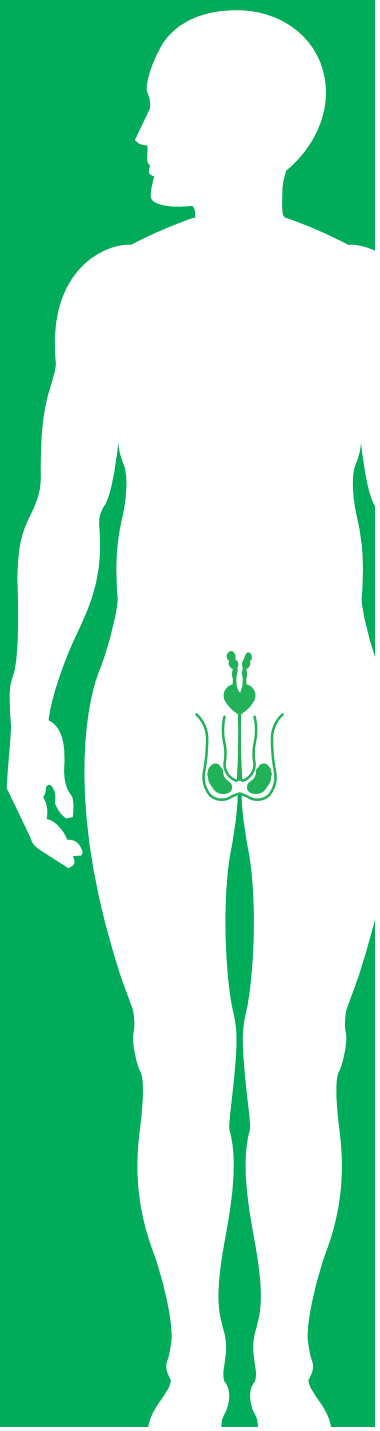
RARE TUMOURS OF THE MALE GENITAL SYSTEM

6%
OF MALE GENITAL SYSTEM TUMOURS ARE RARE

INCIDENCE

2 413
ESTIMATED NEW CASES
ITALY, 2015

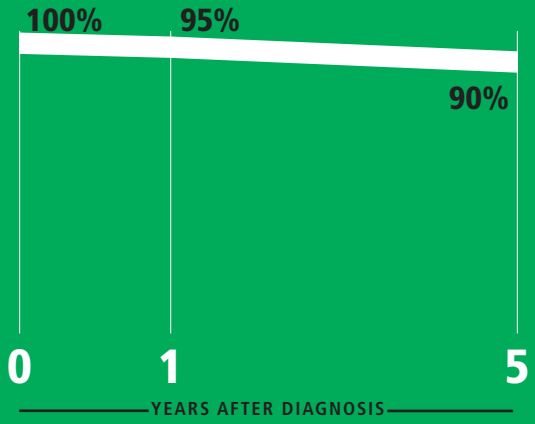
116	RARE EPITHELIAL TUMOURS OF PROSTATE	0.3 % OF RARE TUMOURS OUT OF ALL TUMOURS IN EACH SITE
1 746	TESTICULAR AND PARATESTICULAR TUMOURS	96
473	EPITHELIAL TUMOURS OF PENIS	96
76	EXTRAGONADAL GERM CELL TUMOURS	0.02
1	MESOTHELIOMA OF TUNICA VAGINALIS	



PREVALENCE

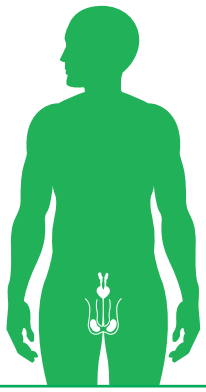
51 030
ESTIMATED PREVALENT CASES
ITALY, 2010

SURVIVAL



SOURCE: AIRTUM. ITALIAN CANCER FIGURES–REPORT 2015

INCIDENCE



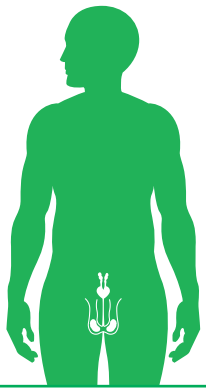
RARE TUMOURS OF THE MALE GENITAL SYSTEM. Crude incidence (rate per 100,000/year) and 95% confidence interval (95% CI), observed cases and proportion of rare cancers on all (common + rare) cancers by site. Rates with 95% CI by sex and age. Estimated new cases at 2015 in Italy.

