECARE web site).[5] Briefly, the great majority of NOS morphology cases were confirmed as NOS. The few NOS cases that changed to a more specific diagnosis generally increased the incidence of the more common cancer forms. For example, 11% of epithelial oral cavity cancers were reclassified from NOS to more specific diagnoses: 8% were reclassified as squamous