

Supplementary data

I tumori rari in Italia / The burden of rare cancer in Italy  
Epidemiol Prev 2016; 40(1 Suppl 2): 1-120

R=rare	Tier	Tumour	Crude incidence rate per 100,000 per year in Europe	95% confidence interval		Number of cases collected in the RARECAREnet database from 2000-2007	ICD-O-3 Topography code	ICD-O-3 Morphology code
	1	<b>EPITHELIAL TUMOURS OF NASAL CAVITY AND SINUSES</b>	0,45	0,44	0,46	7.046	C30.0, C31	8000, 8001, 8003,8004, 8010, 8011, 8012, 8020-8022, 8030,8031, 8032, 8033-8034, 8046, 8050-8076, 8078, 8082-8084, 8120, 8121,8122, 8123, 8130, 8140, 8144, 8560, 8980
R	2	<b>Squamous cell carcinoma with variants of nasal cavity and sinuses</b>	0,35	0,34	0,36	5.465	C30.0, C31	8004, 8120, 8121,8122, 8130, 8032, 8050-8076, 8078, 8083-8084, 8123, 8560, 8980
	3	Squamous carcinoma					C30.0, C31	8070
	3	Verrucous carcinoma					C30.0, C31	8051
	3	Squamous cell carcinoma spindle cell					C30.0, C31	8004, 8032, 8074, 8980
	3	Papillary squamous cell carcinoma					C30.0, C31	8052
	3	Adenosquamous carcinoma					C30.0, C31	8560
	3	Squamous cell carcinoma, adenoid					C30.0, C31	8075
	3	Basaloid squamous cell carcinoma					C30.0, C31	8083
R	2	<b>Lymphoepithelial carcinoma of nasal cavity and sinuses</b>	0,002	0,001	0,003	31	C30.0, C31	8082
R	2	<b>Undifferentiated carcinoma of nasal cavity and sinuses</b>	0,02	0,02	0,02	286	C30.0, C31	8020-8022
R	2	<b>Intestinal type adenocarcinoma of nasal cavity and sinuses</b>	0,003	0,002	0,004	42	C30.0, C31	8144, 8140
	1	<b>EPITHELIAL TUMOURS OF NASOPHARYNX</b>	0,14	0,13	0,14	2.133	C11	8000-8001, 8004, 8010-8011, 8012, 8020-8022, 8032, 8033, 8034, 8046, 8120, 8121, 8122, 8050-8076, 8078, 8082-8084, 8123, 8260, 8560, 8980
R	2	<b>Squamous cell carcinoma with variants of nasopharynx</b>	0,11	0,11	0,12	1.786	C11	8004, 8020-8022, 8032, 8051-8076, 8078, 8082-8084, 8120, 8121, 8122, 8123, 8560, 8980
	3	Squamous carcinoma					C11	8070
	3	Squamous cell carcinoma nonkeratinizing, NOS					C11	8072
	3	Squamous cell carcinoma keratinizing, NOS					C11	8071
	3	Papillary squamous cell carcinoma					C11	8052
	3	Basaloid squamous cell carcinoma					C11	8083
	3	Squamous cell carcinoma, adenoid					C11	8075
	3	Lymphoepithelial carcinoma					C11	8082
	3	Undifferentiated carcinoma					C11	8020-8022
R	2	<b>Papillary adenocarcinoma of nasopharynx</b>	0,0004	#####	0,001	6	C11	8050, 8260
	1	<b>EPITHELIAL TUMOURS OF MAJOR SALIVARY GLANDS AND SALIVARY-GLAND TYPE TUMOURS</b>	1,39	1,37	1,41	21.794	C00-C14, C30.0, C31, C32	<b>C00-C06, C09-C14, C30.0, C31, C32:</b> 8140, 8141,8143, 8144 (not in C30.0,C31), 8147, 8190, 8200, 8201,8211, 8230, 8231, 8261, 8263, 8320, 8323, 8401,8490,8510,8551,8570, 8574, 8675, 8290, 8310, 8430, 8440, 8450, 8480-8481, 8500, 8525, 8550, 8562, 8941, 8982; <b>C7, C8:</b> 8000,8001, 8003, 8004, 8005, 8010, 8011, 8012, 8014, 8015, 8020-8022, 8030,8031, 8032, 8033, 8034, 8035, 8046, 8050-8076, 8078, 8082, 8083, 8084, 8120, 8121, 8122, 8123, 8015, 8020-8022, 8030,8031, 8032, 8033, 8034, 8035, 8046, 8050-8076, 8078, 8082, 8083, 8084, 8120, 8121, 8122, 8123, 8130, 8140, 8141, 8143, 8190, 8147, 8200, 8201, 8211, 8230, 8231, 8255, 8260, 8261, 8262, 8263, 8290, 8310, 8320, 8323, 8401, 8410, 8430, 8440, 8450, 8480-8481, 8490, 8500, 8510, 8525, 8550, 8551, 8560, 8561, 8570, 8574,8575, 8562, 8940, 8941, 8951, 8980, 8981, 8982.
R	2	<b>Epithelial tumours of major salivary glands</b>	0,96	0,95	0,98	15.053	C7, C8	8000,8001, 8003, 8004, 8005, 8010, 8011, 8012, 8014, 8015, 8020-8022, 8030,8031, 8032, 8033, 8034, 8035, 8046, 8050-8076, 8078, 8082, 8083, 8084, 8120, 8121, 8122, 8123, 8130, 8140, 8141, 8143, 8144, 8190, 8147, 8200, 8201, 8211, 8230, 8231, 8255, 8260, 8261, 8262, 8263, 8290, 8310, 8320, 8323, 8401, 8410, 8430, 8440, 8450, 8480-8481, 8490, 8500, 8510, 8525, 8550, 8551, 8560, 8561, 8570, 8574,8575, 8562, 8940, 8941, 8980, 8981, 8982.
	3	Squamous carcinoma					C7, C8	8070
	3	Large cell carcinoma					C7, C8	8012
	3	Lymphoepithelial carcinoma					C7, C8	8082
	3	Carcinosarcoma, NOS					C7, C8	8004, 8032, 8074, 8980
	3	Adenocarcinoma NOS					C7, C8	8140
	3	Clear cell adenocarcinoma NOS					C7, C8	8310
	3	Mucinous adenocarcinoma					C7, C8	8480-8481

R=rare	Tier	Tumour	Crude incidence rate per 100,000 per year in Europe	95% confidence interval		Number of cases collected in the RARECAREnet database from 2000-2007	ICD-O-3 Topography code	ICD-O-3 Morphology code
	3	Ductal carcinoma					C7, C8	8500
	3	Oxyphilic adenocarcinoma					C7, C8	8290
	3	Papillary cystadenocarcinoma, NOS					C7, C8	8450
	3	Adenoid cystic carcinoma					C7, C8	8200
	3	Mucoepidermoid carcinoma					C7, C8	8430
	3	Acinar cell adenocarcinoma					C7, C8	8550
	3	Malignant myoepithelioma					C7, C8	8982
	3	Carcinoma in pleomorphic adenoma					C7, C8	8941
	3	Mixed tumour, malignant					C7, C8	8940
	3	Polymorphous low grade adenocarcinoma					C7, C8	8525
	3	Basal cell adenocarcinoma					C7, C8	8147
	3	Epithelial-myoepithelial carcinoma					C7, C8	8562
	3	Sebaceous adenocarcinoma					C7, C8	8410
	3	Cystadenocarcinoma, NOS					C7, C8	8440
	3	Low grade cribriform cystadenocarcinoma					C7, C8	malignancy without ICD-O morphology code
<b>R</b>	2	<b>Salivary gland type tumours of head and neck</b>	0.43	0.42	0.44	6.741	C00-C06, C09-C14, C30.0, C31, C32	8140, 8141, 8143, 8144 (not in C30.0, C31), 8147, 8190, 8200, 8201, 8211, 8230, 8231, 8261, 8263, 8320, 8323, 8401, 8490, 8510, 8551, 8570, 8574, 8675, 8290, 8310, 8430, 8440, 8450, 8480-8481, 8500, 8525, 8550, 8562, 8941, 8982;
	3	Mucoepidermoid carcinoma					C00-C06, C09-C14, C30.0, C31, C32	8430
	3	Adenocarcinoma NOS					C00-C06, C09-C14, C30.0, C31, C32	8140
	3	Adenoid cystic carcinoma					C00-C06, C09-C14, C30.0, C31, C32	8200
	3	Acinar cell adenocarcinoma					C00-C06, C09-C14, C30.0, C31, C32	8550
	3	Ductal carcinoma					C00-C06, C09-C14, C30.0, C31, C32	8500
	3	Mucinous adenocarcinoma					C00-C06, C09-C14, C30.0, C31, C32	8480-8481
	3	Basal cell adenocarcinoma					C00-C06, C09-C14, C30.0, C31, C32	8147
	3	Clear cell adenocarcinoma, NOS					C00-C06, C09-C14, C30.0, C31, C32	8310
	3	Epithelial-myoepithelial carcinoma					C00-C06, C09-C14, C30.0, C31, C32	8562
	3	Polymorphous low grade adenocarcinoma					C00-C06, C09-C14, C30.0, C31, C32	8525
	3	Carcinoma in pleomorphic adenoma					C00-C06, C09-C14, C30.0, C31, C32	8941
	3	Malignant myoepithelioma					C00-C06, C09-C14, C30.0, C31, C32	8982
	3	Cystadenocarcinoma, NOS					C00-C06, C09-C14, C30.0, C31, C32	8440
	3	Oncocytic carcinoma					C00-C06, C09-C14, C30.0, C31, C32	8290
	3	Papillary cystadenocarcinoma, NOS					C00-C06, C09-C14, C30.0, C31, C32	8450
	1	<b>EPITHELIAL TUMOURS OF HYPOPHARYNX AND LARYNX</b>	6.33	6.29	6.37	99.176	C12, C13, C32	8000-8001, 8003, 8005, 8004, 8010-8011, 8012, 8020-8022, 8030, 8033, 8035, 8031-8032, 8050-8076, 8078, 8082-8084, 8120, 8123, 8560, 8951, 8980
<b>R</b>	2	<b>Squamous cell carcinoma with variants of hypopharynx</b>	1.27	1.25	1.28	19.828	C12, C13	8004, 8020-8022, 8120, 8031-8032, 8050-8076, 8078, 8082-8084, 8123, 8560, 8980
	3	Squamous carcinoma					C12, C13	8070
	3	Lymphoepithelial carcinoma					C12, C13	8082
	3	Verrucous carcinoma					C12, C13	8051

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	3	Squamous cell carcinoma spindle cell					C12, C13	8004, 8032, 8074, 8980
	3	Giant cell carcinoma					C12, C13	8031
	3	Adenosquamous carcinoma					C12, C13	8560
	3	Squamous cell carcinoma, adenoid					C12, C13	8075
	3	Undifferentiated carcinoma					C12, C13	8020-8022
	3	Papillary squamous cell carcinoma					C12, C13	8052
	3	Basaloid squamous cell carcinoma					C12, C13	8083
R	2	<b>Squamous cell carcinoma with variants of larynx</b>	4,61	4,58	4,64	72.210	C32	8004, 8020-8022, 8031-8032, 8050-8076, 8078, 8082-8084, 8120, 8123, 8560, 8980
	3	Squamous carcinoma					C32	8070
	3	Verrucous carcinoma					C32	8051
	3	Squamous cell carcinoma spindle cell					C32	8004, 8074, 8032, 8980
	3	Papillary squamous cell carcinoma					C32	8052
	3	Adenosquamous carcinoma					C32	8560
	3	Squamous cell carcinoma, adenoid					C32	8075
	3	Undifferentiated carcinoma					C32	8020-8022
	3	Lymphoepithelial carcinoma					C32	8082
	3	Giant cell carcinoma					C32	8031
	3	Basaloid squamous cell carcinoma					C32	8083
	1	<b>EPITHELIAL TUMOURS OF OROPHARYNX</b>	3,32	3,29	3,35	52.017	C01.9, C02.4, C02.8, C05.1-C05.2, C05.8-C05.9, C09.0-C10.3, C10.8-10.9, C14.2	8000-8001, 8003, 8005, 8004, 8010-8011, 8012, 8020-8022, 8032, 8033, 8050-8076, 8078, 8082-8084, 8120, 8121, 8123, 8560, 8951, 8980
R	2	<b>Squamous cell carcinoma with variants of oropharynx</b>	3,12	3,09	3,14	48.812	C01.9, C02.4, C02.8, C05.1-C05.2, C05.8-C05.9, C09.0-C10.3, C10.8-10.9, C14.2	8004, 8020-8022, 8032, 8050-8076, 8078, 8082-8084, 8120, 8121, 8123, 8560, 8980
	3	Squamous carcinoma					C01.9, C02.4, C02.8, C05.1-C05.2, C05.8-C05.9, C09.0-C10.3, C10.8-10.9, C14.2	8070
	3	Lymphoepithelial carcinoma					C01.9, C02.4, C02.8, C05.1-C05.2, C05.8-C05.9, C09.0-C10.3, C10.8-10.9, C14.2	8082
	3	Verrucous carcinoma					C01.9, C02.4, C02.8, C05.1-C05.2, C05.8-C05.9, C09.0-C10.3, C10.8-10.9, C14.2	8051
	3	Squamous cell carcinoma spindle cell					C01.9, C02.4, C02.8, C05.1-C05.2, C05.8-C05.9, C09.0-C10.3, C10.8-10.9, C14.2	8004, 8032, 8074, 8980
	3	Papillary squamous cell carcinoma					C01.9, C02.4, C02.8, C05.1-C05.2, C05.8-C05.9, C09.0-C10.3, C10.8-10.9, C14.2	8052
	3	Squamous cell carcinoma, adenoid					C01.9, C02.4, C02.8, C05.1-C05.2, C05.8-C05.9, C09.0-C10.3, C10.8-10.9, C14.2	8075
	3	Basaloid squamous cell carcinoma					C01.9, C02.4, C02.8, C05.1-C05.2, C05.8-C05.9, C09.0-C10.3, C10.8-10.9, C14.2	8083
	3	Adenosquamous carcinoma					C01.9, C02.4, C02.8, C05.1-C05.2, C05.8-C05.9, C09.0-C10.3, C10.8-10.9, C14.2	8560
	3	Undifferentiated carcinoma					C01.9, C02.4, C02.8, C05.1-C05.2, C05.8-C05.9, C09.0-C10.3, C10.8-10.9, C14.2	8020-8022