	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
RARE TUMOURS OF THE MALE GENITAL SYSTEM	**4.08**	**4.00-4.17**	**9 049**	**6%**	**8.36**	**8.19-8.54**	**–**	**–**	**4.60**	**4.49-4.71**	**2.29**	**2.11-2.47**	**3.43**	**3.25-3.60**	**2 413**
RARE EPITHELIAL TUMOURS OF PROSTATE	**0.17**	**0.15-0.19**	**374**	**0.3%**	**0.35**	**0.31-0.39**	**–**	**–**	**<0.01**	**0.00-0.01**	**0.24**	**0.18-0.30**	**0.68**	**0.60-0.76**	**116**
Squamous cell carcinoma with variants of prostate	0.01	0.00-0.01	16		0.01	0.01-0.02	–	–	0.00	–	0.01	0.00-0.03	0.03	0.02-0.05	5
Infiltrating duct carcinoma of prostate	0.11	0.09-0.12	234		0.22	0.19-0.25	–	–	<0.01	0.00-0.01	0.16	0.11-0.21	0.42	0.36-0.49	72
Transitional cell carcinoma of prostate	0.05	0.04-0.06	119		0.11	0.09-0.13	–	–	<0.01	0.00-0.01	0.06	0.03-0.10	0.22	0.18-0.27	38
Basal cell adenocarcinoma of prostate	<0.01	0.00-0.01	5		NE	–	–	–	NE	–	NE	–	NE	–	1
TESTICULAR AND PARATESTICULAR TUMOURS	**3.10**	**3.03-3.18**	**6 877**	**96%**	**6.41**	**6.26-6.56**	**–**	**–**	**4.27**	**4.17-4.38**	**0.83**	**0.72-0.94**	**0.50**	**0.43-0.57**	**1 746**
Paratesticular adenocarcinoma with variants	<0.01	0.00-0.00	2		NE	–	–	–	NE	–	NE	–	NE	–	1
Non seminomatous testicular tumours	1.07	1.02-1.11	2 363		2.20	2.11-2.29	–	–	1.53	1.47-1.60	0.12	0.08-0.17	0.06	0.04-0.08	586
Seminomatous testicular tumours	1.70	1.65-1.76	3 777		3.52	3.41-3.64	–	–	2.35	2.27-2.42	0.55	0.47-0.65	0.22	0.17-0.26	964
Spermatocytic seminoma	0.05	0.04-0.06	103		0.10	0.08-0.12	–	–	0.04	0.03-0.05	0.03	0.02-0.06	0.08	0.05-0.11	29
Teratoma with malignant transformation	<0.01	0.00-0.01	5		NE	–	–	–	NE	–	NE	–	NE	–	1
Testicular sex cord tumours	0.02	0.02-0.03	55		0.05	0.04-0.07	–	–	0.03	0.02-0.03	0.02	0.01-0.05	0.03	0.01-0.04	15
EPITHELIAL TUMOURS OF PENIS	**0.68**	**0.65-0.72**	**1 509**	**96%**	**1.41**	**1.34-1.48**	**–**	**–**	**0.15**	**0.13-0.18**	**1.16**	**1.03-1.29**	**2.19**	**2.05-2.33**	**473**
Squamous cell carcinoma with variants of penis	0.63	0.60-0.67	1 406		1.31	1.24-1.38	–	–	0.14	0.13-0.16	1.10	0.98-1.23	2.02	1.89-2.16	438
Adenocarcinoma with variants of penis	<0.01	0.00-0.01	13		NE	–	–	–	NE	–	NE	–	NE	–	4
EXTRAGONADAL GERM CELL TUMOURS	**0.13**	**0.11-0.14**	**284**	**0.02%**	**0.19**	**0.16-0.22**	**–**	**–**	**0.16**	**0.14-0.18**	**0.06**	**0.04-0.10**	**0.05**	**0.03-0.08**	**76**
Non seminomatous germ cell tumours	0.06	0.05-0.08	142		0.08	0.07-0.10	–	–	0.07	0.06-0.09	0.04	0.02-0.08	0.04	0.02-0.06	38
Seminomatous germ cell tumours	0.01	0.01-0.02	29		0.02	0.02-0.04	–	–	0.02	0.01-0.03	<0.01	0.00-0.03	0.00	–	8
Germ cell tumours of Central Nervous System (CNS)	0.04	0.03-0.05	81		0.06	0.04-0.07	–	–	0.05	0.04-0.06	<0.01	0.00-0.02	<0.01	0.00-0.02	22
MESOTHELIOMA OF TUNICA VAGINALIS	**<0.01**	**0.00-0.01**	**5**		**NE**	**–**	**–**	**–**	**NE**	**–**	**NE**	**–**	**NE**	**–**	**1**

