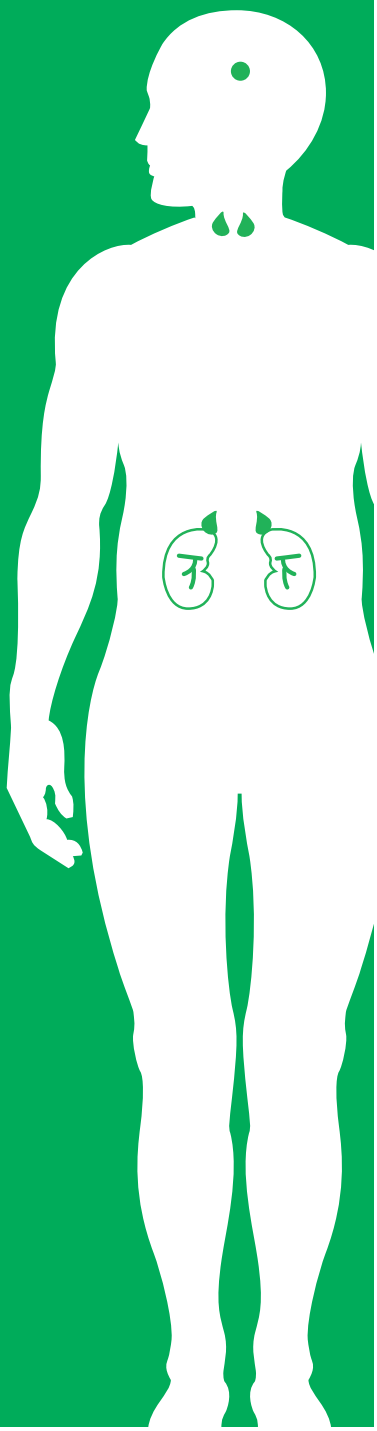
TUMOURS OF THE ENDOCRINE ORGANS

100%
OF ENDOCRINE ORGANS TUMOURS ARE RARE

INCIDENCE

244
ESTIMATED NEW CASES
ITALY, 2015

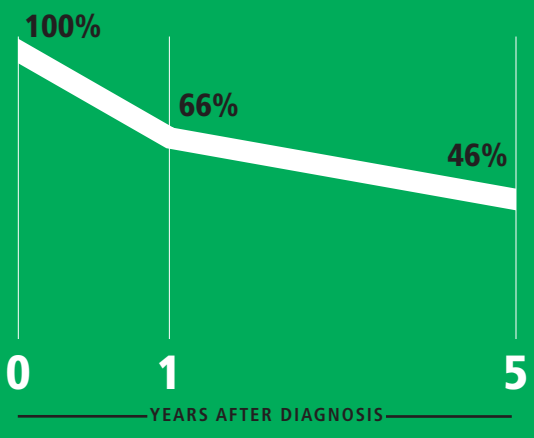
- **22** CARCINOMAS OF PITUITARY GLAND
- **33** CARCINOMAS OF PARATHYROID GLAND
- **189** CARCINOMAS OF ADRENAL CORTEX



PREVALENCE

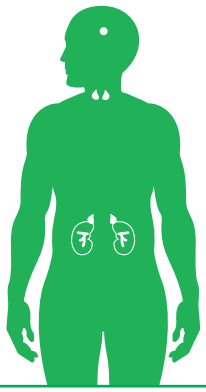
2 222
ESTIMATED PREVALENT CASES
ITALY, 2010

SURVIVAL



SOURCE: AIRTUM. ITALIAN CANCER FIGURES–REPORT 2015

TUMOURS OF THE ENDOCRINE ORGANS

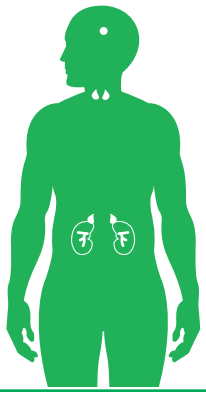


INCIDENCE

TUMOURS OF THE ENDOCRINE ORGANS. 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.