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	1	<b>EPITHELIAL TUMOURS OF ORAL CAVITY AND LIP</b>	4,78	4,75	4,81	74.890	C00.0-C00.9 C02.0-C02.3, C02.9, C03.0-C05.0, C06.0-C06.9	8000-8001, 8003, 8004, 8010-8011, 8012, 8015, 8030, 8031, 8033, 8034, 8020-8022, 8032, 8050-8076, 8078, 8082-8084, 8123, 8560, 8951, 8980, 8981 8093, 8094, 8095, 8097, 8110,8390, 8400, 8407, 8410 <b>C00.0-C00.9:</b> 8090, 8091, 8092,
R	2	<b>Squamous cell carcinoma with variants of oral cavity</b>	3,51	3,48	3,54	54.931	C02.0-C02.3, C02.9, C03.0-C05.0, C06.0-C06.9	8004, 8020-8022, 8032, 8050-8076, 8078, 8082-8084, 8123, 8560, 8980
	3	Squamous carcinoma					C02.0-C02.3, C02.9, C03.0-C05.0, C06.0-C06.9	8070
	3	Verrucous carcinoma					C02.0-C02.3, C02.9, C03.0-C05.0, C06.0-C06.9	8051
	3	Squamous cell carcinoma spindle cell					C02.0-C02.3, C02.9, C03.0-C05.0, C06.0-C06.9	8004, 8032, 8074, 8980
	3	Adenosquamous carcinoma					C02.0-C02.3, C02.9, C03.0-C05.0, C06.0-C06.9	8560
	3	Papillary squamous cell carcinoma					C02.0-C02.3, C02.9, C03.0-C05.0, C06.0-C06.9	8052
	3	Squamous cell carcinoma, adenoid					C02.0-C02.3, C02.9, C03.0-C05.0, C06.0-C06.9	8075
	3	Basaloid squamous cell carcinoma					C02.0-C02.3, C02.9, C03.0-C05.0, C06.0-C06.9	8083
	3	Lymphoepithelial carcinoma					C02.0-C02.3, C02.9, C03.0-C05.0, C06.0-C06.9	8082
	3	Undifferentiated carcinoma					C02.0-C02.3, C02.9, C03.0-C05.0, C06.0-C06.9	8020-8022
R	2	<b>Squamous cell carcinoma with variants of lip</b>	1,02	1,00	1,04	15.984	C00.0-C00.9	8004, 8032, 8050-8076, 8078, 8082-8084, 8090, 8091, 8092, 8093, 8094, 8095, 8097, 8110,8390, 8400, 8407, 8410, 8560, 8980
	3	Squamous carcinoma					C00.0-C00.9	8070
	1	<b>EPITHELIAL TUMOURS OF OESOPHAGUS</b>	7,81	7,77	7,85	122.344	C15	8000-8001, 8003, 8004, 8010-8011, 8012,8020-8022, 8030, 8032, 8035, 8050-8076, 8078, 8082-8084, 8123, 8140-8141, 8143, 8144-8145, 8147, 8190, 8200-8201, 8210-8211, 8221, 8230-8231, 8255, 8260-8263, 8290, 8310, 8315, 8320, 8323, 8333, 8380-8384, 8401, 8430, 8440-8441, 8450, 8480-8482, 8490, 8500, 8503-8504, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8560, 8562, 8570, 8571-8576, 8980
R	2	<b>Squamous cell carcinoma with variants of oesophagus</b>	3,36	3,33	3,39	52.597	C15	8004, 8032, 8050-8076, 8078, 8082-8084, 8123, 8560, 8980
	3	Squamous carcinoma					C15	8070
	3	Adenosquamous carcinoma					C15	8560
	3	Squamous cell carcinoma spindle cell					C15	8004, 8032, 8074, 8980
	3	Verrucous carcinoma					C15	8051
	3	Papillary squamous cell carcinoma					C15	8052
	3	Basaloid squamous cell carcinoma					C15	8083
	3	Squamous cell carcinoma, adenoid					C15	8075
R	2	<b>Adenocarcinoma with variants of oesophagus</b>	3,26	3,24	3,29	51.138	C15	8140-8141, 8143, 8144-8145, 8147, 8190, 8201, 8210-8211, 8221, 8230-8231, 8255, 8260-8263, 8290, 8310, 8315, 8320, 8323, 8333, 8380-8384, 8401, 8440-8441, 8450, 8480-8482, 8490, 8500, 8503-8504, 8510, 8512, 8514, 8525, 8542, 8551, 8570, 8571-8576
R	2	<b>Salivary gland type tumours of oesophagus</b>	0,004	0,003	0,005	63	C15	8200, 8430, 8550, 8562
	3	Mucoepidermoid carcinoma					C15	8430
	3	Adenoid cystic carcinoma					C15	8200
R	2	<b>Undifferentiated carcinoma of oesophagus</b>	0,04	0,04	0,05	695	C15	8020-8022

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	1	<b>EPITHELIAL TUMOURS OF STOMACH</b>	17,43	17,36	17,49	272.978	C16	8000-8001, 8003, 8004, 8005, 8010-8011, 8012, 8020-8022, 8030, 8031, 8032, 8033, 8035, 8040, 8050-8076, 8078, 8082-8084, 8122, 8123, 8130, 8140-8145, 8147, 8190, 8200-8201, 8210-8211, 8214, 8221, 8230-8231, 8255, 8260-8263, 8290, 8310, 8315, 8320, 8323, 8333, 8380-8384, 8401, 8430, 8440-8441, 8450, 8480-8482, 8490, 8500, 8503-8504, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8560, 8562, 8570, 8571-8576, 8951, 8980
	2	<b>Adenocarcinoma with variants of stomach</b>	14,18	14,12	14,24	222.145	C16	8050, 8140-8145, 8147, 8190, 8200, 8201, 8210-8211, 8214, 8221, 8230-8231, 8255, 8260-8263, 8290, 8310, 8315, 8320, 8323, 8333, 8380-8384, 8401, 8440-8441, 8450, 8480-8482, 8490, 8500, 8503-8504, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8560, 8562, 8570, 8571-8576
	3	Adenocarcinoma NOS					C16	8140
	3	Adenocarcinoma intestinal type					C16	8144
	3	Signet ring cell carcinoma					C16	8490
	3	Carcinoma diffuse type					C16	8145
	3	Mucinous adenocarcinoma					C16	8480-8481
	3	Tubular adenocarcinoma					C16	8211
	3	Papillary adenocarcinoma, NOS					C16	8050, 8260
	3	Adenosquamous carcinoma					C16	8560
R	2	<b>Squamous cell carcinoma with variants of stomach</b>	0,12	0,11	0,12	1.807	C16	8004, 8032, 8051-8076, 8078, 8082-8084, 8980
	3	Squamous carcinoma					C16	8070
	3	Squamous cell carcinoma spindle cell					C16	8004, 8032, 8074, 8980
	3	Basaloid carcinoma					C16	8083
	3	Verrucous carcinoma					C16	8051
	3	Squamous cell carcinoma, adenoid					C16	8075
	3	Papillary squamous cell carcinoma					C16	8052
R	2	<b>Salivary gland-type tumours of stomach</b>	0,002	0,002	0,003	39	C16	8200, 8430
	3	Mucoepidermoid carcinoma					C16	8430
	3	Adenoid cystic carcinoma					C16	8200
R	2	<b>Undifferentiated carcinoma of stomach</b>	0,21	0,20	0,22	3.300	C16	8020-8022
	1	<b>EPITHELIAL TUMOURS OF SMALL INTESTINE</b>	0,77	0,76	0,79	12.132	C17	8000-8001, 8003, 8004, 8010-8011, 8012, 8020-8022, 8030, 8031, 8032, 8033-8034, 8050-8076, 8078, 8082-8084, 8140-8141, 8143, 8144, 8145, 8147, 8190, 8200, 8201, 8210-8211, 8221, 8230-8231, 8255, 8260-8263, 8290, 8310, 8315, 8320, 8323, 8333, 8380-8384, 8401, 8430, 8440-8441, 8450, 8480-8482, 8490, 8500, 8503-8504, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8560, 8562, 8570, 8571-8576, 8951, 8980
R	2	<b>Adenocarcinoma with variants of small intestine</b>	0,59	0,58	0,60	9.219	C17	8020-8022, 8140-8141, 8143, 8144-8145, 8147, 8190, 8200, 8201, 8210-8211, 8221, 8230-8231, 8255, 8260-8263, 8290, 8310, 8315, 8320, 8323, 8333, 8380-8384, 8401, 8430, 8440-8441, 8450, 8480-8482, 8490, 8500, 8503-8504, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8560, 8562, 8570, 8571-8576
	3	Adenocarcinoma NOS					C17	8140
	3	Mucinous adenocarcinoma					C17	8480-8481
	3	Signet ring cell carcinoma					C17	8490
	3	Adenosquamous carcinoma					C17	8560
	3	Medullary carcinoma, NOS					C17	8510
	3	Undifferentiated carcinoma					C17	8020-8022
R	2	<b>Squamous cell carcinoma with variants of small intestine</b>	0,01	0,01	0,01	133	C17	8004, 8032, 8050-8076, 8078, 8082-8084, 8980
	3	Squamous carcinoma					C17	8070

R=rare	Tier	Tumour	Crude incidence rate per 100,000 per year in Europe	95% confidence interval		Number of cases collected in the RARECAREnet database from 2000-2007	ICD-O-3 Topography code	ICD-O-3 Morphology code
	1	<b>EPITHELIAL TUMOURS OF COLON</b>	44,01	43,91	44,11	689.460	C18, C19	8000- 8001, 8003, 8004, 8005,8010-8011, 8012, 8020-8022, 8032, 8040,8050-8076, 8078, 8082-8084, 8140-8141, 8143, 8147, 8190, 8201, 8210-8211, 8220-8221, 8230-8231, 8255, 8260-8263, 8290, 8310, 8315, 8320, 8323, 8333, 8380-8384, 8401, 8430, 8440-8441, 8450, 8480-8482, 8490, 8500, 8503-8504, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8560, 8562, 8571-8576, 8951, 8980
	2	<b>Adenocarcinoma with variants of colon</b>	38,85	38,75	38,95	608.637	C18, C19	<b>C18, C19:</b> 8020-8022, 8140-8141, 8143, 8147, 8190, 8201, 8210-8211, 8220-8221, 8230-8231, 8255, 8260-8263, 8290, 8310, 8315, 8320, 8323, 8333, 8380-8384, 8401, 8430, 8440-8441, 8450, 8482, 8490, 8500, 8503-8504, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8560, 8562, 8571-8576, <b>C18.0, C18.2- C18.9, C19:</b> 8480-8481
	3	Adenocarcinoma NOS					C18, C19	8140
	3	Mucinous adenocarcinoma					<b>C18.0,18.2-18.9, C19 (NOT in C18.1)</b>	8480/3-8481
	3	Signet ring cell carcinoma					C18, C19	8490
	3	Adenosquamous carcinoma					C18, C19	8560
	3	Undifferentiated carcinoma					C18, C19	8020-8022
	3	Medullary carcinoma, NOS					C18, C19	8510
R	2	<b>Squamous cell carcinoma with variants of colon</b>	0,03	0,02	0,03	400	C18, C19	8004, 8032, 8050-8076, 8078, 8082-8084, 8980
R	2	<b>Fibromixoma and low grade mucinous adenocarcinoma (pseudomixoma peritonei) of the appendix</b>	0,11	0,10	0,11	1.674	C18.1	8480-8481
	1	<b>EPITHELIAL TUMOURS OF RECTUM</b>	18,09	18,03	18,16	283.461	C20	8000-8001, 8003, 8004, 8010-8011, 8012, 8020-8022, 8030-8031, 8032, 8033,8040, 8050-8076, 8078, 8082-8084, 8120, 8121, 8122, 8123-8124, 8130, 8140-8141, 8143,8144,8145, 8147, 8190, 8200, 8201, 8210-8211, 8220-8221, 8230-8231, 8255, 8260-8263, 8290, 8310, 8315, 8320, 8323, 8333, 8380-8384, 8401, 8430, 8440-8441, 8450, 8480-8482, 8490, 8500, 8503-8504, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8560, 8562,8570,8571-8576, 8951,8980
	2	<b>Adenocarcinoma with variants of rectum</b>	16,45	16,39	16,52	257.723	C20	8020-8022, 8140-8141, 8143,8144,8145, 8147, 8190, 8200,8201, 8210-8211, 8220-8221, 8230-8231, 8255, 8260-8263, 8290, 8310, 8315, 8320, 8323, 8333, 8380-8384, 8401, 8430, 8440-8441, 8450, 8480-8482, 8490, 8500, 8503-8504, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8560, 8562,8570, 8571-8576
	3	Adenocarcinoma NOS					C20	8140
	3	Mucinous adenocarcinoma					C20	8480-8481
	3	Signet ring cell carcinoma					C20	8490
	3	Adenosquamous carcinoma					C20	8560
	3	Undifferentiated carcinoma					C20	8020-8022
	3	Medullary carcinoma, NOS					C20	8510
R	2	<b>Squamous cell carcinoma with variants of rectum</b>	0,11	0,11	0,12	1.764	C20	8004, 8032, 8050-8076, 8078, 8082-8084, 8120, 8121, 8122, 8123-8124, 8130, 8980
	3	Verrucous carcinoma					C20	8051
	3	Basaloid carcinoma of rectum					C20	8123-8124
	1	<b>EPITHELIAL TUMOURS OF ANAL CANAL</b>	1,16	1,14	1,18	18.155	C21	8000-8001, 8004, 8010-8011, 8012, 8014, 8020-8022, 8032, 8033, 8050-8076, 8078, 8082-8084, 8090, 8094,8095, 8120,
R	2	<b>Squamous cell carcinoma with variants of anal canal</b>	0,81	0,80	0,82	12.691	C21	8004, 8020-8022, 8032, 8050-8076, 8078, 8082-8084, 8095, 8120,8122, 8123-8124, 8980,
	3	Verrucous carcinoma					C21	8051
	3	Undifferentiated carcinoma					C21	8020-8022
	3	Basaloid carcinoma of anal canal					C21	8123-8124
R	2	<b>Adenocarcinoma with variants of anal canal</b>	0,25	0,25	0,26	3.970	C21	8140-8141, 8143, 8144, 8147, 8190, 8200, 8201, 8210-8211, 8215, 8221, 8230-8231, 8255, 8260-8263, 8290, 8310, 8315,
	3	Mucinous adenocarcinoma					C21	8480-8481
R	2	<b>Paget's disease of anal canal</b>	0,001	0,001	0,002	21	C21	8542

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R=rare	Tier	Tumour	Crude incidence rate per 100,000 per year in Europe	95% confidence interval		Number of cases collected in the RARECAREnet database from 2000-2007	ICD-O-3 Topography code	ICD-O-3 Morphology code
	1	<b>EPITHELIAL TUMOURS OF PANCREAS</b>	12,92	12,86	12,97	202.338	C25	8000-8001, 8003, 8004, 8005, 8010-8011, 8012, 8020-8022, 8030-8031, 8032, 8033-8034, 8035, 8040, 8050-8076, 8078, 8082-8084, 8140-8141, 8143, 8144, 8145, 8147, 8190, 8200, 8201, 8210-8211, 8221, 8230-8231, 8255, 8260-8263, 8290, 8310, 8315, 8320, 8323, 8333, 8380-8384, 8401, 8430, 8440-8441, 8450, 8452-8453, 8470, 8471, 8480-8482, 8490, 8500, 8503-8504, 8510, 8512, 8514, 8521, 8523, 8525, 8542, 8550-8551, 8560, 8562, 8570, 8571-8576, 8941, 8980
	2	<b>Adenocarcinoma with variants of pancreas</b>	7,96	7,92	8,01	124.744	C25	8010, 8020-8022, 8140-8141, 8143, 8144, 8145, 8147, 8190, 8200, 8201, 8210-8211, 8221, 8230-8231, 8255, 8260-8263, 8290, 8310, 8315, 8320, 8323, 8333, 8380-8384, 8401, 8430, 8440, 8450, 8471, 8480-8482, 8490, 8500, 8503-8504, 8510, 8512, 8514, 8521, 8523, 8525, 8542, 8551, 8560, 8562, 8570, 8571-8576, 8941
	3	Adenocarcinoma NOS					C25	8140
	3	Mucinous adenocarcinoma					C25	8480-8481
	3	Undifferentiated carcinoma					C25	8020-8022
	3	Signet ring cell carcinoma					C25	8490
	3	Adenosquamous carcinoma					C25	8560
R	2	<b>Squamous cell carcinoma with variants of pancreas</b>	0,02	0,02	0,03	361	C25	8004, 8032, 8050-8076, 8078, 8082-8084, 8980
R	2	<b>Acinar cell carcinoma of pancreas</b>	0,03	0,03	0,03	449	C25	8550
R	2	<b>Mucinous cystadenocarcinoma of pancreas</b>	0,01	0,01	0,01	109	C25	8470
R	2	<b>Intraductal papillary mucinous carcinoma invasive of pancreas</b>	0,01	0,01	0,01	173	C25	8453
R	2	<b>Solid pseudopapillary carcinoma of pancreas</b>	0,003	0,002	0,004	44	C25	8452
R	2	<b>Serous cystadenocarcinoma of pancreas</b>	0,0003	#####	0,001	4	C25	8441
R	2	<b>Carcinoma with osteoclast-like giant cells of pancreas</b>	0,001	0,001	0,002	19	C25	8035
	1	<b>EPITHELIAL TUMOURS OF LIVER AND INTRAHEPATIC BILE TRACT (IBT)</b>	7,10	7,06	7,14	111.271	C22	8000-8001, 8003, 8004, 8010-8011, 8012, 8020-8022, 8030, 8031, 8032, 8033, 8034, 8050, 8051-8076, 8078, 8082-8084, 8140-8141, 8143, 8147, 8160-8162, 8170-8175, 8180, 8190, 8201, 8210-8211, 8221, 8230-8231, 8255, 8260-8263, 8290, 8310, 8315, 8320, 8323, 8333, 8380-8384, 8430, 8440-8441, 8450, 8480-8482, 8490, 8500, 8503-8504, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8560, 8562, 8571-8576, 8980, 8981,
R	2	<b>Hepatocellular carcinoma of Liver and IBT</b>	3,22	3,19	3,25	50.461	C22	8170, 8172-8175, 8180
	3	Combined hepatocellular carcinoma and cholangiocarcinoma					C22	8180
R	2	<b>Hepatocellular carcinoma, fibrolamellar</b>	0,02	0,02	0,03	387	C22	8171
R	2	<b>Cholangiocarcinoma of IBT</b>	0,97	0,95	0,99	15.201	C22	8160
R	2	<b>Adenocarcinoma with variants of liver and IBT</b>	0,41	0,40	0,42	6.457	C22	8140-8141, 8143, 8147, 8162, 8190, 8201, 8210-8211, 8221, 8230-8231, 8255, 8260-8263, 8290, 8310, 8315, 8320, 8323, 8333, 8380-8384, 8430, 8440-8441, 8450, 8480-8482, 8490, 8500, 8503-8504, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8560, 8562, 8571-8576
R	2	<b>Undifferentiated carcinoma of liver and IBT</b>	0,02	0,01	0,02	240	C22	8020-8022
R	2	<b>Squamous cell carcinoma with variants of liver and IBT</b>	0,01	0,01	0,01	147	C22	8004, 8032, 8051-8076, 8078, 8082-8084, 8980
R	2	<b>Bile duct cystadenocarcinoma of IBT</b>	0,002	0,002	0,003	38	C22	8161
	1	<b>EPITHELIAL TUMOURS OF GALLBLADDER AND EXTRAHEPATIC BILIARY TRACT (EBT)</b>	4,44	4,41	4,48	69.590	C23.9, C24	8000-8001, 8003, 8005, 8004, 8010-8011, 8012, 8014, 8015, 8020-8022, 8030, 8031, 8032, 8033, 8035, 8050-8076, 8078, 8082-8084, 8140-8141, 8143, 8144, 8145, 8147, 8160-8162, 8190, 8201, 8210-8211, 8221, 8230-8231, 8255, 8260-8263, 8290, 8310, 8315, 8320, 8323, 8333, 8380-8384, 8430, 8440-8441, 8450, 8480-8482, 8490, 8500, 8503-8504, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8560, 8562, 8571-8576, 8980