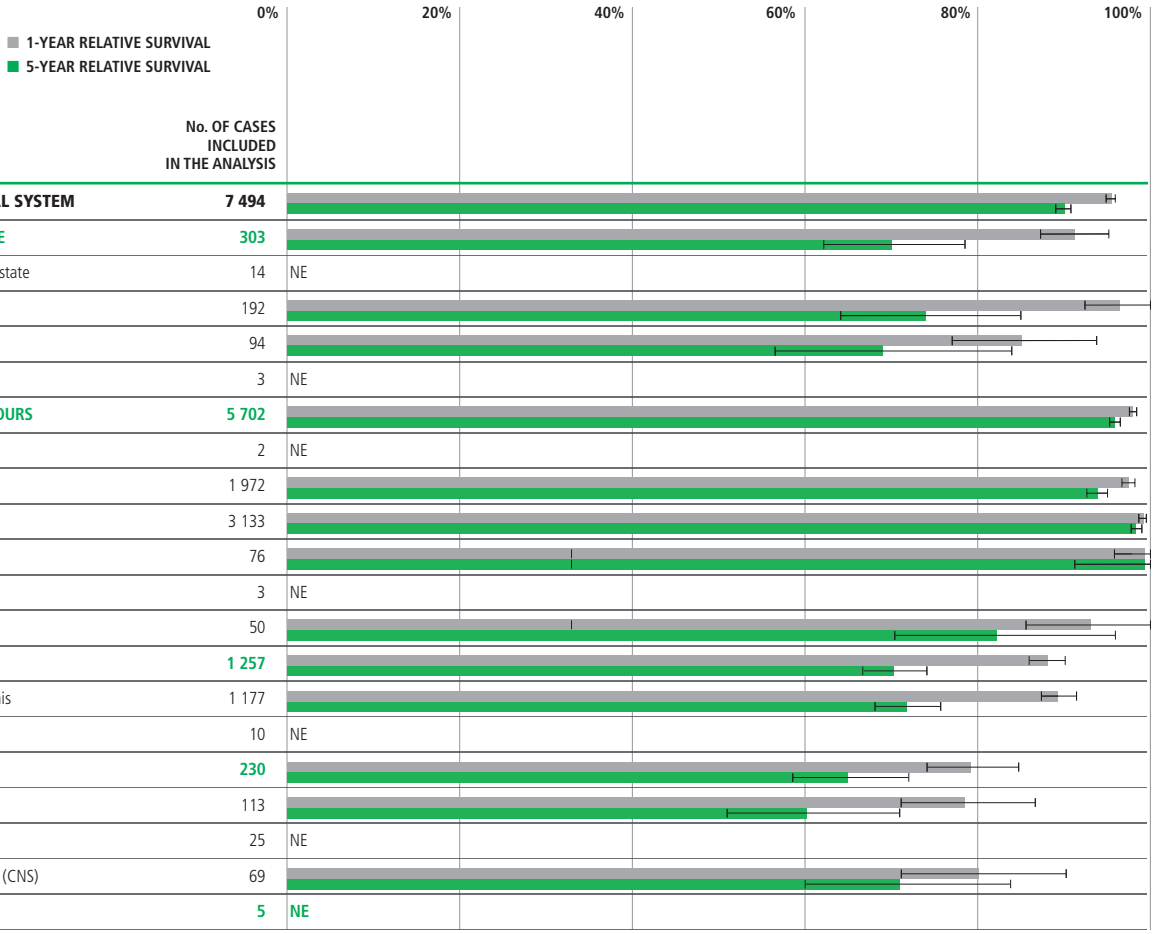
NE: not estimable because 15 or less incident cases were observed

