

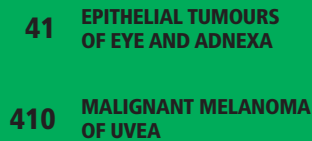
TUMOURS OF THE EYE

100%
OF EYE TUMOURS
ARE RARE

INCIDENCE

451

ESTIMATED NEW CASES
ITALY, 2015

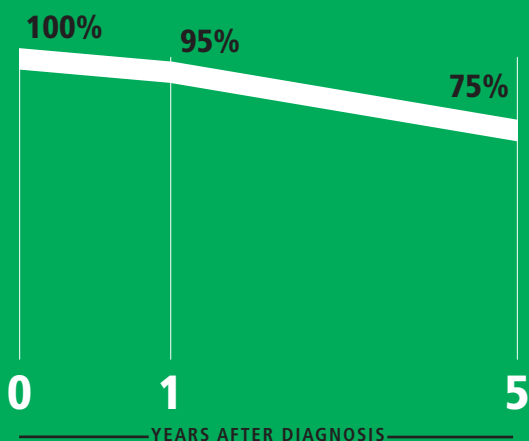


PREVALENCE

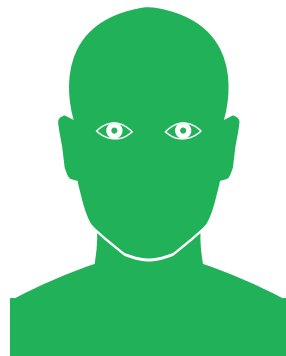
5 869

ESTIMATED PREVALENT CASES
ITALY, 2010

SURVIVAL



INCIDENCE

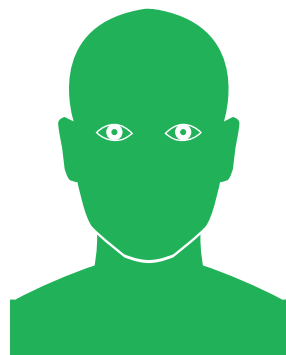


TUMOURS OF THE EYE. Crude incidence (rate per 100,000/year) and 95% confidence interval (95% CI), observed cases and proportion of rare cancers on all (common + rare) cancers by site. Rates with 95% CI by sex and age. Estimated new cases at 2015 in Italy.

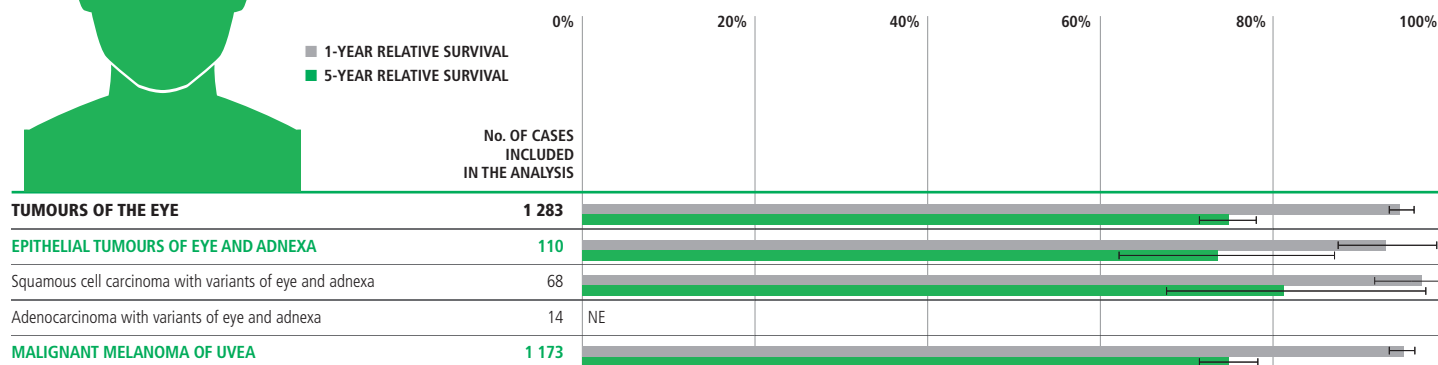
	AIRTUM POOL (period of diagnosis 2000-2010)													ITALY	
	RATE	95% CI	OBSERVED CASES (No.)	RARE CANCERS BY SITE (%)	SEX				AGE						ESTIMATED NEW CASES 2015
					MALE		FEMALE		0-54 yrs		55-64 yrs		65+ yrs		
					RATE	95% CI	RATE	95% CI	RATE	95% CI	RATE	95% CI	RATE	95% CI	
TUMOURS OF THE EYE	0.69	0.66-0.73	1 530	100%	0.70	0.65-0.75	0.68	0.64-0.73	0.26	0.23-0.28	1.21	1.08-1.35	1.85	1.72-1.98	451
EPITHELIAL TUMOURS OF EYE AND ADNEXA	0.06	0.05-0.07	133	NA	0.09	0.07-0.11	0.04	0.03-0.05	0.03	0.02-0.04	0.07	0.04-0.11	0.17	0.13-0.21	41
Squamous cell carcinoma with variants of eye and adnexa	0.03	0.03-0.04	77	NA	0.05	0.04-0.07	0.02	0.01-0.03	0.01	0.01-0.02	0.05	0.03-0.08	0.11	0.08-0.14	24
Adenocarcinoma with variants of eye and adnexa	<0.01	0.00-0.01	18	NA	0.01	0.01-0.02	<0.01	0.00-0.01	<0.01	0.00-0.01	<0.01	0.00-0.03	0.02	0.01-0.04	5
MALIGNANT MELANOMA OF UVEA	0.63	0.60-0.66	1 397	NA	0.61	0.56-0.66	0.65	0.60-0.70	0.23	0.21-0.26	1.14	1.02-1.28	1.68	1.56-1.80	410