**Supplementary data**

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R=rare	Tier	Tumour	Crude incidence rate per 100,000 per year in Europe	95% confidence interval			Number of cases collected in the RARECAREnet database from 2000-2007	ICD-O-3 Topography code	ICD-O-3 Morphology code
R	2	<b>Adenocarcinoma with variants of gallbladder</b>	1,35	1,33	1,36	21.085	C23.9	8020-8022, 8050, 8140-8141, 8143, 8144, 8147, 8160-8162, 8190, 8200, 8201, 8210-8211, 8221, 8230-8231, 8251, 8255, 8260-8263, 8290, 8310, 8315, 8320, 8323, 8333, 8350, 8380-8384, 8430, 8440-8441, 8450, 8480-8482, 8490, 8500, 8503-8504, 8510, 8512, 8514, 8521, 8525, 8542, 8550-8551, 8560, 8562, 8570, 8571-8576	
	3	Cholangiocarcinoma					C23.9	8160	
	3	Papillary adenocarcinoma, NOS					C23.9	8050, 8260	
	3	Adenocarcinoma intestinal type					C23.9	8144	
	3	Mucinous adenocarcinoma					C23.9	8480-8481	
	3	Clear cell adenocarcinoma, NOS					C23.9	8310	
	3	Signet ring cell carcinoma					C23.9	8490	
	3	Adenosquamous carcinoma					C23.9	8560	
	3	Undifferentiated carcinoma					C23.9	8020-8022	
	3	Bile duct cystadenocarcinoma					C23.9	8161	
R	2	<b>Adenocarcinoma with variants of EBT</b>	1,44	1,42	1,46	22.510	C24	8020-8022, 8050, 8140-8141, 8143, 8144, 8147, 8160-8162, 8190, 8200, 8201, 8210-8211, 8221, 8230-8231, 8251, 8255, 8260-8263, 8290, 8310, 8315, 8320, 8323, 8333, 8350, 8380-8384, 8430, 8440-8441, 8450, 8480-8482, 8490, 8500, 8503-8504, 8510, 8512, 8514, 8521, 8525, 8542, 8550-8551, 8560, 8562, 8570, 8571-8576	
	3	Cholangiocarcinoma					C24	8160	
	3	Papillary adenocarcinoma, NOS					C24	8050, 8260	
	3	Adenocarcinoma intestinal type					C24	8144	
	3	Mucinous adenocarcinoma					C24	8480-8481	
	3	Clear cell adenocarcinoma, NOS					C24	8310	
	3	Signet ring cell carcinoma					C24	8490	
	3	Adenosquamous carcinoma					C24	8560	
	3	Undifferentiated carcinoma					C24	8020-8022	
	3	Bile duct cystadenocarcinoma					C24	8161	
R	2	<b>Squamous cell carcinoma of gallbladder and EBT</b>	0,03	0,03	0,03	496	C23, C24	8004, 8032, 8051-8076, 8078, 8082-8084, 8980	
	1	<b>EPITHELIAL TUMOUR OF TRACHEA</b>	0,11	0,11	0,12	1.771	C33	8000-8001, 8004, 8010-8011, 8012, 8020-8022, 8031-8032, 8033, 8050-8076, 8078, 8082-8084, 8140-8141, 8143- 8144, 8147, 8190, 8200-8201, 8210-8211, 8221, 8230-8231, 8255, 8260-8263, 8290, 8310, 8315, 8320, 8323, 8333, 8380-8384, 8430, 8440-8441, 8470, 8480-8482, 8490, 8504, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8560, 8562-8576, 8980, 8982	
R	2	<b>Squamous cell carcinoma with variants of trachea</b>	0,06	0,06	0,07	1.017	C33	8004, 8020-8022, 8031-8032, 8050-8076, 8078, 8082-8084, 8560, 8980	
	3	Squamous carcinoma					C33	8070	
	3	Papillary squamous cell carcinoma					C33	8052	
	3	Basaloid squamous cell carcinoma					C33	8083	
	3	Squamous cell carcinoma clear cell type					C33	8084	
	3	Verrucous carcinoma					C33	8051	
	3	Squamous cell carcinoma, adenoid					C33	8075	
	3	Spindle cell carcinoma					C33	8004, 8032, 8074, 8980	
	3	Adenosquamous carcinoma					C33	8560	
	3	Lymphoepithelial carcinoma					C33	8082	
	3	Giant cell carcinoma					C33	8031	
	3	Undifferentiated carcinoma					C33	8020-8022	
R	2	<b>Adenocarcinoma with variants of trachea</b>	0,01	0,01	0,01	164	C33	8140-8141, 8143, 8144, 8147, 8190, 8201, 8210-8211, 8221, 8230, 8231, 8255, 8260-8263, 8290, 8310, 8315, 8320, 8323, 8333, 8380-8384, 8440-8441, 8470, 8480-8482, 8490, 8504, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8562-8576	
	3	Adenocarcinoma NOS					C33	8140	
	3	Mucinous adenocarcinoma					C33	8480-8481	
	3	Papillary adenocarcinoma, NOS					C33	8260	
	3	Solid carcinoma, NOS					C33	8230	
	3	Clear cell adenocarcinoma, NOS					C33	8310	
	3	Acinar cell adenocarcinoma					C33	8550	



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	3	Signet ring cell carcinoma					C33	8490
	3	Mucinous cystadenocarcinoma, NOS					C33	8470
R	2	Salivary gland type tumours of trachea	0,01	0,01	0,01	175	C33	8200, 8430, 8982
	3	Adenoid cystic carcinoma					C33	8200
	3	Mucoepidermoid carcinoma					C33	8430
	3	Myoepithelial carcinoma					C33	8982
		<b>EPITHELIAL TUMOUR OF LUNG</b>	57,39	57,27	57,51	899.066	C34	8000-8001, 8002, 8003, 8004, 8005, 8010-8014, 8020-8021, 8022, 8030-8031, 8032, 8033, 8034, 8035, 8040, 8041-8046, 8050-8084, 8123, 8140-8141, 8143, 8144, 8146, 8147, 8190, 8200-8201, 8210-8211, 8221, 8230, 8231, 8246, 8250-8253, 8254, 8255, 8260-8263, 8290, 8310, 8315, 8320, 8323, 8333, 8380-8384, 8401, 8430, 8440-8441, 8460, 8470, 8480-8482, 8490, 8504, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8560, 8562-8576, 8951, 8972, 8980, 8981, 8982
	2	Squamous cell carcinoma with variants of lung	12,31	12,25	12,36	192.771	C34	8050-8070, 8073, 8075-8084,
	3	Squamous carcinoma					C34	8070
	3	Papillary squamous cell carcinoma					C34	8052
	3	Basaloid squamous cell carcinoma					C34	8083
	3	Squamous cell carcinoma small cell nonkeratinizing					C34	8073
	3	Squamous cell carcinoma clear cell type					C34	8084
	2	Adenocarcinoma with variants of lung	11,63	11,58	11,68	182.175	C34	8140-8141, 8143, 8144, 8147, 8190, 8201, 8210-8211, 8221, 8230, 8231, 8250-8254, 8255, 8260-8263, 8290, 8310, 8315, 8320, 8323, 8333, 8380-8384, 8401, 8440-8441, 8460, 8470, 8480-8482, 8490, 8504, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8562-8576
	3	Adenocarcinoma NOS					C34	8140
	3	Mucinous adenocarcinoma					C34	8480-8481
	3	Papillary adenocarcinoma, NOS					C34	8260
	3	Solid carcinoma, NOS					C34	8230
	3	Clear cell adenocarcinoma, NOS					C34	8310
	3	Acinar cell adenocarcinoma					C34	8550
	3	Signet ring cell carcinoma					C34	8490
	3	Mucinous cystadenocarcinoma, NOS					C34	8470
	3	Adenocarcinoma with mixed subtypes					C34	8255
	3	Fetal adenocarcinoma					C34	8333
	3	Bronchiolo-alveolar carcinoma,					C34	8252-8254
R	2	Adenosquamous carcinoma of lung	0,29	0,29	0,30	4.607	C34	8560
R	2	Large cell carcinoma of lung	3,81	3,78	3,84	59.714	C34	8012, 8014, 8034, 8071-72, 8123
	2	Poorly differentiated endocrine carcinoma of lung	7,91	7,86	7,95	123.888	C34	8002, 8013, 8041-8045, 8246
	3	Small cell endocrine carcinoma of the lung					C34	8002, 8041-8045
	3	Large cell endocrine carcinoma of the lung					C34	8013
R	2	Salivary gland type tumours of lung	0,06	0,05	0,06	879	C34	8200, 8430, 8982
	3	Adenoid cystic carcinoma					C34	8200
	3	Mucoepidermoid carcinoma					C34	8430
	3	Myoepithelial carcinoma					C34	8982
R	2	Sarcomatoid carcinoma of lung	0,21	0,20	0,22	3.255	C34	8004, 8022, 8030-8033, 8074, 8980, 8972
	3	Sarcomatoid carcinoma					C34	8033
	3	Pleomorphic carcinoma					C34	8022
	3	Giant cell carcinoma					C34	8031
	3	Carcinosarcoma, NOS					C34	8980
	3	Spindle cell carcinoma					C34	8004, 8032, 8074
	3	Pulmonary blastoma					C34	8972
	1	<b>EPITHELIAL TUMOURS OF THYMUS</b>	0,18	0,17	0,19	2.795	C37	8000-8001, 8003, 8010-8011, 8012, 8020-8022, 8032, 8033, 8050-8076, 8078, 8082-8084, 8123, 8140-8141, 8143-8144, 8147, 8190, 8200-8201, 8210-8211, 8221, 8230-8231, 8255, 8260-8263, 8290, 8310, 8315, 8320, 8323, 8333, 8380-8384, 8430, 8440-8441, 8480-8482, 8490, 8504, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8560, 8562-8576, 8580-8586, 8588, 8589, 8980,
R	2	Malignant thymoma	0,14	0,14	0,15	2.268	C37	8580-8586
	3	Thymoma, malignant, NOS					C37	8580

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	3	Malignant thymoma, type AB						C37	8582
	3	Malignant thymoma, type A						C37	8581
	3	Malignant thymoma, type B						C37	8583, 8584, 8585
	3	Malignant thymoma, type C						C37	8586
R	2	<b>Squamous cell carcinoma of thymus</b>	0,01	0,01	0,01	114	C37	8051-8076, 8078, 8083-8084, 8123	
R	2	<b>Undifferentiated carcinoma of thymus</b>	0,002	0,002	0,003	36	C37	8020-8022	
R	2	<b>Lymphoepithelial carcinoma of thymus</b>	0,001	#####	0,001	12	C37	8082	
R	2	<b>Adenocarcinoma with variants of thymus</b>	0,003	0,002	0,004	45	C37	8050, 8140-8141, 8143, 8144, 8147, 8190, 8200-8201, 8210-8211, 8221, 8230, 8231, 8255, 8260-8263, 8290, 8310, 8315, 8320, 8323, 8333, 8380-8384, 8430, 8440-8441, 8480-8482, 8490, 8504, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8560, 8562-8576	
	3	Adenocarcinoma NOS					C37	8140	
	3	Papillary adenocarcinoma, NOS					C37	8050, 8260	
	1	<b>EPITHELIAL TUMOURS OF BREAST</b>	67,16	67,03	67,29	1.052.063	C50	8000-8001,8003, 8004,8005, 8010-8011,8012,8020,8021, 8022, 8050, 8070, 8030-8033, 8035, 8140-8141, 8143, 8145, 8147, 8190, 8200-8201, 8210-8211, 8221, 8230- 8231, 8255, 8260-8263, 8290, 8310, 8314-8315, 8320, 8323, 8333, 8401, 8430, 8440, 8480-8481, 8490, 8500-8504, 8505, 8508, 8510, 8512-8514, 8520-8525, 8530, 8540-8541, 8543, 8550-8551, 8560, 8562, 8570-8576, 8951, 8980, 8982, 8983	
	2	<b>Invasive ductal carcinoma of breast</b>	46,56	46,45	46,66	729.345	C50	8500, 8501, 8503,8507, 8514, 8521, 8523	
	2	<b>Invasive lobular carcinoma of breast</b>	7,75	7,71	7,80	121.455	C50	8022,8520, 8524	
	3	Classic type					C50	8520	
	3	Pleomorphic carcinoma					C50	8022	
R	2	<b>Mammary Paget's disease of breast</b>	0,41	0,40	0,42	6.488	C50	8540, 8541, 8543	
R	2	<b>Special types of adenocarcinoma of breast</b>	3,06	3,04	3,09	48.012	C50	8050, 8201, 8211, 8260, 8290, 8314-8315, 8401, 8480-8481, 8502, 8510	
	3	Tubular adenocarcinoma					C50	8211	
	3	Mucinous carcinoma					C50	8480-8481	
	3	Medullary carcinoma, NOS					C50	8510	
	3	Papillary adenocarcinoma, NOS					C50	8050, 8260	
	3	Cribriform carcinoma					C50	8201	
	3	Apocrine adenocarcinoma					C50	8401	
	3	Secretory carcinoma					C50	8502	
	3	Glycogen-rich clear cell carcinoma					C50	8315	
	3	Lipid-rich carcinoma					C50	8314	
	3	Oncocytic carcinoma					C50	8290	
R	2	<b>Metaplastic carcinoma of breast</b>	0,10	0,10	0,11	1.576	C50	8070, 8560, 8571, 8572, 8575	
	3	Squamous carcinoma					C50	8070	
	3	Adenosquamous carcinoma					C50	8560	
	3	Adenocarcinoma with cartilaginous and osseous metaplasia					C50	8571	
	3	Adenocarcinoma with spindle cell metaplasia					C50	8572	
R	2	<b>Salivary gland type tumours of breast</b>	0,06	0,05	0,06	868	C50	8200, 8430, 8550, 8982	
	3	Mucoepidermoid carcinoma					C50	8430	
	3	Adenoid cystic carcinoma					C50	8200	
	3	Myoepithelial carcinoma					C50	8982	
	3	Acinar cell adenocarcinoma					C50	8550	
R	2	<b>Epithelial tumour of male breast</b>	1,06	1,04	1,08	8.098	C50	8000-8001,8003, 8004,8005, 8010-8011,8012,8020,8021, 8022, 8050, 8070, 8030-8033, 8035, 8140-8141, 8143, 8145, 8147, 8190, 8200-8201, 8210-8211, 8221, 8230- 8231, 8255, 8260-8263, 8290, 8310, 8314-8315, 8320, 8323, 8333, 8401, 8430, 8440, 8480-8481, 8490, 8500-8504, 8505, 8508, 8510, 8512-8514, 8520-8525, 8530, 8540-8541, 8543, 8550-8551, 8560, 8562, 8570-8576, 8951, 8980, 8982, 8983	