RARE TUMOURS OF THE MALE GENITAL SYSTEM

SURVIVAL



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

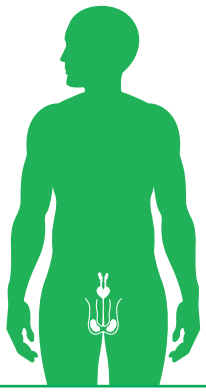


	No. OF CASES INCLUDED IN THE ANALYSIS
RARE TUMOURS OF THE MALE GENITAL SYSTEM	**7 494**
RARE EPITHELIAL TUMOURS OF PROSTATE	**303**
Squamous cell carcinoma with variants of prostate	14
Infiltrating duct carcinoma of prostate	192
Transitional cell carcinoma of prostate	94
Basal cell adenocarcinoma of prostate	3
TESTICULAR AND PARATESTICULAR TUMOURS	**5 702**
Paratesticular adenocarcinoma with variants	2
Non seminomatous testicular tumours	1 972
Seminomatous testicular tumours	3 133
Spermatocytic seminoma	76
Teratoma with malignant transformation	3
Testicular sex cord tumours	50
EPITHELIAL TUMOURS OF PENIS	**1 257**
Squamous cell carcinoma with variants of penis	1 177
Adenocarcinoma with variants of penis	10
EXTRAGONADAL GERM CELL TUMOURS	**230**
Non seminomatous germ cell tumours	113
Seminomatous germ cell tumours	25
Germ cell tumours of Central Nervous System (CNS)	69
MESOTHELIOMA OF TUNICA VAGINALIS	**5**

	NE
RARE EPITHELIAL TUMOURS OF PROSTATE	
Squamous cell carcinoma with variants of prostate	NE
Infiltrating duct carcinoma of prostate	
Transitional cell carcinoma of prostate	
Basal cell adenocarcinoma of prostate	NE
TESTICULAR AND PARATESTICULAR TUMOURS	
Paratesticular adenocarcinoma with variants	NE
Non seminomatous testicular tumours	
Seminomatous testicular tumours	
Spermatocytic seminoma	
Teratoma with malignant transformation	NE
Testicular sex cord tumours	
EPITHELIAL TUMOURS OF PENIS	
Squamous cell carcinoma with variants of penis	
Adenocarcinoma with variants of penis	NE
EXTRAGONADAL GERM CELL TUMOURS	
Non seminomatous germ cell tumours	
Seminomatous germ cell tumours	NE
Germ cell tumours of Central Nervous System (CNS)	
MESOTHELIOMA OF TUNICA VAGINALIS	**NE**