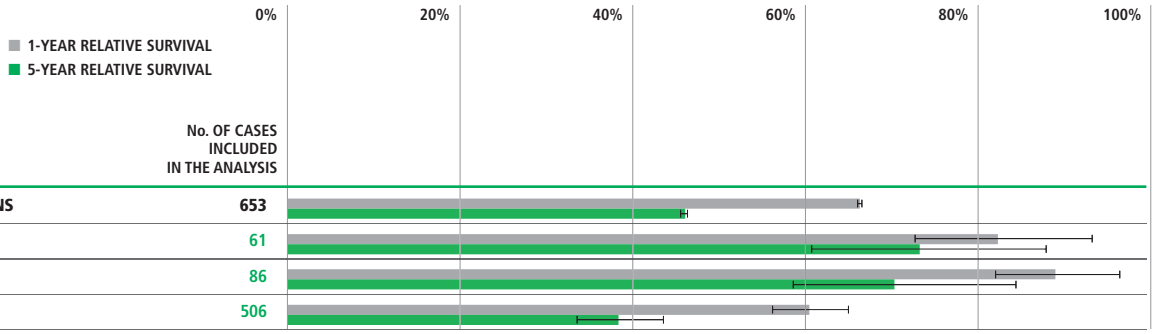
	AIRTUM POOL (period of diagnosis 2000-2010)														ITALY
					SEX				AGE						
					MALE		FEMALE		0-54 yrs		55-64 yrs		65+ yrs		
	RATE	95% CI	OBSERVED CASES (No.)	RARE CANCERS BY SITE (%)	RATE	95% CI	RATE	95% CI	RATE	95% CI	RATE	95% CI	RATE	95% CI	ESTIMATED NEW CASES 2015
TUMOURS OF THE ENDOCRINE ORGANS	**0.37**	**0.35-0.40**	**830**	**100%**	**0.39**	**0.35-0.42**	**0.36**	**0.33-0.40**	**0.20**	**0.18-0.23**	**0.51**	**0.43-0.60**	**0.88**	**0.80-0.97**	**244**
CARCINOMAS OF PITUITARY GLAND	0.03	0.03-0.04	76	NA	0.04	0.03-0.05	0.03	0.02-0.04	0.02	0.01-0.03	0.04	0.02-0.07	0.08	0.05-0.11	22
CARCINOMAS OF PARATHYROID GLAND	0.05	0.04-0.06	110	NA	0.05	0.04-0.06	0.05	0.04-0.07	0.02	0.02-0.03	0.08	0.05-0.13	0.12	0.09-0.16	33
CARCINOMAS OF ADRENAL CORTEX	0.29	0.27-0.31	644	NA	0.30	0.27-0.33	0.28	0.25-0.31	0.16	0.14-0.18	0.38	0.31-0.46	0.68	0.61-0.77	189

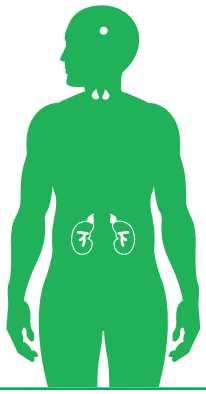
NA: not applicable



SURVIVAL

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





PREVALENCE

TUMOURS OF THE ENDOCRINE ORGANS. 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		
	≤2 YEARS		2-5 YEARS		≤15 YEARS				
	PROPORTION	95% CI	PROPORTION	95% CI	PROPORTION	95% CI	PROPORTION	95% CI	ESTIMATED PREVALENT CASES 2010
TUMOURS OF THE ENDOCRINE ORGANS	**0.46**	**0.33-0.63**	**0.56**	**0.42-0.74**	**2.33**	**2.02-2.67**	**3.74**	**3.21-4.27**	**2 222**
CARCINOMAS OF PITUITARY GLAND	0.05	0.01-0.12	0.21	0.12-0.33	0.62	0.47-0.81	1.07	0.77-1.36	627
CARCINOMAS OF PARATHYROID GLAND	0.08	0.03-0.17	0.11	0.06-0.21	0.47	0.34-0.64	0.77	0.53-1.01	448
CARCINOMAS OF ADRENAL CORTEX	0.33	0.22-0.48	0.24	0.15-0.37	1.24	1.02-1.50	1.91	1.54-2.27	1 147

The list of rare cancers proposed by the European RARECARE project (surveillance of rare cancers in Europe, www.rarecare.eu) considers thyroid cancer arising from the follicular epithelium rare, reporting an incidence rate lower than 6 per 100,000 in Europe. Even if rare at European level, thyroid carcinoma is not presented in the endocrine grouping of this monograph. This choice is due to the fact that the methodological assumptions used to estimate the epidemiological indicators for rare cancers (many of them extremely rare) are not applicable to thyroid cancer epidemiology in Italy (see «Materials and Methods», pp. 14-21).

This section deals in detail with tumours that arise from hormone-secreting endocrine glands, which are all rare:

- **carcinomas of pituitary gland**;
- **carcinomas of parathyroid gland**;
- **carcinomas of adrenal cortex**.

WHAT DO WE KNOW ABOUT THESE CANCERS?