**Supplementary data**

**I tumori rari in Italia / The burden of rare cancer in Italy**  
*Epidemiol Prev* 2016; 40(1 Suppl 2): 1-120

R=rare	Tier	Tumour	Crude incidence rate per 100,000 per year in Europe	95% confidence interval		Number of cases collected in the RARECAREnet database from 2000-2007	ICD-O-3 Topography code	ICD-O-3 Morphology code
	1	<b>EPITHELIAL TUMOURS OF CORPUS UTERI</b>	11,35	11,30	11,40	177.783	C54	8000, 8001,8003-8005, 8010, 8011, 8012, 8014, 8015, 8020-8022, 8030-8033, 8050-8052, 8070-8078, 8082-8084, 8120, 8122, 8123, 8130, 8313, 8140-8141, 8143, 8144, 8147, 8190, 8200, 8201, 8210-8211, 8221, 8230, 8231, 8255, 8260-8263, 8290, 8310, 8315, 8320, 8323, 8333, 8380-8384, 8401, 8430, 8460, 8440-8441, 8470-8471, 8480-8482, 8490, 8503, 8504, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8560, 8562-8576, 8950-8951, 8980, 8981, 9000, 9110
	2	<b>Adenocarcinoma with variants of corpus uteri</b>	9,93	9,88	9,98	155.550	C54	8050-8051,8120, 8122, 8123, 8130, 8140-8141, 8143, 8144, 8147, 8190, 8201, 8210-8211, 8221, 8230, 8231, 8255, 8260-8263, 8290, 8313, 8315, 8320, 8323, 8333, 8380-8384, 8401, 8430, 8440, 8460, 8470, 8471, 8480-8482, 8490,8503, 8504, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8562-8576, 9000, 9110
	3	Adenocarcinoma, NOS					C54	8140
	3	Endometrioid adenocarcinoma, NOS					C54	8380
	3	Adenosquamous carcinoma					C54	8560
	3	Mucinous adenocarcinoma					C54	8480-8481
	3	Mixed cell adenocarcinoma					C54	8323
	3	Endometrioid adenocarcinoma secretory variant					C54	8382
	3	Villous adenocarcinoma					C54	8262
	3	Brenner tumor, malignant						9000
	3	Transitional cell carcinoma						8120
	3	Transitional cell ca spindle cell						8122
	3	Basaloid carcinoma						8123
	3	Endometrioid adenocarcinoma ciliated cell variant					C54	8383
R	2	<b>Squamous cell carcinoma with variants of corpus uteri</b>	0,06	0,06	0,07	1.003	C54	8052, 8070-8078, 8082-8084
R	2	<b>Adenoid cystic carcinoma of corpus uteri</b>	0,0003	#####	#####	5	C54	8200
R	2	<b>Clear cell adenocarcinoma, NOS</b>	0,16	0,16	0,17	2.527	C54	8310
R	2	<b>Serous (papillary) carcinoma</b>	0,08	0,07	0,08	1.227	C54	8441
R	2	<b>Mullerian mixed tumour</b>	0,40	0,39	0,41	6.276	C54	8950, 8951,8980, 8981
	2	<b>Small cell carcinoma ≠ NET</b>						new morphology. ICD-O code not available
	1	<b>EPITHELIAL TUMOURS OF CERVIX UTERI</b>	6,28	6,24	6,32	98.321	C53	8000, 8001, 8003-8005, 8010, 8011, 8012, 8015, 8020-8022, 8030-8031, 8032, 8033, 8050-8084, 8098, 8120, 8123,8130, 8140-8141, 8143, 8144, 8147, 8190, 8200-8201, 8210-8211, 8221, 8230, 8231, 8255, 8260-8263, 8290, 8310, 8315, 8320, 8323, 8333, 8380-8384,8410, 8430, 8460, 8440-8441, 8480-8482, 8490, 8504, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8560, 8562-8576, 8950-8951, 8980, 9110
R	2	<b>Squamous cell carcinoma with variants of cervix uteri</b>	4,73	4,70	4,76	74.105	C53	8015, 8120, 8123, 8050-8084, 8130
	3	Squamous carcinoma					C53	8070
	3	Squamous cell carcinoma nonkeratinizing, NOS					C53	8072
	3	Squamous cell carcinoma keratinizing, NOS					C53	8071
	3	Papillary squamous cell carcinoma					C53	8052
	3	Papillary carcinoma, NOS					C53	8050
	3	Verrucous/Warty carcinoma					C53	8051
	3	Basaloid carcinoma					C53	8123
	3	Squamous cell carcinoma spindle cell					C53	8074
	3	Lymphoepithelial carcinoma					C53	8082
	3	Transitional cell carcinoma, NOS					C53	8120
	3	Glassy cell carcinoma					C53	8015
R	2	<b>Adenocarcinoma with variants of cervix uteri</b>	0,91	0,89	0,92	14.252	C53	8098, 8140-8141, 8143, 8144, 8147, 8190, 8200-8201, 8210-8211, 8221, 8230, 8231, 8255, 8260-8263, 8290, 8310, 8315, 8320, 8323, 8333, 8380-8384, 8410, 8430, 8440-8441, 8460, 8480-8482, 8490, 8504, 8510, 8512, 8514, 8525, 8542, 8550-8551,8562-8576, 9110
	3	Adenocarcinoma, NOS					C53	8140

R=rare	Tier	Tumour	Crude incidence rate per 100,000 per year in Europe	95% confidence interval		Number of cases collected in the RARECAREnet database from 2000-2007	ICD-O-3 Topography code	ICD-O-3 Morphology code
	3	Adenocarcinoma with squamous metaplasia					C53	8570
	3	Mucinous adenocarcinoma					C53	8480-8481
	3	Clear cell adenocarcinoma, NOS					C53	8310
	3	Endometrioid adenocarcinoma, NOS					C53	8380
	3	Serous cystadenocarcinoma, NOS					C53	8441
	3	Signet ring cell carcinoma					C53	8490
	3	Mesonephroma malignant					C53	9110
	3	Villous adenocarcinoma					C53	8262
	3	Mucinous adenocarcinoma, endocervical type					C53	8482
	3	Adenocarcinoma intestinal type					C53	8144
	3	Mixed cell adenocarcinoma					C53	8323
R	2	<b>Undifferentiated carcinoma of cervix uteri</b>	0,03	0,03	0,03	480	C53	8020-8022
R	2	<b>Mullerian mixed tumour of cervix uteri</b>	0,02	0,01	0,02	257	C53	8950, 8951, 8980
	2	<b>Small cell carcinoma ≠ NET</b>						new morphology. ICD-O code not available
	1	<b>EPITHELIAL TUMOURS OF OVARY AND FALLOPIAN TUBE</b>	9,38	9,33	9,43	146.908	C56.9, C57.0-C57.3	<b>C56.9, C57.0-C57.3:</b> 8000, 8001, 8003-8005, 8010, 8011, 8012, 8020-8022, 8030-8034, 8040, 8050, 8051-8052, 8070-8076, 8083-8084, 8120, 8122, 8123, 8130, 8140-8141, 8143, 8146, 8147, 8190, 8200-8201, 8210-8211, 8221, 8230, 8231, 8255, 8260-8263, 8290, 8310, 8313, 8315, 8320, 8323, 8330, 8333, 8380-8384, 8401, 8430, 8440-8463, 8470-8490, 8500, 8504, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8560, 8562-8576, 8950-8951, 8980, 9000, 9014-9015, 9110 <b>C48:</b> 8050, 8260, 8441, 8450, 8460, 8461
R	2	<b>Adenocarcinoma with variants of ovary</b>	5,95	5,92	5,99	93.263	C56.9	8050, 8120, 8122, 8123, 8130, 8140-8141, 8143, 8146, 8147, 8190, 8200-8201, 8210-8211, 8221, 8230, 8231, 8255, 8260-8263, 8290, 8313, 8315, 8320, 8323, 8330, 8333, 8380-8384, 8401, 8430, 8440-8463, 8504, 8510, 8512, 8514, 8525, 8542, 8500, 8550-8551, 8562-8576, 9000, 9014-9015, 9110
	3	Serous (papillary) cystadenocarcinoma					C56.9	8441, 8460, 8461
	3	Adenocarcinoma NOS					C56.9	8140
	3	Endometrioid adenocarcinoma, NOS					C56.9	8380
	3	Papillary adenocarcinoma, NOS					C56.9	8050, 8260, 8450
	3	Endometrioid adenofibroma malignant					C56.9	8381
	3	Clear cell adenocarcinofibroma					C56.9	8313
	3	Serous adenocarcinofibroma					C56.9	9014
	3	Mucinous adenocarcinofibroma					C56.9	9015
	3	Brenner tumor, malignant					C56.9	9000
	3	Transitional cell carcinoma					C56.9	8120
	3	Transitional cell carcinoma spindle cell					C56.9	8122
	3	Basaloid carcinoma					C56.9	8123
R	2	<b>Mucinous adenocarcinoma of ovary</b>	0,77	0,76	0,78	12.066	C56.9	8470- 8490
R	2	<b>Clear cell adenocarcinoma of ovary</b>	0,30	0,29	0,31	4.753	C56.9	8310
R	2	<b>Primary peritoneal serous/papillary carcinoma</b>	0,08	0,08	0,09	1.280	C48	8050, 8260, 8441, 8450, 8460, 8461
R	2	<b>Mullerian mixed tumour of ovary</b>	0,14	0,14	0,15	2.255	C56.9	8950, 8951, 8980
	2	<b>Small cell ≠ NET</b>						new morphology. ICD-O code not available
R	2	<b>Adenocarcinoma with variants of fallopian tube</b>	0,17	0,16	0,18	2.683	C57.0-C57.3	8050, 8140-8141, 8143, 8147, 8190, 8200-8201, 8210-8211, 8221, 8230, 8231, 8255, 8260-8263, 8290, 8310, 8315, 8320, 8323, 8333, 8380-8384, 8430, 8440-8441, 8460, 8480-8482, 8490, 8504, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8560, 8562-8576, 9014-9015, 9110
	3	Adenocarcinoma NOS					C57.0-C57.3	8140
	3	Papillary serous cystadenocarcinoma					C57.0-C57.3	8460
	3	Serous cystadenocarcinoma, NOS					C57.0-C57.3	8441
	3	Papillary adenocarcinoma, NOS					C57.0-C57.3	8050, 8260

**Supplementary data**

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*Epidemiol Prev* 2016; 40(1 Suppl 2): 1-120

R=rare	Tier	Tumour	Crude incidence rate per 100,000 per year in Europe	95% confidence interval		Number of cases collected in the RARECAREnet database from 2000-2007	ICD-O-3 Topography code	ICD-O-3 Morphology code
	1	<b>NON EPITHELIAL TUMOURS OF OVARY</b>	0,25	0,25	0,26	3.977	C56	8590,8600, 8620, 8630-8631, 8634, 8640, 8650, 8670, 9060-9065, 9070-9072, 9073,9080-9085, 9090, 9091, 9100-9102, 9391, 9440, 9473, 9501, 8621, 8622, 8623,
R	2	<b>Sex cord tumours of ovary</b>	0,13	0,12	0,13	2.006	C56	8590, 8600, 8620, 8621,8622, 8623, 8630-8631, 8634, 8640, 8650, 8670,
	3	Granulosa cell tumour malignant					C56	8620/3
	3	Granulosa cell tumour malignant, adult type					C56	8620/1 this is included in the list however it will not be included in the analyses because border line
	3	Granulosa cell tumour malignant, juvenile type					C56	8622/1 this is included in the list however it will not be included in the analyses because border line
	3	Sertoli-Leydig cell tumour poorly differentiated					C56	8631
	3	Steroid cell tumour, malignant					C56	8670
R	2	<b>Malignant/Immature teratomas of ovary</b>	0,05	0,05	0,06	833	C56	9080-9084, 9090, 9102, 9391, 9440, 9473, 9501, 9091
	3	Teratoma, malignant, NOS					C56	9080
	3	Struma ovarii, malignant					C56	9090
	3	Primitive neuroectodermal tumour, NOS					C56	9473
	3	Ependymoma					C56	9391
	3	Medulloepithelioma, NOS					C56	9501
	3	Glioblastoma					C56	9440
R	2	<b>Germ cell tumour of ovary</b>	0,07	0,07	0,08	1.138	C56	9060-9065, 9070-9072, 9073, 9085, 9100-9101
	3	Dysgerminoma/Seminoma					C56	9060-9065
	3	Yolk sac tumour					C56	9071
	3	Mixed germ cell tumour					C56	9085
	3	Embryonal adenocarcinoma					C56	9070
	3	Choriocarcinoma, NOS					C56	9100
	3	Polyembryoma					C56	9072
	1	<b>EPITHELIAL TUMOURS OF VULVA AND VAGINA</b>	1,97	1,95	2,00	30.938	C51, C52	8000, 8001, 8003-8004, 8010, 8011, 8012, 8020-8022,8030, 8032-8033, 8050-8084,8090-8095, 8097-8098,8102,8120, 8123, 8140-8141, 8143, 8144,8147, 8190-8211, 8230-8231, 8255-8263, 8290, 8310, 8315, 8320, 8323, 8333, 8380-8384, 8390-8420, 8430,8440,8460, 8480-8490, 8500, 8503-8504, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8560, 8562-8576, 8950,8951, 8980,8982, 9110
R	2	<b>Squamous cell carcinoma with variants of vulva and vagina</b>	1,69	1,67	1,71	26.422	C51, C52	8050-8084, 8090-8095, 8097,8120, 8123, 8130
R	2	<b>Adenocarcinoma with variants of vulva and vagina</b>	0,07	0,07	0,08	1.120	C51, C52	8098, 8102, 8140-8141, 8143, 8147, 8190-8211, 8230-8231, 8255-8263, 8290, 8310, 8315, 8320, 8323, 8333, 8380-8384, 8390-8420, 8430, 8440, 8446, 8480-8490, 8500, 8503-8504, 8510, 8512, 8514, 8525, 8550-8551, 8562-8576, 8982, 9110
R	2	<b>Paget's disease of vulva and vagina</b>	0,05	0,04	0,05	746	C51, C52	8542
R	2	<b>Undifferentiated carcinoma of vulva and vagina</b>	0,01	0,00	0,01	85	C51, C52	8020-8022
	1	<b>TROPHOBLASTIC TUMOUR OF PLACENTA</b>	0,02	0,02	0,03	363	C58.9	9100, 9104, 9105
R	2	<b>Choriocarcinoma of placenta</b>	0,02	0,02	0,02	352	C58.9	9100
	1	<b>EPITHELIAL TUMOURS OF PROSTATE</b>	55,66	55,55	55,78	872.015	C61, C63.7	8000-8001, 8003, 8005, 8010-8011, 8012, 8015, 8020-8022,8030, 8031, 8032, 8033, 8034, 8040,8050, 8051-8052, 8070-8072,8073, 8074, 8075-8076, 8082, 8084, 8120, 8121, 8122-8123, 8130-8131, 8140-8141, 8143, 8145, 8147, 8190, 8200-8201, 8210-8211, 8230-8231, 8251, 8255, 8260, 8261, 8262-8263, 8290, 8310, 8320, 8323, 8333, 8380-8384,8430, 8440, 8441, 8450, 8480-8490, 8500, 8503-8504, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8560, 8562-8576, 8941, 8951, 8980
	2	<b>Adenocarcinoma with variants of prostate</b>	48,86	48,75	48,97	765.405	C61	8030, 8032, 8033,8074, 8082, 8140-8141, 8143,8145, 8190, 8210-8211, 8231, 8251, 8255, 8261, 8262-8263, 8290, 8310, 8320, 8323, 8333, 8380-8384,8430, 8440, 8441, 8450,8480-8490, 8504, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8562-8576, 8941, 8980
	3	Adenocarcinoma NOS					C61	8140
	3	Acinar cell adenocarcinoma					C61	8550
	3	Mucinous adenocarcinoma					C61	8480-8481