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

PREVALENCE



RARE TUMOURS OF THE MALE GENITAL SYSTEM. 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
RARE TUMOURS OF THE MALE GENITAL SYSTEM	**7.83**	**7.26-8.44**	**9.93**	**9.28-10.62**	**41.95**	**40.60-43.34**	**88.72**	**84.78-92.66**	**51 030**
RARE EPITHELIAL TUMOURS OF PROSTATE	**0.21**	**0.12-0.33**	**0.18**	**0.11-0.30**	**0.88**	**0.70-1.11**	**0.93**	**0.72-1.14**	**539**
Squamous cell carcinoma with variants of prostate	0.01	0.00-0.06	NE	–	0.02	0.00-0.08	0.02	0.00-0.06	14
Infiltrating duct carcinoma of prostate	0.14	0.07-0.24	0.06	0.02-0.13	0.56	0.42-0.74	0.58	0.42-0.74	333
Transitional cell carcinoma of prostate	0.06	0.02-0.13	0.13	0.06-0.23	0.30	0.20-0.44	0.33	0.20-0.45	192
Basal cell adenocarcinoma of prostate	NE	–	NE	–	NE	–	NE	–	NE
TESTICULAR AND PARATESTICULAR TUMOURS	**6.27**	**5.76-6.82**	**8.54**	**7.94-9.18**	**35.66**	**34.42-36.94**	**78.39**	**74.62-82.17**	**45 055**
Paratesticular adenocarcinoma with variants	NE	–	NE	–	NE	–	NE	–	NE
Non seminomatous testicular tumours	1.92	1.64-2.23	2.80	2.46-3.18	11.86	11.14-12.60	29.53	26.37-32.69	16 779
Seminomatous testicular tumours	3.87	3.47-4.31	5.06	4.60-5.55	21.49	20.52-22.48	44.49	41.55-47.42	25 649
Spermatocytic seminoma	0.06	0.02-0.13	0.09	0.04-0.18	0.42	0.30-0.59	1.33	0.76-1.91	23 047
Teratoma with malignant transformation	NE	–	NE	–	0.01	0.00-0.06	0.01	0.00-0.04	6
Testicular sex cord tumours	NE	–	0.06	0.02-0.13	0.20	0.11-0.31	0.41	0.21-0.60	232
EPITHELIAL TUMOURS OF PENIS	**1.17**	**0.96-1.42**	**0.99**	**0.79-1.22**	**4.39**	**3.96-4.86**	**6.16**	**5.54-6.78**	**3 562**
Squamous cell carcinoma with variants of penis	1.15	0.93-1.40	0.95	0.76-1.18	4.22	3.80-4.68	5.85	5.25-6.45	3 377
Adenocarcinoma with variants of penis	NE	–	NE	–	0.01	0.00-0.06	0.01	0.00-0.04	7
EXTRAGONADAL GERM CELL TUMOURS	**0.18**	**0.11-0.30**	**0.22**	**0.13-0.34**	**1.02**	**0.82-1.26**	**3.23**	**2.30-4.16**	**1 874**
Non seminomatous germ cell tumours	0.08	0.03-0.17	0.09	0.04-0.18	0.47	0.34-0.64	1.11	0.73-1.49	652
Seminomatous germ cell tumours	0.03	0.01-0.10	0.05	0.01-0.12	0.17	0.10-0.28	0.25	0.12-0.39	150
Germ cell tumours of Central Nervous System (CNS)	0.06	0.02-0.13	0.05	0.01-0.12	0.30	0.20-0.44	0.42	0.22-0.62	244
MESOTHELIOMA OF TUNICA VAGINALIS	**NE**	**–**	**NE**	**–**	**NE**	**–**	**NE**	**–**	**NE**

NE: not estimable in observed prevalence if no cases were observed within ≤2, 2-5, ≤15 years prior to prevalence date, in complete prevalence if the 15-year prevalence is NE

This group includes a heterogeneous number of rare cancers:
- **rare epithelial tumours of the prostate** (squamous cell carcinoma, infiltrating duct carcinoma, transitional cell carcinoma, basal cell adenocarcinoma);
- **testicular cancers** (paratesticular adenocarcinoma, non-seminomatous testicular cancer, seminomatous testicular cancer; spermatocytic seminoma, teratoma with malignant transformation, testicular sex cord cancers);
- **epithelial tumours of the penis** (squamous cell carcinomas, adenocarcinomas);
- **extragonadal germ cell tumours** (non-seminomatous germ cell tumours, seminomatous germ cell tumours, and germ cell tumours of the Central Nervous System);
- **mesothelioma of the tunica vaginalis testis**.

Extragonadal germ cell tumours include a few entities that occur even in women; these are also considered in this group since clinical management is the same for females as for males. Overall, rare cancers account for 6% of all male genital system cancers.

RARE EPITHELIAL TUMOURS OF THE PROSTATE

WHAT DO WE KNOW ABOUT THESE CANCERS?