Pituitary tumours are indolent tumours representing approximately 15%-20% of intracranial neoplasms.[1] They can be characterised by pituitary dysfunction, neurological deficits (especially visual impairment), and/or invasion of the parasellar compartment and/or the sphenoid sinuses. Initially considered as sporadic tumours, some of them are associated with familial syndromes such as multiple endocrine neoplasia type 1, Carney complex, or familial isolated pituitary adenomas. Pituitary tumours can be typed based on their hormone-secreting properties into lactotropic (prolactin secreting, 35%), gonadotropic (follicle-stimulating hormone and luteinizing hormone, 35%), somatotropic (growth hormone, GH, 10%-15%), and other tumour types, including tumours with mixed secreting patterns and non-secreting adenomas. Primary pituitary carcinoma is a rare entity defined as any tumour of adenohypophyseal origin with demonstrated craniospinal and/or extracranial metastatic dissemination, fortunately very uncommon and accounting for only 0.1% of all pituitary tumours.[2] Both hormonally active (ACTH-, GH-, and PRL-producing) and hormonally inactive forms of pituitary carcinoma have been reported. Pituitary carcinoma can present in patients with preexisting pituitary adenomas with initial indistinguishable clinical course. The majority of **adrenal cancers a**rise sporadically but can develop as a part of a constellation of tumours in inherited familial cancer syndromes such as Li Fraumeni syndrome, Beckwith-Widerman syndrome, Gardner syndrome, and multiple endocrine neoplasia type 1, each syndrome is associated with unique germ-line mutation.[3] Steroid overproduction is present in over 60% of patients with adrenal cancers. Despite radical surgery with curative intent, the majority of patients with localised adrenal cancers will develop metastases within 6-24 months from resection.[4] **Parathyroid carcinoma** is a rare cause of primary hyperparathyroidism. It can occur either sporadically or in family members affected by hyperparathyroidism-jaw tumour syndrome or associated with multiple endocrine neoplasia.[5] Most parathyroid cancers secrete parathyroid hormone and cause hypercalcaemia. An increase in incidence (from 0.03 to 0.05) was observed in a population-based study in the USA in the period 1988-2003, probably due to increased serum calcium screening.[6]

Pituitary, adrenal, and parathyroid carcinomas are rare, and population-based studies are scarce. Thus, this population-based analysis, which uses data from the pool of the AIRTUM cancer registries, offers the first opportunity to describe the burden of these cancers in Italy.

THE EPIDEMIOLOGICAL DATA IN ITALY

Incidence

Incidence Rate per 100,000 (IR) of endocrine tumours is 0.37; 244 new cases of endocrine tumours are expected in 2015. The first tumour in order of frequency is adrenal cortex cancer. With 189 cases expected, it represents 78% of endocrine tumours considered, with an IR of 0.29 (incidence table, p. 99), slightly higher than the RARECARENet database (IR 0.22) (www.rarecarenet.eu) and equally distributed by sex. The IR progressively increases with age.

One hundred and ten cases of parathyroid cancers were detected, with an IR of 0.05 (see table p. 99), slightly higher compared to the European RARECARENet database (IR of 0.03) representing 13% of this group of rare tumours. Despite the few cases, the incidence seems to increase with age, with no differences between sexes. Since only pituitary carcinoma, based on a morphology code of the ICD-O with a malignant behaviour, has been included in the analysis, only 76 cases of pituitary tumours were detected, with an IR of 0.03 (see table p. 99) similar to that observed in the larger RARECARENet database.

Survival

Relative survival (RS) of endocrine tumours is 66% at 1 year and 46% at 5 years. This value is influenced by the lower survival of adrenal cortex tumours compared to other endocrine tumours. Despite surgical, medical, and chemotherapeutic advances, patients with adrenal cancer show a poor prognosis, with an RS of 38% at 5 years (survival figure, p. 99) in line with European data of the RARECARENet database. This prognosis is most probably due to the high tendency to metastasise within 6-24 months from resection. Parathyroid cancer has an RS of 89% and 70% at 1 and 5 years, respectively (see figure p. 99). Morbidity and mortality for this cancer usually are caused by metabolic complications rather than tumour burden. Similar RS was observed for pituitary tumours (RS 82% at 1 year and 73% at 5 years) (see figure p. 99). These results are similar to those observed in the wider European RARECARENet database.

Prevalence

Slightly more than 2,000 persons are estimated to be living in 2010 with a diagnosis of carcinoma of the endocrine organs (excluding thyroid cancer), of whom more than 50% have a diagnosis of adrenal cortex carcinoma (prevalence table, p. 99). The distribution of prevalent cases by time since diagnosis shows that cases where time since diagnosis is over 15 years are 42% for pituitary carcinoma, 38% for parathyroid carcinoma, and 35% for adrenal cortex carcinoma.

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5. Sharretts JM, Simonds WF. Clinical and molecular genetics of parathyroid neoplasms. *Best Pract Res Clin Endocrinol Metab* 201;24(3):491-502.
6. Lee PK, Jarosek SL, Virnig BA, Evasovich M, Tuttle TM. Trends in the incidence and treatment of parathyroid cancer in the United States. *Cancer* 2007;109(9):1736-1741.