**Supplementary data**

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R=rare	Tier	Tumour	Crude incidence rate per 100,000 per year in Europe	95% confidence interval		Number of cases collected in the RARECAREnet database from 2000-2007	ICD-O-3 Topography code	ICD-O-3 Morphology code
	3	Signet ring cell carcinoma					C61	8490
	3	Adenocarcinoma with neuroendocrine differentiation					C61	8574
	3	Oxyphilic adenocarcinoma					C61	8290
	3	Spindle cell adenocarcinoma					C61	8032, 8074, 8572
	3	Lymphoepithelial carcinoma					C61	8082
R	2	<b>Squamous cell carcinoma with variants of prostate</b>	0,02	0,02	0,02	291	C61	8051, 8052, 8070-8072, 8073, 8075, 8076, 8084, 8121, 8123 ,8560,
	3	Squamous carcinoma					C61	8070
	3	Adenosquamous carcinoma					C61	8560
	3	Basaloid carcinoma					C61	8123
R	2	<b>Infiltrating duct carcinoma of prostate</b>	0,51	0,50	0,53	8.064	C61	8050, 8201, 8230, 8260, 8500, 8503
	3	Cribriform carcinoma					C61	8201
	3	Solid carcinoma, NOS					C61	8230
	3	Papillary adenocarcinoma, NOS					C61	8050, 8260
R	2	<b>Transitional cell carcinoma of prostate</b>	0,06	0,06	0,07	960	C61	8120, 8122, 8130-8131
R	2	<b>Basal cell adenocarcinoma of prostate</b>	0,01	0,01	0,01	122	C61	8147, 8200
	3	Adenoid cystic carcinoma					C61	8200
	3	Basal cell adenocarcinoma					C61	8147
	1	<b>TESTICULAR AND PARATESTICULAR CANCERS</b>	3,29	3,27	3,32	51.609	C62, C63.0, C63.1	<b>C63.0, C63.1:</b> 8000, 8001, 8004, 8010,8011, 8012, 8120, 8123, 8140-8141, 8147, 8190, 8200, 8210-8211, 8221, 8230, 8231, 8255, 8260, 8261-8263, 8290, 8310, 8320, 8323, 8333, 8380-8384, 8401, 8430, 8440-8441, 8460-8461, 8470, 8480-8490, 8504, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8560, 8562-8576, <b>C62:</b> 8000, 8001, 8590-8592, 8630-8640, 8650, 9060-9085, 9100-9102
R	2	<b>Paratesticular adenocarcinoma with variants</b>	0,001	0,001	0,002	22	C63.0, C63.1	8120, 8123, 8140-8141, 8147, 8190, 8200, 8210-8211, 8221, 8230, 8231, 8255, 8260, 8261-8263, 8290, 8310, 8320, 8323, 8333, 8380-8384, 8401, 8430, 8440-8441, 8460, 8470, 8480-8490, 8504, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8560, 8562-8576
	3	Endometrioid adenocarcinoma, NOS					C63.0, C63.1	8380
	3	Clear cell adenocarcinoma, NOS					C63.0, C63.1	8310
	3	Serous cystadenocarcinoma, NOS					C63.0, C63.1	8441
	3	Mucinous cystadenocarcinoma, NOS					C63.0, C63.1	8470
	3	Collecting duct and rete testis carcinoma					C63.0, C63.1	malignancy without ICD-O morphology code
	3	Transitional cell carcinoma, NOS					C63.0, C63.1	8120
R	2	<b>Non seminomatous testicular cancer</b>	1,27	1,25	1,28	19.835	C62	9080-9083, 9085, 9100-9102, 9065, 9070-9072
	3	Mixed germ cell tumour					C62	9085
	3	Teratocarcinoma					C62	9081
	3	Choriocarcinoma combined with other germ cell elements					C62	9101
	3	Embryonal adenocarcinoma, NOS					C62	9070
	3	Yolk sac tumour					C62	9071
	3	Choriocarcinoma, NOS					C62	9100
R	2	<b>Seminomatous testicular cancer</b>	1,82	1,80	1,84	28.516	C62	9060-9062
	3	Seminoma, NOS					C62	9061
R	2	<b>Spermatocytic seminoma</b>	0,03	0,03	0,03	502	C62	9063
R	2	<b>Teratoma with malignant transformation</b>	0,001	0,001	0,002	20	C62	9084
R	2	<b>Testicular sex cord cancer</b>	0,02	0,02	0,02	340	C62	8630-8640, 8650, 8590-8592
	3	Leydig cell tumour malignant					C62	8650
	3	Sertoli cell carcinoma					C62	8640
	3	Malignant sex cord/gonadal stromal tumours					C62	8590-8592 borderline malignancy

**Supplementary data**

**I tumori rari in Italia / The burden of rare cancer in Italy**  
*Epidemiol Prev* 2016; 40(1 Suppl 2): 1-120

R=rare	Tier	Tumour	Crude incidence rate per 100,000 per year in Europe	95% confidence interval		Number of cases collected in the RARECAREnet database from 2000-2007	ICD-O-3 Topography code	ICD-O-3 Morphology code
	1	<b>EPITHELIAL TUMOURS OF PENIS</b>	0,66	0,65	0,67	10.368	C60	8000-8001, 8003, 8010-8011, 8012, 8020-8022, 8031, 8032, 8033, 8050-8084, 8090, 8094, 8095, 8123, 8140, 8141, 8147, 8190, 8200-8201, 8210-8211, 8221, 8230-8231, 8255-8263, 8310, 8320, 8323, 8390, 8402, 8410, 8430-8440, 8480-8490, 8504, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8560-8576, 8940, 8980, 8982
R	2	<b>Squamous cell carcinoma with variants of penis</b>	0,62	0,60	0,63	9.646	C60	8020-8022, 8050-8084, 8090, 8094, 8123, 8320, 8410, 8980
	3	Squamous carcinoma					C60	8070
	3	Adenosquamous carcinoma					C60	8560
	3	Verrocous carcinoma					C60	8051
	3	Squamous cell carcinoma, sarcomatoid					C60	8074
	3	Basaloid carcinoma					C60	8123
R	2	<b>Adenocarcinoma with variants of penis</b>	0,01	0,005	0,01	88	C60	8140-8141, 8147, 8190, 8200-8201, 8210-8211, 8221, 8230-8231, 8255-8263, 8310, 8323, 8402, 8440, 8480-8490, 8504, 8510, 8512, 8514, 8525, 8542, 8550-8576, 8940, 8982
	3	Extramammary Paget's disease					C60	8542
	3	Sebaceous adenocarcinoma					C60	8410
	3	Papillary adenocarcinoma, NOS					C60	8260
	3	Mixed tumour malignant, NOS					C60	8940
	3	Clear cell adenocarcinoma, NOS					C60	8310
	3	Basal cell adenocarcinoma					C60	8147
	1	<b>EPITHELIAL TUMOURS OF KIDNEY</b>	12,71	12,65	12,76	199.062	C64	8000-8001, 8003-8005, 8010-8011, 8012, 8014, 8020, 8022, 8030-8035, 8050-8084, 8123, 8140-8141, 8143, 8147, 8190, 8200, 8201, 8210-8211, 8221, 8230-8231, 8251, 8255-8263, 8270, 8290, 8300, 8310, 8311, 8312, 8315-8320, 8323, 8330, 8333, 8380-8384, 8401, 8430, 8440, 8450, 8480-8490, 8500, 8503-8504, 8510-8512, 8514, 8525, 8542, 8550-8551, 8560, 8562-8576, 8980, 9110,
	2	<b>Renal cell carcinoma with variants</b>	10,08	10,03	10,13	157.886	C64	8005, 8050, 8140-8141, 8143, 8147, 8190, 8200, 8201, 8211, 8221, 8230-8231, 8251, 8255-8263, 8270, 8290, 8300, 8310, 8311, 8312, 8315-8320, 8323, 8330, 8333, 8401, 8430, 8440, 8342, 8450, 8480-8490, 8500, 8503-8504, 8510-8512, 8514, 8525, 8542, 8550-8551, 8562, 9110
	3	Adenocarcinoma NOS					C64	8140
	3	Renal cell adenocarcinoma					C64	8312
	3	Clear cell adenocarcinoma, NOS					C64	8310
	3	Papillary adenocarcinoma, NOS					C64	8050, 8260
	3	Renal cell carcinoma chromophobe type					C64	8317
	3	Collecting duct carcinoma					C64	8319
	3	Xp translocation carcinoma					C64	malignancy without ICD-O morphology code
	3	Carcinoma associated with neuroblastoma					C64	malignancy without ICD-O morphology code
	3	Renal medullary carcinoma					C64	8510
	3	Mucinous tubular and spindle cell carcinoma					C64	malignancy without ICD-O morphology code
R	2	<b>Squamous cell carcinoma spindle cell type of kidney</b>	0,01	0,01	0,01	190	C64	8004, 8032, 8074, 8980
R	2	<b>Squamous cell carcinoma with variants of kidney</b>	0,03	0,03	0,04	533	C64	8020-8022, 8051-8073, 8075-8084, 8560
	1	<b>EPITHELIAL TUMOURS OF PELVIS AND URETER</b>	1,58	1,57	1,60	24.826	C65.9,C66.9	8000-8001, 8003-8005, 8010-8011, 8012, 8014, 8020, 8021, 8022, 8030, 8031, 8032, 8033, 8050-8078, 8082-8084, 8120, 8121, 8122, 8123, 8130-8131, 8140-8141, 8143, 8145, 8147, 8190, 8200, 8201, 8210-8211, 8221, 8230-8231, 8251, 8255-8263, 8270, 8280, 8290, 8300, 8310, 8320, 8323, 8330, 8342, 8430-8440, 8480-8490, 8500, 8504, 8510, 8512, 8514, 8525, 8542, 8550, 8560-8576, 8980, 9110
R	2	<b>Transitional cell carcinoma of pelvis and ureter</b>	1,41	1,39	1,43	22.099	C65.9,C66.9	8020-8022, 8031, 8050 8082, 8120, 8121, 8122, 8130-8131
	3	Undifferentiated carcinoma					C65.9,C66.9	8020-8022
	3	Transitional cell carcinoma spindle cell					C65.9,C66.9	8122
	3	Lymphoepithelioma-like carcinoma					C65.9,C66.9	8082
	3	Transitional cell carcinoma, giant cell					C65.9,C66.9	8031



**Supplementary data**

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R=rare	Tier	Tumour	Crude incidence rate per 100,000 per year in Europe	95% confidence interval		Number of cases collected in the RARECAREnet database from 2000-2007	ICD-O-3 Topography code	ICD-O-3 Morphology code
	3	Transitional cell carcinoma, micropapillary					C65.9,C66.9	8131
R	2	<b>Squamous cell carcinoma with variants of pelvis and ureter</b>	0,02	0,02	0,03	372	C65.9,C66.9	8004, 8051-8076; 8123
	3	Basaloid carcinoma					C65.9,C66.9	8123
	3	Verrucous carcinoma					C65.9,C66.9	8051
R	2	<b>Adenocarcinoma with variants of pelvis and ureter</b>	0,02	0,02	0,02	326	C65.9,C66.9	8005, 8140-8141, 8143, 8147, 8190, 8200, 8201, 8211, 8221, 8230-8231, 8251, 8255, 8260-8263, 8270, 8280, 8290, 8300, 8310, 8320, 8323, 8330, 8342, 8430, 8480-8490,8500, 8510, 8512, 8514, 8525, 8542, 8560, 8570, 9110
	3	Mucinous adenocarcinoma					C65.9,C66.9	8480-8481
	3	Clear cell adenocarcinoma, NOS					C65.9,C66.9	8310
	3	Signet ring cell carcinoma					C65.9,C66.9	8490
	3	Adenoid cystic carcinoma					C65.9,C66.9	8200
	1	<b>EPITHELIAL TUMOURS OF URETHRA</b>	0,13	0,13	0,14	2.077	C68.0	8000-8001, 8010-8011, 8012, 8020, 8021, 8022, 8031, 8033, 8050-8078, 8082-8084, 8094, 8120, 8121, 8122, 8123, 8130-8131, 8140-8141, 8143, 8147, 8190, 8200, 8201, 8210-8211, 8221, 8230-8231, 8255-8263, 8290, 8310, 8320, 8323,8440, 8480-8490, 8504, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8560-8576, 8980, 9110,
R	2	<b>Transitional cell carcinoma of urethra</b>	0,09	0,08	0,09	1.390	C68.0	8020-8022, 8031, 8050, 8082, 8120, 8121, 8122, 8130-8131
	3	Undifferentiated carcinoma					C68.0	8020-8022
	3	Transitional cell carcinoma spindle cell					C68.0	8122
	3	Lymphoepithelioma-like carcinoma					C68.0	8082
	3	Transitional cell carcinoma, giant cell					C68.0	8031
	3	Transitional cell carcinoma, micropapillary					C68.0	8131
R	2	<b>Squamous cell carcinoma with variants of urethra</b>	0,02	0,02	0,02	329	C68.0	8051-8076; 8094, 8123
	3	Basaloid carcinoma					C68.0	8123
	3	Verrucous carcinoma					C68.0	8051
R	2	<b>Adenocarcinoma with variants of urethra</b>	0,01	0,01	0,01	190	C68.0	8140-8141, 8143, 8147, 8190, 8200-8201, 8210, 8211, 8221, 8230-8231, 8255, 8260-8263, 8290, 8310, 8320, 8323, 8430, 8480-8490, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8560, 8570, 9110
	3	Mucinous adenocarcinoma					C68.0	8480-8481
	3	Clear cell adenocarcinoma, NOS					C68.0	8310
	3	Signet ring cell carcinoma					C68.0	8490
	3	Adenoid cystic carcinoma					C68.0	8200
	1	<b>EPITHELIAL TUMOURS OF BLADDER</b>	18,20	18,13	18,27	285.122	C67	8000-8001, 8004, 8010-8011, 8012, 8020, 8021,8022,8030, 8031, 8032, 8033, 8035, 8050-8078, 8083-8084, 8120,8121, 8122, 8123, 8130-8131, 8140-8141, 8143,8145, 8147, 8190, 8200, 8201, 8210-8211, 8221, 8230-8231, 8255-8263, 8290, 8310, 8320, 8323, 8380, 8430-8440, 8460, 8480-8490, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8560-8575, 8951, 8980, 9110
	2	<b>Transitional cell carcinoma of bladder</b>	15,68	15,62	15,74	245.681	C67	8020-8022, 8030, 8031, 8032, 8033, 8050, 8082, 8120, 8121, 8122, 8130-8131, 8145
	3	Undifferentiated carcinoma					C67	8020-8022
	3	Transitional cell carcinoma spindle cell					C67	8122
	3	Transitional cell carcinoma with squamous differentiation					C67	malignancy without ICD-O morphology code
	3	Lymphoepithelial carcinoma					C67	8082
	3	Giant cell carcinoma					C67	8031
	3	Transitional cell carcinoma micropapillary					C67	8131
R	2	<b>Squamous cell carcinoma with variants of bladder</b>	0,36	0,35	0,36	5.566	C67	8051-8076; 8078, 8083-8084, 8123
	3	Basaloid carcinoma					C67	8123
	3	Verrucous carcinoma					C67	8051
R	2	<b>Adenocarcinoma with variants of bladder</b>	0,30	0,29	0,31	4.653	C67	8140-8141, 8143, 8147, 8190, 8201, 8210, 8211, 8221, 8230-8231, 8255, 8260-8263, 8290, 8310, 8320, 8323, 8380, 8440,8460, 8480-8490, 8510, 8512, 8514, 8525, 8542, 8560, 8570, 9110
	3	Mucinous adenocarcinoma					C67	8480-8481
	3	Clear cell adenocarcinoma, NOS					C67	8310