Tumours with **squamous cell differentiation** may arise from the urothelial cells of the prostatic urethra, the periurethral glands, as from a stem cell.[1] They may be in the pure squamous carcinoma form, which usually does not express serum PSA,[2] or associated with acinar adenocarcinoma (adenosquamous carcinoma), urothelial carcinoma, or sarcoma;[1] they are associated with early development of osteolytic metastases and poor responsiveness to antiandrogen therapy.[2]

Regarding infiltrating duct carcinomas, the mixed variant (adenocarcinoma-ductal carcinoma) is more common than pure ductal carcinoma; the latter arises in periurethral prostatic ducts and shows Gleason score 8;[2] this histotype may have a prevalent papillary, or solid or complex cribriform pattern of growth, and is more likely to metastasise to testis and penis.[2] **Transitional cell carcinoma** in the absence of bladder carcinoma may arise from the prostatic urethra or major prostatic ducts, which are lined by urothelial epithelium; it is often difficult to distinguish the origin of this neoplasm, due the possibility of an intraprostatic extension from bladder carcinoma.

Basal cell adenocarcinoma of the prostate is a very rare histological type described for the first time in 1974; it arises in the basal cell layer of the prostate gland and is very likely to cause urinary obstructive symptoms at clinical presentation; it is reported to behave less aggressively than other histotypes.[2] It is frequently associated with the acinar variant and only about 50 cases are described in the literature.[3]

Risk factors associated with common prostatic adenocarcinoma (age, familiarity, obesity, lifestyle, environmental exposure) most likely have a role even in the aetiology of rare prostatic cancers, except for squamous cell carcinoma. About half of squamous cell carcinomas arise after androgen deprivation therapy or radiation treatment for a conventional adenocarcinoma. However, some cases have been reported as *de novo* cancers in patients without prostate disease.

THE EPIDEMIOLOGICAL DATA IN ITALY

Incidence

Infiltrating duct carcinoma is the most common (63%), followed by transitional carcinoma (32%), squamous (4%), and basal cell adenocarcinoma (1%). Like the common acinar adenocarcinoma of the prostate, even these rare epithelial tumours are typical of advanced age (>65 years), with exceptional cases of early-onset prostate cancer (diagnosed <55 years). Approximately 120 Italians were estimated to be diagnosed with rare tumours of the prostate in 2015.

Survival

Infiltrating duct carcinoma and transitional carcinomas have a good prognosis (5-year relative survival: 74% and 69%, respectively). In the AIRTUM database, the number of cases is too limited to provide survival of squamous cell carcinoma and basal cell adenocarcinoma. However, data from the European RARECAREnet database (www.rarecarenet.eu) show that squamous cell carcinomas have a poor survival rate (40% at 5 years), while basal cell adenocarcinoma have a good survival rate (80%).

The European RARECAREnet data show that rare epithelial cancers of the prostate are characterised by a worse prognosis than the common acinar adenocarcinoma of the prostate (88%). The lower prognosis of these rare cancers seems mainly due to the more advanced stage at diagnosis and some resistance to treatment, particularly to hormonal therapy. They usually occur in people older than those diagnosed with acinar adenocarcinoma and survival decreases with increasing age.

Prevalence

In Italy, slightly more than 500 persons were estimated to be alive in 2010 with a diagnosis of rare epithelial tumours of the prostate. The most prevalent cases are infiltrating duct carcinomas of the prostate, followed by transitional cell carcinomas.

TESTICULAR CANCERS

WHAT DO WE KNOW ABOUT THESE CANCERS?

Testicular cancers are all rare, but are the most common cancers in young men, and their incidence is increasing.[4] The incidence varies in different geographical areas, thus testicular cancer may not be perceived as rare in countries of Northern and Western Europe.[4] However, as described in «Material and methods» (pp. 14-21), the definition of rarity is based on the European population and not on the country-specific incidence. Thus, even though the country-specific incidence rate of testicular cancer can be higher than 6 in some countries, testicular cancer is considered rare because its incidence is <6 per 100,000 in the EU.