NA: not applicable

SURVIVAL

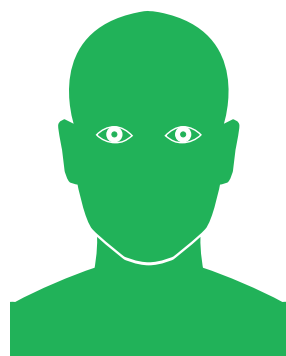


TUMOURS OF THE EYE. One and 5-year relative survival. Error bars are 95% confidence interval. Cohort approach (complete analysis), period of diagnosis 2000-2008.



NE: not estimable because 30 or less incident cases were observed

PREVALENCE



TUMOURS OF THE EYE. Observed prevalence (proportion per 100,00 and 95% confidence interval - 95% CI) by duration (≤ 2 , 2-5, ≤ 15 years) prior to prevalence date (1st January 2007), and complete prevalence. Estimated prevalent cases in 2010 in Italy.

	AIRTUM POOL								ITALY
	OBSERVED PREVALENCE BY DURATION						COMPLETE PREVALENCE		ESTIMATED PREVALENT CASES 2010
	≤ 2 YEARS		2-5 YEARS		≤ 15 YEARS		PROPORTION	95% CI	
	PROPORTION	95% CI	PROPORTION	95% CI	PROPORTION	95% CI			
TUMOURS OF THE EYE	1.57	1.32-1.86	1.89	1.62-2.21	6.89	6.35-7.47	10.11	9.28-10.93	5 869
EPITHELIAL TUMOURS OF EYE AND ADNEXA	0.15	0.08-0.26	0.09	0.04-0.18	0.52	0.38-0.69	0.78	0.55-1.01	454
Squamous cell carcinoma with variants of eye and adnexa	0.10	0.05-0.20	0.06	0.02-0.13	0.38	0.26-0.53	0.48	0.31-0.64	271
Adenocarcinoma with variants of eye and adnexa	0.01	0.00-0.06	0.01	0.00-0.06	0.05	0.01-0.12	0.06	0.00-0.13	39
MALIGNANT MELANOMA OF UVEA	1.42	1.18-1.70	1.80	1.53-2.11	6.37	5.86-6.93	9.33	8.54-10.12	5 415

This group of tumours includes the major cancers occurring in the eye:

- epithelial tumours and adnexa;
- uveal melanoma.

Both are extremely rare, with annual incidence rates (IR) <0.5 per 100,000year.¹ These exceptional cancers have already been described in previous papers based on two large datasets, the EURO-CARE study and the RARECARE project.²⁻⁴ These two large population-based studies used data from almost 100 cancer registries, thus providing solid measures of incidence, survival, and prevalence for these very rare cancers in Europe.

WHAT DO WE KNOW ABOUT THESE CANCERS?