R=rare	Tier	Tumour	Crude incidence rate per 100,000 per year in Europe	95% confidence interval		Number of cases collected in the RARECAREnet database from 2000-2007	ICD-O-3 Topography code	ICD-O-3 Morphology code
	3	Signet ring cell carcinoma					C67	8490
R	2	Salivary gland type tumours of bladder	0,000	0,000	0,001	7	C67	8200, 8430
	3	Adenoid cystic carcinoma					C67	8200
	3	Mucoepidermoid carcinoma					C67	8430
	1	<b>EPITHELIAL TUMOURS OF EYE AND ADNEXA</b>	0,04	0,04	0,05	679	C69.0, C69.5	8000-8001, 8010-8011, 8020, 8032, 8050-8084, 8090, 8120-8121, 8123, 8140-8141, 8147, 8190, 8200, 8221, 8230-8231, 8255-8263, 8290, 8310, 8320, 8323, 8410, 8430-8440, 8480-8490, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8560-8576, 8941
R	2	Squamous cell carcinoma with variants of eye and adnexa	0,03	0,02	0,03	421	C69.0, C69.5	8020, 8050-8082, 8083-8084, 8090
	3	Squamous carcinoma					C69.0, C69.5	8070
	3	Basal cell carcinoma					C69.0, C69.5	8090
R	2	Adenocarcinoma with variants of eye and adnexa	0,01	0,01	0,01	134	C69.0, C69.5	8140, 8147, 8190, 8200, 8211, 8221, 8230-8231, 8255-8263, 8290, 8310, 8320, 8323, 8410, 8430-8440, 8480-8490, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8560-8576, 8941
	3	Adenocarcinoma NOS					C69.0, C69.5	8140
	3	Adenoid cystic carcinoma					C69.0, C69.5	8200
	3	Mucoepidermoid carcinoma					C69.0, C69.5	8430
	1	<b>EPITHELIAL TUMOURS OF MIDDLE EAR</b>	0,03	0,03	0,04	524	C30.1	8000-8001, 8003, 8010-8011, 8012, 8020, 8022, 8032, 8050-8084, 8090, 8094, 8120-8121, 8123, 8130, 8140-8141, 8147, 8190, 8200-8201, 8211, 8221, 8230-8231, 8255-8263, 8290, 8310, 8320, 8323, 8410, 8430-8440, 8480-8490, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8560-8576
R	2	Squamous cell carcinoma with variants middle ear	0,02	0,02	0,03	377	C30.1	8050-8084, 8090, 8094, 8120, 8121, 8123, 8130
	3	Squamous carcinoma					C30.1	8070
R	2	Adenocarcinoma with variants of middle ear	0,003	0,002	0,004	50	C30.1	8140, 8147, 8190, 8200, 8211, 8221, 8230-8231, 8255-8263, 8290, 8310, 8320, 8323, 8410, 8430-8440, 8480-8490, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8560-8576
	3	Adenocarcinoma NOS					C30.1	8140
	3	Adenoid cystic carcinoma					C30.1	8200
	1	<b>MALIGNANT MESOTHELIOMA</b>	2,14	2,12	2,16	33.552	all cancer sites	9050-9053
R	2	Mesothelioma of pleura and pericardium	1,83	1,81	1,85	28.676	C38	9050-9053
	3	Epithelioid malignant mesothelioma					C38	9052
	3	Sarcomatoid malignant mesothelioma					C38	9051
R	2	Mesothelioma of peritoneum and tunica vaginalis	0,13	0,13	0,14	2.065	C48, C63.7	9050-9053
	3	Epithelioid malignant mesothelioma					C48, C63.7	9052
	3	Sarcomatoid malignant mesothelioma					C48, C63.7	9051
	1	<b>MALIGNANT SKIN MELANOMA</b>	14,06	14,00	14,12	220.206	C44, C63.2, C51, C60	8720-8780
	2	Malignant skin melanoma	14,06	14,00	14,12	220.206	C44, C63.2, C51, C60	8720-8780
	3	Superficial spreading melanoma					C44, C63.2, C51, C60	8743
	3	Nodular melanoma					C44, C63.2, C51, C60	8721
	3	Lentigo maligna melanoma					C44, C63.2, C51, C60	8742
	3	Acral lentiginous melanoma malignant					C44, C63.2, C51, C60	8744
	1	<b>MALIGNANT MELANOMA OF MUCOSA AND EXTRACUTANEOUS</b>	0,15	0,14	0,15	2.279	C00-C26, C30-C34, C52, C53, C54, C55, C56, C57, C61, C62, C64, C65-C68,	8720-8780
R	2	Malignant melanoma of mucosa and extracutaneous	0,15	0,14	0,15	2.279	C00-C26, C30-C34, C52, C53, C54, C55, C56, C57, C61, C62, C64, C65-C68,	8720-8780
	1	<b>MALIGNANT MELANOMA OF UVEA</b>	0,70	0,69	0,72	11.022	C69.3-C69.4	8000, 8001, 8720-8780
R	2	Malignant melanoma of uvea	0,70	0,69	0,72	11.022	C69.3-C69.4	8000, 8001, 8720-8780
	1	<b>EPITHELIAL TUMOURS OF SKIN</b>	55,03	54,91	55,14	862.038	C44, C63.2,	8011, 8032, 8050-8052, 8070-8078, 8090-8098, 8560
	2	Basal cell carcinoma of skin	40,75	40,65	40,85	638.347	C44, C63.2,	8090-8098

**Supplementary data**

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R=rare	Tier	Tumour	Crude incidence rate per 100,000 per year in Europe	95% confidence interval		Number of cases collected in the RARECAREnet database from 2000-2007	ICD-O-3 Topography code	ICD-O-3 Morphology code
	3	Basal cell carcinoma NOS					C44, C63.2,	8090, 8092
	3	Basosquamous carcinoma					C44, C63.2,	8094
	3	Multifocal superficial basal cell carcinoma					C44, C63.2,	8091
	3	Basal cell carcinoma nodular					C44, C63.2,	8097
	3	Basal cell carcinoma fibroepithelial					C44, C63.2,	8093
	3	Adenoid basal carcinoma					C44, C63.2,	8098
	2	<b>Squamous cell carcinoma with variants of skin</b>	14,28	14,22	14,34	223.691	C44, C63.2,	8011, 8032, 8050-8052, 8070-8078, 8560
	3	Squamous carcinoma					C44, C63.2,	8070
	3	Verrucous carcinoma					C44, C63.2,	8051
	3	Spindle cell carcinoma					C44, C63.2,	8032, 8074
	3	Pseudo vascular/ Squamous cell carcinoma adenoid					C44, C63.2,	8075
	3	Adenosquamous carcinoma					C44, C63.2	8560
	1	<b>ADNEXAL CARCINOMA OF SKIN</b>	0,30	0,29	0,31	4.684	C44, C63.2	8100-8110, 8200, 8211, 8390-8420, 8480-8481, 8542, 8940
R	2	<b>Adnexal carcinoma of skin</b>	0,30	0,29	0,31	4.684	C44, C63.2	8100-8110, 8200, 8211, 8390-8420, 8480-8481, 8542, 8940
	3	Nodular hidradenoma, malignant					C44, C63.2	8402
	3	Sebaceous adenocarcinoma					C44, C63.2	8410
	3	Adenoid cystic carcinoma					C44, C63.2	8200
	3	Paget's disease extramammary					C44, C63.2	8542
	3	Apocrine adenocarcinoma					C44, C63.2	8401
	3	Mucinous adenocarcinoma					C44, C63.2	8480-8481
	3	Pilomatrix carcinoma					C44, C63.2	8110
	3	Eccrine poroma, malignant					C44, C63.2	8409
	3	Mixed tumour malignant, NOS					C44, C63.2	8940
	3	Sclerosing sweat duct carcinoma					C44, C63.2	8407
	3	Malignant eccrine spiradenoma					C44, C63.2	8403
	3	Tubular adenocarcinoma					C44, C63.2	8211
	3	Eccrine papillary adenocarcinoma					C44, C63.2	8408
	1	<b>NEUROBLASTOMA AND GANGLIONEUROBLASTOMA</b>	0,10	0,10	0,11	1.566	all cancers sites, except Central Nervous System and Autonomic Nervous	9490, 9500
R	2	<b>Neuroblastoma and ganglioneuroblastoma</b>	0,10	0,10	0,11	1.566	all cancers sites, except Central Nervous System and Autonomic Nervous	9490, 9500
	1	<b>NEPHROBLASTOMA</b>	0,13	0,12	0,13	1.965	C64.9	8960
R	2	<b>Nephroblastoma</b>	0,13	0,12	0,13	1.965	C64.9	8960
	1	<b>RETINOBLASTOMA</b>	0,05	0,05	0,06	860	C69.2	9510-9514
R	2	<b>Retinoblastoma</b>	0,05	0,05	0,06	860	C69.2	9510-9514
	1	<b>HEPATOBLASTOMA</b>	0,02	0,02	0,03	357	C22.0	8970
R	2	<b>Heptatoblastoma</b>	0,02	0,02	0,03	357	C22.0	8970
	1	<b>PLEUROPULMONARY BLASTOMA</b>	0,001	0,000	0,001	9	C34	8973
R	2	<b>Pleuropulmonary blastoma</b>	0,001	0,000	0,001	9	C34	8973
	1	<b>PANCREATOBLASTOMA</b>	0,002	0,002	0,003	39	C25	8971
R	2	<b>Pancreatoblastoma</b>	0,002	0,002	0,003	39	C25	8971
	1	<b>OLFACTORY NEUROBLASTOMA</b>	0,03	0,03	0,03	498	C30.0-C31.9	9520, 9522, 9523
R	2	<b>Olfactory neuroblastoma</b>	0,03	0,03	0,03	498	C30.0-C31.9	9520, 9522, 9523
	1	<b>ODONTOGENIC MALIGNANT TUMORS</b>	0,004	0,003	0,01	69	all sites	9270; 9290;9330; 9342
R	2	<b>Odontogenic malignant tumours</b>	0,004	0,003	0,01	69	all sites	9270; 9290;9330; 9342
	1	<b>EXTRAGONADAL GERM CELL TUMOURS</b>	0,12	0,11	0,12	1.862	all cancers sites except C56, C58 and C62	9060-9063, 9064, 9070, 9071, 9080, 9082, 9083, 9081, 9084, 9085, 9100,9101, 9102, 9105

R=rare	Tier	Tumour	Crude incidence rate per 100,000 per year in Europe	95% confidence interval		Number of cases collected in the RARECAREnet database from 2000-2007	ICD-O-3 Topography code	ICD-O-3 Morphology code
R	2	<b>Non seminomatous germ cell tumours</b>	0,06	0,05	0,06	915	all cancer sites except C56, C58, C62, C71-C72, C75.1, C75.3	9070, 9071, 9081, 9080, 9082, 9083, 9084, 9085, 9100,9101,9102,9105
	3	Embryonal adenocarcinoma, NOS					all cancer sites except C56, C58, C62, C71-C72, C75.1, C75.3	9070
	3	Yolk sac tumour					all cancer sites except C56, C58, C62, C71-C72, C75.1, C75.3	9071
	3	Teratocarcinoma					all cancer sites except C56, C58, C62, C71-C72, C75.1, C75.3	9081
	3	Teratoma with malignant transformation					all cancer sites except C56, C58, C62, C71-C72, C75.1, C75.3	9084
	3	Mixed germ cell tumour					all cancer sites except C56, C58, C62, C71-C72, C75.1, C75.3	9085
	3	Choriocarcinoma, NOS					all cancer sites except C56, C58, C62, C71-C72, C75.1, C75.3	9100
	3	Choriocarcinoma combined with other germ cell elements					all cancer sites except C56, C58, C62, C71-C72, C75.1, C75.3	9101
R	2	<b>Seminomatous germ cell tumors</b>	0,01	0,01	0,01	130	all cancer sites except C56, C58, C62, C71-C72, C75.1, C75.3	9060-9063
	3	Dysgerminoma					all cancer sites except C56, C58, C62, C71-C72, C75.1, C75.3	9060
	3	Seminoma, NOS					all cancer sites except C56, C58, C62, C71-C72, C75.1, C75.3	9061
	3	Seminoma, anaplastic					all cancer sites except C56, C58, C62, C71-C72, C75.1, C75.3	9062
	3	Spermatocytic seminoma					all cancer sites except C56, C58, C62, C71-C72, C75.1, C75.3	9063
R	2	<b>Germ cell tumors of Central Nervous System (CNS)</b>	0,04	0,03	0,04	574	C71-C72, C75.1, C75.3	9060-9063, 9064, 9070, 9071, 9080, 9081, 9082, 9083, 9084, 9085, 9100,9101, 9102, 9105
	3	Dysgerminoma					C71-C72, C75.1, C75.3	9060
	3	Seminoma, NOS					C71-C72, C75.1, C75.3	9061
	3	Seminoma, anaplastic					C71-C72, C75.1, C75.3	9062
	3	Spermatocytic seminoma					C71-C72, C75.1, C75.3	9063
	3	Embryonal adenocarcinoma, NOS					C71-C72, C75.1, C75.3	9070
	3	Yolk sac tumour					C71-C72, C75.1, C75.3	9071
	3	Teratocarcinoma					C71-C72, C75.1, C75.3	9081
	3	Teratoma with malignant transformation					C71-C72, C75.1, C75.3	9084
	3	Mixed germ cell tumour					C71-C72, C75.1, C75.3	9085
	3	Choriocarcinoma, NOS					C71-C72, C75.1, C75.3	9100
	3	Choriocarcinoma combined with other germ cell elements					C71-C72, C75.1, C75.3	9101

**Supplementary data**

**I tumori rari in Italia / The burden of rare cancer in Italy**  
*Epidemiol Prev* 2016; 40(1 Suppl 2): 1-120

R=rare	Tier	Tumour	Crude incidence rate per 100,000 per year in Europe	95% confidence interval		Number of cases collected in the RARECAREnet database from 2000-2007	ICD-O-3 Topography code	ICD-O-3 Morphology code
	1	<b>SOFT TISSUE SARCOMA</b>	4,71	4,68	4,74	73.796	all cancers sites except C40.0-C41.9	<b>all cancers sites except C40.0-C41.9 (includes unknown primary sites)</b> : 8710-8711, 8800-8935, 8959, 8963-8964, 8990-8991, 9020, 9040-9044, 9120-9133, 9150, 9170, 9180, 9231, 9240, 9251, 9260, 9364-9365, 9540, 9560-9571, 9580-9581 <b>C8, C60, C44, C63.2</b> : 8940 <b>all cancer sites except C40.0-C41.9, C56, C62, C71, C72</b> : 9473,
<b>R</b>	2	<b>Soft tissue sarcoma of head and neck</b>	0,26	0,25	0,27	4.087	C00.0-C14.8, C30.0-C32.9, C49.0, C73.9, C75.2, C75.4, C76.0	8710-8711, 8800-8902, 8912, 8921, 8933-8935, 8963, 8990-8991, 9040-9044, 9120-9133, 9150, 9170, 9180, 9231, 9240, 9364-9365, 9540, 9560-9571, 9580-9581
	3	Sarcoma NOS					C00.0-C14.8, C30.0, C31.0-C32.9, C47.0, C49.0, C73.9, C75.2, C75.4	8800
	3	Epithelioid sarcoma					C00.0-C14.8, C30.0, C31.0-C32.9, C47.0, C49.0, C73.9, C75.2, C75.4	8804
	3	Desmoplastic small round cell tumour					C00.0-C14.8, C30.0, C31.0-C32.9, C47.0, C49.0, C73.9, C75.2, C75.4	8806
	3	Fibrosarcoma					C00.0-C14.8, C30.0, C31.0-C32.9, C47.0, C49.0, C73.9, C75.2, C75.4	8810
	3	Fibromyxosarcoma					C00.0-C14.8, C30.0, C31.0-C32.9, C47.0, C49.0, C73.9, C75.2, C75.4	8811
	3	Infantile fibrosarcoma					C00.0-C14.8, C30.0, C31.0-C32.9, C47.0, C49.0, C73.9, C75.2, C75.4	8814
	3	Myxoinflammatory fibroblastic sarcoma					C00.0-C14.8, C30.0, C31.0-C32.9, C47.0, C49.0, C73.9, C75.2, C75.4	8825.1 borderline malignancy
	3	Malignant fibrous histiocytoma					C00.0-C14.8, C30.0, C31.0-C32.9, C47.0, C49.0, C73.9, C75.2, C75.4	8830
	3	DFSP					C00.0-C14.8, C30.0, C31.0-C32.9, C47.0, C49.0, C73.9, C75.2, C75.4	8832-8833
	3	Well differentiated liposarcoma					C00.0-C14.8, C30.0, C31.0-C32.9, C47.0, C49.0, C73.9, C75.2, C75.4	8851
	3	Myxoid liposarcoma					C00.0-C14.8, C30.0, C31.0-C32.9, C47.0, C49.0, C73.9, C75.2, C75.4	8852
	3	Pleomorphic liposarcoma					C00.0-C14.8, C30.0, C31.0-C32.9, C47.0, C49.0,	8854
	3	Mixed type liposarcoma					C00.0-C14.8, C30.0, C31.0-C32.9, C47.0, C49.0,	8855
	3	Dedifferentiated liposarcoma					C00.0-C14.8, C30.0, C31.0-C32.9, C47.0, C49.0, C73.9, C75.2, C75.4	8858
	3	Leiomyosarcoma					C00.0-C14.8, C30.0, C31.0-C32.9, C47.0, C49.0, C73.9, C75.2, C75.4	8890
	3	Pleomorphic rhabdomyosarcoma					C00.0-C14.8, C30.0, C31.0-C32.9, C47.0, C49.0, C73.9, C75.2, C75.4	8901