Seminomatous and non-seminomatous cancers are the most common germ cell tumours of the testis. Seminomatous cancers are more often localised, metastasise via the lymphatic system, and are radiosensitive; they occur in somewhat older patients. By contrast, non-seminomatous cancers are prone to haematogenous as well as lymphatic spread and are less radiosensitive.[4] **Spermatocytic seminoma** is clinically and pathologically distinct from classic seminomatous cancer, in particular for its almost complete inability to metastasise.[4] On rare occasions, teratomas undergo somatic ma-

lignant transformation.[5] The most common transformations are to sarcoma, primitive neuroectodermal tumour, and adenocarcinoma. Most **sex cord cancers** have a benign clinical course following surgery, but about 20% are metastatic at diagnosis and 10%-12% behave aggressively, often with fatal outcome.[4]

THE EPIDEMIOLOGICAL DATA IN ITALY

Incidence

Seminomatous testicular cancer is the most common entity (55%), followed by non-seminomatous testicular cancer (34%). The other germ cell tumours are very rare. For seminomatous cancers, the incidence rate (IR) peaks in the 30-34 age group (0.5 per 100,000; data not shown). For non-seminomatous cancers, the IR peaks in the 25-29 age group (4 per 100,000; data not shown). Until around 24 years of age non-seminomatous cancers are the predominant histologic type, after around 29 years of age seminomas are more common than non seminomas. Among the elderly (65 years and over) 62% of germ cell tumours are seminomas. A total of 1,746 testicular cancers were estimated to be diagnosed in Italy at 2015; 90% of these cancers are germ cell tumours.

Survival

Among testicular and paratesticular cancers, seminomas and spermatocytic seminoma have the highest survival, followed by non seminomas. Five-year relative survival (RS) is also good for sex cord tumours. Survival differences between seminomatous and non-seminomatous cancers are explained by their different biology. The good prognosis of spermatocytic seminoma and sex cord tumours is mainly explained by their benign clinical course. The AIRTUM database has too few cases to provide survival of the other entities. However, regarding teratoma with malignant transformation, studies suggest that transformation has a negative impact on prognosis compared to the non-transformed counterpart.[5]

Prevalence

Around 45,000 persons were estimated to be living with a diagnosis of testicular cancers in Italy in 2010. The most prevalent testicular cancers are seminomatous and non-seminomatous cancers. Young age of patients may play an important role in so relatively high prevalence figures. For testicular cancers, however, the high survival rate appears as the most important contributor to the prevalence. Our estimates are slightly higher than those published in the AIRTUM monograph on prevalence[6] because of the different methodology used (see «Materials and methods», pp.14-21).

EPITHELIAL TUMOURS OF THE PENIS

WHAT DO WE KNOW ABOUT THESE CANCERS?

Epithelial tumours of the penis are all rare. Risk factors for the development of penile cancer are multifactorial and include: phimosis, infection with human papilloma virus (HPV), HIV infection, cigarette smoking, history of trauma and chronic balanitis, lichen sclerosus, and PUVA treatment.[2] Circumcision in infancy is associated with a protective effect for penile cancer.[2] Penile cancer is a very rare tumour for which a referral to a centre of expertise is recommended, since it deserves special attention in the diagnosis process and treatment options. Penectomy is disfiguring and can have an intense effect on the patient's quality of life, sexual function, self-esteem, and general mental health. Therefore, there is an increased trend for penile preserving strategies (radiotherapy), despite the fact that recurrence rates may be higher than those of radical surgical procedures.

THE EPIDEMIOLOGICAL DATA IN ITALY

Incidence

Cancer of the penis is the rarest cancer of the male genital system. The IR in Italy is 0.7 per 100,000 and it is equal to that observed in the European RARECAREnet database. As HPV infection is an important risk for penile cancer and circumcision is protective, the intermediate position of Italy and Europe can be explained by moderate rates of HPV-infection in a generally uncircumcised male population. Squamous cell carcinoma is the most common morphological type of penis cancer (93%). This tumour is very rare before 50 years old; the IR increases with age, with the highest IR in patients 75 years or older. About 470 penile cancers were estimated to be diagnosed in Italy in 2015.