Uveal melanoma is an adult intraocular tumour, arising from melanocytes in the uvea. **Adnexal skin tumours** are extremely diverse group of neoplasms, arising from cutaneous appendages, particularly the sebaceous, apocrine, and eccrine glands. Because of their rarity, even the basic descriptive epidemiology of these tumour types is sparse.⁴

Uveal melanoma is the most common ocular tumour. A very large study based on data published in *Cancer Incidence In 5 Continents*, Volumes VI-VIII covering a long period of registration (1983 to 1997) showed the highest IR in Northern Europe and Australia and the lowest rates among Asian, Hispanic, and black populations, consistent with other observations of lower rates in pigmented race and a positive association with fair skin.⁵ One of the largest European studies² confirmed the results of the previous paper, but added that IR increased with age and reached a plateau after 75 years. A few hypotheses were provided for this levelling off in older age: susceptible individuals develop cancer due to environmental exposure in adulthood and the pattern is due to a 1- to 2-decade biological lag between the beginning of the exposure and the clinical onset of uveal melanoma; the internal environment of the eye is less stimulating for malignant cells after age 70; or tumour ascertainment is lower for elderly people.

Epidemiological studies have demonstrated that individual exposure to UV radiation is related to the risk of these cancers.⁶ Lesions were reported to occur more often on the left than the right side of the face, and the left side is expected to receive more UV radiation through the driver's side window.⁷ Individuals treated with ionizing radiation as children or adolescents may be at particularly high risk. From the early 1920s to the late 1950s, ionizing radiation was commonly used to treat acne or other inflammatory and benign conditions of the head and neck in the US.⁸

Organ transplant recipients who are immunosuppressed have a greatly increased risk of cutaneous appendageal tumours compared with apparently immunocompetent individuals. In addition, their tumours are more likely to be malignant and of sebaceous origin.⁹ The International Agency for Research on Cancer (IARC) has classified welding with sufficient evidence and solar radiation with limited evidence as risk factors for epithelial tumours of the eye. Occupational exposure to ultraviolet radiation has been described to increase uveal melanoma in workers exposed during outdoor occupational activities and welders.¹⁰

THE EPIDEMIOLOGICAL DATA IN ITALY

Incidence

In 2015, we estimate 451 **tumours of the eye**, most of them are **uveal melanoma** (No. 410). The crude annual (IRs) per 100,000 are 0.1 for epithelial tumours of the eye and adnexa, and 0.6 for uveal melanoma. Slightly more than 50% of cases are over 65 years of age.

Incidence increases with age: in people over 65 years old, IRs are 0.2 and 1.7 for epithelial tumour and melanoma, respectively. The occurrence of uveal melanoma shows an increase from 1.7 in cases in the 65-69 age group, up to 1.9 in cases aged 75-79, then incidence decreases to 1.2 (data not shown).

No difference between genders is relevant statistically. From the SEER program database in the US11 and the RARECARE database in Europe⁴ the same IRs were reported for uveal melanoma for the periods 1973-2008 and 1995-2002, respectively. The age-adjusted incidence trend remained unchanged in the US from 1973 to 2008.¹¹ Cutaneous appendageal carcinoma IRs were reported to increase in the US, especially for sebaceous carcinoma. The authors attributed the increase in trends to improved recognition and classification, and did not exclude factors such as UV exposure and immunosuppression.¹²

Survival

Based on about 1,300 cases, survival analysis shows good 5-year relative survival for both epithelial tumours of the eye and uveal melanoma. Relative survival at 1 and 5 years is 93% and 95%, and 74% and 75%, respectively. Squamous cell carcinoma is characterised by the best prognosis: 82% at 5 years (based on 68 cases). Treatments of uveal melanoma have changed with the progressive introduction of conservative management for smaller tumours during the 1980s.^{11,13} With this therapeutic shift, 5-year RS is reported to be stable both in the US and in Europe.^{11,13}

The Collaborative Ocular Melanoma Study (COMS) demonstrated that metastatic disease survival rate and overall survival was not significantly different between those treated with enucleation and radiotherapy (brachytherapy). Treatment of epithelial tumours of the eye and uveal melanoma is concentrated in high-volume and specialized hospitals.⁴ Patients for which treatments are not available should enter clinical trials. In consideration of the rarity of the disease, international cooperation for research should be arranged.¹

Prevalence

Around 6,000 persons were estimated to be living with a diagnosis of epithelial tumours of the eye and uveal melanoma in Italy in 2010. The majority of these persons (>90%) have a previous diagnosis of uveal melanomas. Our prevalence estimates of uveal melanoma differ from those published in the AIRTUM prevalence monograph,¹⁴ because of the different sites and morphology definition. Here we included ICD-O-3 topographies C69.3-C69.4 and morphologies M8000, M8001, M8720-M8780; the AIRTUM prevalence monograph included ICD-10 C69 + ICD-O-3 morphologies M8720-M8790.¹⁴

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