R=rare	Tier	Tumour	Crude incidence rate per 100,000 per year in Europe	95% confidence interval		Number of cases collected in the RARECAREnet database from 2000-2007	ICD-O-3 Topography code	ICD-O-3 Morphology code
	3	Extrarenal rhabdoid tumour					C00.0-C14.8, C30.0, C31.0-C32.9, C47.0, C49.0, C73.9, C75.2, C75.4	8963
	3	Synovial sarcoma					C00.0-C14.8, C30.0, C31.0-C32.9, C47.0, C49.0, C73.9, C75.2, C75.4	9040
	3	Clear cell sarcoma of soft tissue					C00.0-C14.8, C30.0, C31.0-C32.9, C47.0, C49.0, C73.9, C75.2, C75.4	9044
	3	Epithelioid angiosarcoma					C00.0-C14.8, C30.0, C31.0-C32.9, C47.0, C49.0, C73.9, C75.2, C75.4	9120
	3	Epithelioid hemangioendothelioma					C00.0-C14.8, C30.0, C31.0-C32.9, C47.0, C49.0, C73.9, C75.2, C75.4	9133
	3	Extraskelatal osteosarcoma					C00.0-C14.8, C30.0, C31.0-C32.9, C47.0, C49.0, C73.9, C75.2, C75.4	9180
	3	Extraskelatal myxoid chondrosarcoma					C00.0-C14.8, C30.0, C31.0-C32.9, C47.0, C49.0, C73.9, C75.2, C75.4	9231
	3	Malignant peripheral nerve sheath tumours					C00.0-C14.8, C30.0, C31.0-C32.9, C47.0, C49.0, C73.9, C75.2, C75.4	9540, 9561, 9571
	3	Malignant glomus tumours (glomangiosarcoma)					C00.0-C14.8, C30.0, C31.0-C32.9, C47.0, C49.0, C73.9, C75.2, C75.4	8710, 8711
	3	Intimal Sarcoma					C00.0-C14.8, C30.0, C31.0-C32.9, C47.0, C49.0, C73.9, C75.2, C75.4	malignancy without ICD-O morphology code
	3	Low grade fibromyxoid sarcoma					C00.0-C14.8, C30.0, C31.0-C32.9, C47.0, C49.0, C73.9, C75.2, C75.4	malignancy without ICD-O morphology code
	3	Low grade myofibroblastic sarcoma					C00.0-C14.8, C30.0, C31.0-C32.9, C47.0, C49.0, C73.9, C75.2, C75.4	malignancy without ICD-O morphology code
	3	PEComas					C00.0-C14.8, C30.0, C31.0-C32.9, C47.0, C49.0, C73.9, C75.2, C75.4	malignancy without ICD-O morphology code
	3	Sclerosing epithelioid fibrosarcoma					C00.0-C14.8, C30.0, C31.0-C32.9, C47.0, C49.0, C73.9, C75.2, C75.4	malignancy without ICD-O morphology code
	3	Undifferentiated pleomorphic sarcoma					C00.0-C14.8, C30.0, C31.0-C32.9, C47.0, C49.0, C73.9, C75.2, C75.4	8805, 8803
R	2	<b>Soft tissue sarcoma of limbs *</b>	1,10	1,08	1,11	17.178	C49.1-C49.2, C76.4-C76.5	8710-8711, 8800-8902, 8912, 8921, 8933-8935, 8940, 8963, 8990-8991, 9040-9044, 9120-9133, 9150, 9170, 9180, 9231, 9240, 9365, 9540, 9560-9571, 9580-9581
R	2	<b>Soft tissue sarcoma of superficial trunk *</b>	0,50	0,49	0,51	7.813	C49.3-C49.4, C49.6, C76.1-C76.2, C76.7	8710-8711, 8800-8902, 8912, 8921, 8933-8935, 8940, 8963, 8990-8991, 9040-9044, 9120-9133, 9150, 9170, 9180, 9231, 9240, 9365, 9540, 9560-9571, 9580-9581
R	2	<b>Soft tissue sarcoma of mediastinum *</b>	0,03	0,03	0,03	465	C38.1-C38.3, C38.8	8710-8711, 8800-8902, 8912, 8921, 8933-8935, 8940, 8963, 8990-8991, 9040-9044, 9120-9133, 9150, 9170, 9180, 9231, 9240, 9365, 9540, 9560-9571, 9580-9581
R	2	<b>Soft tissue sarcoma of heart *</b>	0,01	0,01	0,02	216	C38.0	8710-8711, 8800-8902, 8912, 8921, 8933-8935, 8940, 8963, 8990-8991, 9040-9044, 9120-9133, 9150, 9170, 9180, 9231, 9240, 9365, 9540, 9560-9571, 9580-9581

**Supplementary data**

**I tumori rari in Italia / The burden of rare cancer in Italy**  
*Epidemiol Prev* 2016; 40(1 Suppl 2): 1-120

R=rare	Tier	Tumour	Crude incidence rate per 100,000 per year in Europe	95% confidence interval		Number of cases collected in the RARECAREnet database from 2000-2007	ICD-O-3 Topography code	ICD-O-3 Morphology code
R	2	Soft tissue sarcoma of breast *	0,18	0,18	0,19	2.865	C50.0-C50.9	8710-8711, 8800-8902, 8912, 8921, 8933-8935, 8940, 8963, 8990-8991, 9020, 9040-9044, 9120-9133, 9150, 9170, 9180, 9231, 9240, 9365, 9540, 9560-9571, 9580-9581
R	2	Soft tissue sarcoma of uterus *	0,55	0,54	0,56	8.657	C53.0-C55.9	8710-8711, 8800-8902, 8912, 8921-8931, 8933, 8934-8935, 8940, 8963, 8990-8991, 9040-9044, 9120-9133, 9150, 9170, 9180, 9231, 9240, 9365, 9540, 9560-9571, 9580-9581
R	2	Soft tissue sarcoma of paratestis *	0,03	0,03	0,04	510	C63.0-C63.7	8710-8711, 8800-8902, 8912, 8921, 8933-8935, 8940, 8963, 8990-8991, 9040-9044, 9120-9133, 9150, 9170, 9180, 9231, 9240, 9365, 9540, 9560-9571, 9580-9581
R	2	Soft tissue sarcomas of other genitourinary tract (vulva, vagina, ovary, penis, prostate, testis, kidney, renal pelvis, ureter, bladder, urethra)	0,20	0,19	0,21	3.160	C51.0-C52.9, C56.9-C57.9, C60.0-C62.9, C63.8-C63.9, C64.9-C68.9	<b>C51.0-C52.9, C56.9-C57.9, C60.0-C62.9, C63.8-C63.9, C64.9-C68.9:</b> 8710-8711, 8800-8902, 8912, 8921, 8933, 8934-8935, 8959, 8963-8964, 8990-8991, 9040-9044, 9120-9133, 9150, 9170, 9180, 9231, 9240, 9365, 9540, 9560-9571, 9580-9581 <b>C51.0-C52.9, C56.9-C57.9, C61.9-C62.9, C63.8-C63.9, C64.9-C68.9:</b> 8940
	3	Clear cell sarcoma of kidney					64,9	8964
	3	Malignant rhabdoid tumor of kidney					64,9	8963
R	2	Soft tissue sarcoma of viscera *	0,38	0,37	0,39	6.004	C15.0-C26.9, C33.9-C37.9, C38.4, C39.0-C39.9, C42.2	8710-8711, 8800-8902, 8912, 8921, 8933-8935, 8940, 8963, 8990-8991, 9040-9044, 9120-9133, 9150, 9170, 9180, 9231, 9240, 9365, 9540, 9560-9571, 9580-9581
R	2	Soft tissue sarcoma of retroperitoneum and peritoneum *	0,31	0,30	0,32	4.911	C48.0-C48.8	8710-8711, 8800-8902, 8912, 8921, 8933-8935, 8940, 8963, 8990-8991, 9040-9044, 9120-9133, 9150, 9170, 9180, 9231, 9240, 9365, 9540, 9560-9571, 9580-9581
R	2	Soft tissue sarcoma of pelvis *	0,20	0,19	0,20	3.090	C49.5, C76.3	8710-8711, 8800-8902, 8912, 8921, 8933-8935, 8940, 8963, 8990-8991, 9040-9044, 9120-9133, 9150, 9170, 9180, 9231, 9240, 9365, 9540, 9560-9571, 9580-9581
R	2	Soft tissue sarcoma of skin *	0,30	0,29	0,31	4.737	C44.0-C44.9	8710-8711, 8800-8902, 8912, 8921, 8933-8935, 8963, 8990-8991, 9040-9044, 9120-9133, 9150, 9170, 9180, 9231, 9240, 9365, 9540, 9560-9571, 9580-9581
R	2	Soft tissue sarcoma of paraorbit*	0,01	0,01	0,01	117	C69.0-C69.9	8710-8711, 8800-8902, 8912, 8921, 8933-8935, 8940, 8963, 8990-8991, 9040-9044, 9120-9133, 9150, 9170, 9180, 9231, 9240, 9365, 9540, 9560-9571, 9580-9581
R	2	Soft tissue sarcoma of brain and other parts of the nervous system *	0,17	0,17	0,18	2.723	C47.0-C47.9, C70.0-C72.9, C75.1, C75.3	8710-8711, 8800-8902, 8912, 8921, 8933-8935, 8940, 8963, 8990-8991, 9040-9044, 9120-9133, 9150, 9170, 9180, 9231, 9240, 9365, 9540, 9560-9571, 9580-9581
		* includes all the entities listed for the soft tissue of the Head and Neck						
R	2	Embryonal rhabdomyosarcoma of soft tissue	0,05	0,05	0,06	836	all cancers sites except C40.0-C41.9	8910
R	2	Alveolar rhabdomyosarcoma of soft tissue	0,03	0,03	0,04	519	all cancers sites except C40.0-C41.9, C56 and C62	8920
R	2	Ewing's sarcoma of soft tissue	0,06	0,06	0,07	998	all cancers sites except C40.0-C41.9, C56 and C62, C71, C72	9260, 9364, 9473
	3	Ewing Sarcoma					all cancers sites except C40.0-C41.9, C56 and C62, C71, C72	9260
	3	Peripheral neuroectodermal tumor					all cancers sites except C40.0-C41.9, C56 and C62, C71, C72	9364
	3	Primitive neuroectodermal tumor, NOS					all cancers sites except C40.0-C41.9, C56 and C62, C71, C72	9473





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R=rare	Tier	Tumour	Crude incidence rate per 100,000 per year in Europe	95% confidence interval		Number of cases collected in the RARECAREnet database from 2000-2007	ICD-O-3 Topography code	ICD-O-3 Morphology code
	1	<b>BONE SARCOMA</b>	0,85	0,84	0,87	13.376	C40.0-C41.9	8800-8801, 8803-8806, 8810, 8811-8812, 8815, 8830, 8840, 8850-8855, 8890-8891, 8894-8896, 8900-8902, 8910, 8912, 8920, 9040-9044, 9120-9133, 9150, 9170, 9180-9250, 9260-9261, 9310, 9364, 9370-9372, 9540-9581
R	2	<b>Osteogenic sarcoma</b>	0,21	0,21	0,22	3.330	C40.0-C41.9	9180,9181,9182,9183, 9185, 9187, 9192-9194
	3	Conventional osteosarcoma					C40.0-C41.9	9180
	3	Teleangiectatic osteosarcoma					C40.0-C41.9	9183
	3	Small cell osteosarcoma					C40.0-C41.9	9185
	3	Parosteal osteosarcoma					C40.0-C41.9	9192
	3	High grade surface osteosarcoma					C40.0-C41.9	9194
	3	Low grade central osteosarcoma					C40.0-C41.9	9187
	3	Periosteal osteosarcoma					C40.0-C41.9	9193
	3	Secondary osteosarcoma					C40.0-C41.9	malignancy without ICD-O morphology code
R	2	<b>Chondrogenic sarcomas</b>	0,26	0,25	0,27	4.107	C40.0-C41.9	9220, 9240, 9242-9243
	3	Chondrosarcoma					C40.0-C41.9	9220
	3	Mesenchymal chondrosarcoma					C40.0-C41.9	9240
	3	Clear cell chondrosarcoma					C40.0-C41.9	9242
	3	Dedifferentiated chondrosarcoma					C40.0-C41.9	9243
R	2	<b>Notochordal sarcomas, chordoma</b>	0,07	0,07	0,08	1.145	C40.0-C41.9	9370, 9371, 9372
R	2	<b>Vascular sarcomas</b>	0,01	0,01	0,01	129	C40.0-C41.9	9120-9133, 9150, 9170
	3	Angiosarcoma					C40.0-C41.9	9120
R	2	<b>Ewing's sarcoma</b>	0,12	0,12	0,13	1.943	C40.0-C41.9	9260, 9364
	3	Peripheral neuroectodermal tumor					C40.0-C41.9	9364
R	2	<b>Epithelial tumours, adamantinoma</b>	0,01	0,01	0,02	213	C40.0-C41.9	9261, 9310
R	2	<b>Other high grade sarcomas (fibrosarcoma, malignant fibrous histiocytoma)</b>	0,02	0,02	0,02	304	C40.0-C41.9	8810, 8830
	3	Fibrosarcoma					C40.0-C41.9	8810
	3	Malignant fibrous histiocytoma					C40.0-C41.9	8830
	1	<b>GASTROINTESTINAL STROMAL SARCOMA</b>	0,30	0,29	0,31	4.706	any sites	8936
R	2	<b>Gastrointestinal stromal sarcoma</b>	0,30	0,29	0,31	4.706	any sites	8936
	1	<b>KAPOSI'S SARCOMA</b>	0,25	0,24	0,26	3.893	any sites	9140
R	2	<b>Kaposi's sarcoma</b>	0,25	0,24	0,26	3.893	any sites	9140
	1	<b>NEUROENDOCRINE TUMOURS</b>	3,51	3,48	3,54	54.942	all sites (except poorly diff in the lung C34.0-34.9)	<b>C15.0-C26.9, C48.0-C48.8:</b> 8002, 8013, 8041-8045, 8150, 8151-8153, 8154, 8155-8157, 8240-8245, 8246, 8248-8249 <b>C73.9:</b> 8002, 8013,8041,8240,8246,8249,8345-8347,8510, 8512 <b>C44.0-C44.9, C51.0-C51.9,C60.0,C60.9,C63.2:</b> 8002, 8013,8041,8240,8246 <b>C00.0-80.9:</b> 8247 <b>C34.0-34.9:</b> 8240, 8249, 8241, 8242, 8243, 8244, 8245, <b>C74.0-74.9:</b> 8700 <b>all cancer sites except: C15.0-C26.9,C34.0-8246,8248-8249,</b> <b>C00.0-C80.9:</b> 8680, 8693
R	2	<b>Well differentiated not functioning endocrine carcinoma of pancreas and digestive tract</b>	1,01	1,00	1,03	15.852	C15-26, C48	8150, 8240-8245, 8248-8249,
	3	Carcinoid tumour					C15-26, C48	8240-8245, 8248-8249
	3	Islet cell carcinoma					C15-26, C48	8150
R	2	<b>Well differentiated functioning endocrine carcinoma of pancreas and digestive tract</b>	0,03	0,02	0,03	411	C15-26, C48	8151-8153, 8155-8157
	3	Insulinoma, malignant					C15-26, C48	8151