Survival

Survival of squamous cell carcinoma of the penis is good and similar to that observed in the European RARECAREnet database. In the AIRTUM database the number of cases is too limited to provide data for adenocarcinoma of the penis. The European RARECAREnet database reports a 5-year RS of 50%. Although survival for penile cancers is relatively good, it has not improved in the past few years, probably because of the limited advances in curative treatment and the limited centralisation of treatment.[7,8]

Prevalence

In Italy, 3,500 persons were estimated to be alive with a diagnosis of penile cancer in 2010. The most prevalent penile cancer is squamous cell carcinoma.

EXTRAGONADAL GERM CELL TUMOURS (EGCTs)

WHAT DO WE KNOW ABOUT THESE CANCERS?

Extragonadal germ cell tumours are all rare. Germ cell tumours of the CNS are typical of children and adolescents and, therefore, are mainly managed by paediatric oncologists. EGCTs represent a minor part of all germ cell tumours and are more aggressive than those of the gonads. They usually occur along the midline of the body along which the primordial germ cells migrate from the proximal epiblast to the genital ridge.

THE EPIDEMIOLOGICAL DATA IN ITALY

Incidence

EGCTs account for 4% of all germ cell tumours in the AIRTUM database (data not shown). EGCTs are more common in males than females and, in all sites, non seminoma is the predominant histology. Germ cell tumours of the CNS are typical of children and adolescents (10-19 years old). Non seminomas have a first incidence peak in the 0-4-year age group and a second in the 25-29-year age group. Seminomas are very rare and have an incidence peak in the 35-39-year age group. Sites are predominantly the me-

diastinum (29%), CNS (17%), endocrine system (12%), female genital system (9%), retroperitoneum (5%), with primary site unknown in 3%. The remaining cases occur at disparate sites including head and neck, digestive tract, lung, prostate, urinary tract, and other not specified sites (ICD-O3 C76.0-76.8).

Survival

Five-year RS of EGCTs is always worse than that of gonadal germ cell tumours. Five-year RS is 71% for CNS EGCTs and 60% for non-seminomatous EGCTs (data not shown). Among the latter, those of the mediastinum (most common site in the AIRTUM database) has the worst survival. This could be due to generally large tumour bulk at diagnosis, resistance to chemotherapy, difficulty of removing all residual disease after chemotherapy, and a predisposition to develop haematologic neoplasia and other non-germ cell malignancies. In the AIRTUM database the number of cases is too limited to provide data for seminomatous EGCTs. However, the European RARECAREnet database reports a 5-year RS of 85%.

Prevalence

About 1,900 persons were estimated to be living with a diagnosis of extragonadal germ cell cancer in Italy in 2010.

MESOTHELIOMA OF THE TUNICA VAGINALIS TESTIS

Only 5 cases of **malignant mesothelioma of the tunica vaginalis** (MTVT) were observed in the AIRTUM database in the period of diagnosis 2000-2010, confirming the extreme rarity of this tumour. The data of the National Italian malignant Mesothelioma Registry (ReNAM) reported 51 cases of MTVT diagnosed in the period 1993-2008, corresponding to 0.3% of all mesothelioma cases in Italy.[9] Differences in number of cases observed are due to the national coverage of the ReNAM, which is a mesothelioma-dedicated registry, and the partial coverage of the AIRTUM registries (which are general registries). The highest incidence is observed in the 65-75-year age group.

An Italian study found exposure to asbestos in 67% of cases of MTVT and clarified that the difference in the percentage of asbestos-related MTVT reported in the literature (30%-40%) might be the result of an incomplete investigation of exposure history for MTVT patients.[10] Thus, asbestos exposure is the main risk factor for MTVT, as well as for malignant mesotheliomas that occur at other sites, although the mechanism of involvement of the tunica vaginalis by asbestos fibres still remains unclear.

In the AIRTUM database the number of cases is too limited to estimate survival. However, according to the literature, survival is very bad, ranging from a minimum of 18 months without treatment[11] to a maximum of 23 months for patients treated with surgery.[12]

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