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	3	Glucagonoma, malignant						C15-26, C48	8152 and 8157
	3	Somatostatinoma, malignant						C15-26, C48	8156
	3	Gastrinoma, malignant						C15-26, C48	8153
	3	VIP-oma						C15-26, C48	8155
	3	Other ectopic hormone producing tumours						C15-26, C48	malignancy without ICD-O morphology code
R	2	Poorly differentiated endocrine carcinoma of pancreas and digestive tract	0,67	0,65	0,68	10.421	C15-26, C48	8013, 8041-8045, 8246, 8002	
	3	Large cell endocrine carcinoma					C15-26, C48	8013	
	3	Small cell endocrine carcinoma					C15-26, C48	8041-8045, 8002	
R	2	Mixed endocrine-exocrine carcinoma of pancreas and digestive tract	0,01	0,01	0,01	147	C15-26	8154	
R	2	Endocrine carcinoma of thyroid gland	0,24	0,23	0,25	3.796	C73.9	8002, 8013, 8041, 8240, 8246, 8249, 8345-8347, 8510, 8512	
	3	Medullary carcinoma					C73.9	8510, 8345, 8246	
	3	Mixed medullary-follicular carcinoma					C73.9	8346	
	3	Well differentiated endocrine tumours					C73.9	8240, 8249	
	3	Poorly differentiated endocrine carcinoma					C73.9	8013, 8041, 8002	
R	2	Neuroendocrine carcinoma of skin	0,19	0,19	0,20	3.024	C44.0-C44.9, CC51.0-C51.9, C60.0,C60.9 C63.2	8013, 8041, 8002, 8240, 8246 and 8247 in all sites	
	3	Merkel cell carcinoma					C44.0-C44.9, CC51.0-C51.9, C60.0,C60.9 C63.2	8013, 8041, 8240, 8246 and 8247 in all sites	
R	2	Typical and atypical carcinoid of the lung	0,39	0,38	0,40	6.160	C34	8240, 8241, 8242, 8243, 8244, 8245, 8249	
R	2	Neuroendocrine carcinoma of other sites	0,90	0,89	0,92	14.120	All cancer sites except C15-C26, C34, C44, C48, C51, C60, C63.2, C73	8002, 8013, 8041-8045, 8150-8153, 8155-8157, 8240-8246, 8248-8249,	
	3	Well differentiated endocrine tumours, carcinoid					All cancer sites except C15-C26, C34, C44, C48, C51, C60, C63.2, C73	8150-8153, 8155-8157, 8240-8245, 8248,	
	3	Well differentiated endocrine tumours, atypical carcinoid					All cancer sites except C15-C26, C34, C44, C48, C51, C60, C63.2, C73	8249	
	3	Poorly differentiated endocrine carcinoma					All cancer sites except C15-C26, C34, C44, C48, C51, C60, C63.2, C73	8013, 8041-8045, 8246	
R	2	Pheochromocytoma, malignant	0,04	0,04	0,04	650	C74	8700	
R	2	Paraganglioma	0,02	0,02	0,02	347	any site	8680, 8693,	
	1	CARCINOMAS OF PITUITARY GLAND	0,04	0,03	0,04	582	C75.1	8000-8001, 8010, 8140-8147, 8180-8231, 8250-8381	
R	2	Carcinoma of pituitary gland	0,04	0,03	0,04	582	C75.1	8000-8001, 8010, 8140-8147, 8180-8231, 8250-8381	
	3	Pituitary carcinoma					C75.1	8272	
	1	CARCINOMAS OF THYROID GLAND	5,07	5,03	5,11	79.420	C73.9	8000-8001, 8003-8004, 8005, 8010, 8011, 8012, 8014-8035, 8050-8084, 8140, 8141, 8190, 8200, 8201, 8211, 8230, 8260, 8261-8263, 8290, 8310, 8320-8323, 8330-8333, 8334-8344, 8350, 8360, 8380, 8430, 8440, 8450, 8480, 8481, 8490, 8500, 8504, 8514, 8520, 8550, 8560, 8562, 8574, 8588-8589, 8941, 8980, 8982	
R	2	Carcinoma of thyroid gland	5,07	5,03	5,11	79.420	C73.9	8000-8001, 8003-8004, 8005, 8010, 8011, 8012, 8014-8035, 8050-8084, 8140, 8141, 8190, 8200, 8201, 8211, 8230, 8260, 8261-8263, 8290, 8310, 8320-8323, 8330-8333, 8334-8344, 8350, 8360, 8380, 8430, 8440, 8450, 8480, 8481, 8490, 8500, 8504, 8514, 8520, 8550, 8560, 8562, 8574, 8588-8589, 8941, 8980, 8982	
	3	Papillary adenocarcinoma, NOS					C73.9	8050, 8260	
	3	Follicular carcinoma, NOS					C73.9	8330	
	3	Undifferentiated/anaplastic carcinoma					C73.9	8020-8033, 8041	



**Supplementary data**  
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R=rare	Tier	Tumour	Crude incidence rate per 100,000 per year in Europe	95% confidence interval		Number of cases collected in the RARECAREnet database from 2000-2007	ICD-O-3 Topography code	ICD-O-3 Morphology code
	3	Mucoepidermoid carcinoma					C73.9	8430
	3	Mucinous carcinoma					C73.9	8480-8481
	3	Spindle cell tumour with thymus-like differentiation (SETTLE)					C73.9	8588
	3	Carcinoma showing thymus-like differentiation (CASTLE)					C73.9	8589
	1	<b>CARCINOMAS OF PARATHYROID GLAND</b>	0,03	0,02	0,03	410	C75.0.	8000-8001, 8010-8012, 8014-8035, 8050-8076, 8140, 8147, 8190, 8200, 8211, 8230-8231, 8255, 8260-8263, 8290, 8310, 8320, 8322, 8323, 8430, 8440, 8480, 8481, 8490, 8504, 8510, -8512, 8514, 8525, 8542, 8550-8551, 8560, 8562, 8570-8573, 8575-8576, 8980
R	2	<b>Carcinomas of parathyroid gland</b>	0,03	0,02	0,03	410	C75.0.	8000-8001, 8010-8012, 8014-8035, 8050-8076, 8140, 8147, 8190, 8200, 8211, 8230-8231, 8255, 8260-8263, 8290, 8310, 8320, 8322, 8323, 8430, 8440, 8480, 8481, 8490, 8504, 8510, -8512, 8514, 8525, 8542, 8550-8551, 8560, 8562, 8570-8573, 8575-8576, 8980
	1	<b>CARCINOMA OF ADRENAL CORTEX</b>	0,22	0,21	0,23	3.424	C74	8000-8001, 8003, 8005, 8010-8012, 8014-8035, 8050-8076, 8140, 8147, 8190, 8211, 8230-8231, 8255, 8260-8263, 8290, 8310, 8315, 8320, 8323, 8370, 8430, 8440, 8480, 8481, 8490, 8504, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8560, 8562, 8570-8573, 8575-8576, 8671, 8980,
R	2	<b>Carcinoma of adrenal cortex</b>	0,22	0,21	0,23	3.424	C74	8000-8001, 8003, 8005, 8010-8012, 8014-8035, 8050-8076, 8140, 8147, 8190, 8211, 8230-8231, 8255, 8260-8263, 8290, 8310, 8315, 8320, 8323, 8370, 8430, 8440, 8480, 8481, 8490, 8504, 8510, 8512, 8514, 8525, 8542, 8550-8551, 8560, 8562, 8570-8573, 8575-8576, 8671, 8980,
	3	Adrenal cortical carcinoma					C74	8370
	1	<b>TUMOURS OF CENTRAL NERVOUS SYSTEM (CNS)</b>	7,36	7,32	7,40	115.289	C70, C71, C72,	<b>C71, C72:</b> 8000, 8001, 9380-9382, 9384, 9400-9442, 9450-9451, 9460, 9383, 9391-9394, 9505, 9521 <b>C71.5:</b> 8000, 8001, 9390 <b>C70:</b> 8000, 8001, 9530, 9538-9539
R	2	<b>Astrocytic tumours of CNS</b>	4,99	4,95	5,02	78.118	C71, C72	9380-9382, 9384, 9400-9442,
	3	Astrocytoma, NOS					C71, C72	9400
	3	Astrocytoma, anaplastic					C71, C72	9401
	3	Glioblastoma					C71,	9440
	3	Gliosarcoma					C71, C72	9442
	3	Giant cell glioblastoma					C71, C72	9441
	3	Astroblastoma					C71, C72	9430
	3	Protoplasmic astrocytoma					C71, C72	9410
	3	Gliomatosis cerebri					C71, C72	9381
	3	Pleomorphic xanthoastrocytoma					C71, C72	9424
	3	Pilocytic and Pilo-mixoid astrocytoma					C71, C72	9421/1 borderline tumour
	3	Oligoastrocytoma					C71, C72	9382
	3	Subependymal giant cell astrocytoma					C71, C72	9384/1 borderline tumour
R	2	<b>Oligodendroglial tumours of CNS</b>	0,39	0,38	0,40	6.148	C71, C72	9450-9451, 9460
	3	Oligodendroglioma					C71, C72	9450
	3	Oligodendroglioma, anaplastic					C71, C72	9451
R	2	<b>Ependymal tumours of CNS</b>	0,21	0,20	0,21	3.212	C71, C72	9383, 9391-9394
	3	Ependymoma, NOS					C71, C72	9391
	3	Ependymoma, anaplastic					C71, C72	9392/33 (poorly differentiated, WHO grading III)
	3	Subependymoma					C71, C72	9383/1 borderline malignancy
	3	Myxopapillary ependymoma					C71, C72	9394/1 borderline malignancy
R	2	<b>Neuronal and mixed neuronal-glia tumors</b>	0,005	0,004	0,01	74	C71, C72	9493/0, 9412/1, 9413/0, 9492/0, 9505/1, 9506/1, 9505/3, 9521
R	2	<b>Choroid plexus carcinoma of CNS</b>	0,01	0,01	0,01	98	C71.5	9390
R	2	<b>Malignant meningiomas</b>	0,16	0,16	0,17	2.564	C70	9530, 9538-9539

**Supplementary data**

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R=rare	Tier	Tumour	Crude incidence rate per 100,000 per year in Europe	95% confidence interval		Number of cases collected in the RARECAREnet database from 2000-2007	ICD-O-3 Topography code	ICD-O-3 Morphology code
	1	<b>EMBRYONAL TUMORS OF CNS</b>	0,20	0,19	0,21	3.102	C71, C72, C75.3	<b>C71, C72:</b> 9392/34, 9470-9474, 9490, 9500-9504, 9508
R	2	<b>Embryonal tumors of CNS</b>	0,20	0,19	0,21	3.102	C71, C72, C75.3	<b>C75.3:</b> 9362 9508 <b>C71, C72:</b> 9392/34, 9470-9474, 9490, 9500-9504,
	3	Medulloblastoma					C71, C72	9470
	3	Desmoplastic nodular medulloblastoma					C71, C72	9471
	3	CNS-PNET					C71, C72	9473, 9490, 9500
	3	Atypical teratoid/rhabdoid tumour					C71, C72	9508
	3	Medulloepithelioma, NOS					C71, C72	9501
	3	Pineoblastoma					C75.3	9362
	3	Ependymoblastoma					C71, C72	9392/34
	3	Neuroepithelioma, NOS					C71, C72	9503
	1	<b>LYMPHOID DISEASES</b>	29,12	29,04	29,21	456.200		9590, 9591, 9596, 9650-9655, 9659, 9661-9667, 9670-9671, 9673, 9675, 9678-9680, 9684, 9687, 9689-9691, 9695, 9698-9702, 9705, 9708-9709, 9714, 9716-9719, 9727-9729, 9731-9734, 9760-9762, 9764, 9820, 9823, 9826-9827, 9831-9837, 9940, 9948
R	2	<b>Hodgkin lymphoma, classical</b>	2,46	2,44	2,49	38.588		9596, 9650-9655, 9661-9667
	3	Lymphocyte-rich						9651
	3	Mixed cellularity						9652
	3	Lymphocyte-depleted						9653-9655
	3	Nodular sclerosis						9663-9667
	3	Composite Hodgkin and Non Hodgkin lymphoma						9596
R	2	<b>Hodgkin lymphoma nodular lymphocyte predominance</b>	0,09	0,09	0,10	1.483		9659
R	2	<b>Precursor B/T lymphoblastic leukaemia/lymphoblastic lymphoma (and Burkitt leukemia/lymphoma)</b>	1,46	1,44	1,47	22.795		9687, 9727-9729, 9826, 9835-9837
	3	Precursor B/T lymphoblastic leukaemia/lymphoblastic lymphoma						9727-9729, 9835-9837
	3	Burkitt leukaemia/lymphoma						9687, 9826
R	2	<b>T cutaneous lymphoma (Sezary syn, Mycosis fung)</b>	0,35	0,34	0,36	5.526		9700-9701, 9709, 9718
R	2	<b>Other T cell lymphomas and NK cell neoplasms</b>	0,62	0,60	0,63	9.656		9702, 9705, 9708, 9714, 9716-9717, 9719, 9827, 9831, 9834, 9948
R	2	<b>Diffuse B lymphoma</b>	4,32	4,29	4,35	67.645		9675, 9678-9680, 9684
R	2	<b>Follicular B lymphoma</b>	2,19	2,17	2,22	34.346		9690-9691, 9695, 9698
R	2	<b>Hairy cell leukaemia</b>	0,28	0,27	0,29	4.375		9940
R	2	<b>Plasmacytoma/Multiple Myeloma (and Heavy chain diseases)</b>	5,71	5,67	5,75	89.440		9731-9734, 9762
	3	Plasmacytoma/Multiple Myeloma						9731-9734
	3	Heavy chain disease						9762
	2	<b>Other non Hodgkin, Mature B cell lymphoma</b>	6,37	6,33	6,41	99.729		9670, 9671, 9689, 9699, 9761, 9764, 9823
	3	Small cell lymphocytic lymph/chronic lymphatic leukaemia						9670, 9823
	3	Lymphoma lymphoplasmatic/macroglobulinemia Waldstrom						9671, 9761
	3	Splenic/nodal marginal zone/extra nodal MALT type						9689, 9699, 9764
R	2	<b>Mantle cell lymphoma</b>	0,56	0,55	0,57	8.748		9673
R	2	<b>Prolymphocytic leukaemia, B cell</b>	0,05	0,05	0,06	804		9832-9833
	1	<b>ACUTE MYELOID LEUKEMIA AND RELATED PRECURSOR NEOPLASMS</b>	3,81	3,77	3,84	59.608		9805, 9840, 9860-9861, 9866-9867, 9870-9874, 9891, 9895-9897, 9910, 9920, 9930-9931, 9984, 9987

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R	2	<b>Acute promyelocytic leukemia (AML with t(15;17) with variants</b>	0,12	0,11	0,13	1.876		9866
R	2	<b>AML</b>	3,50	3,47	3,53	54.789		9805, 9840, 9861, 9867, 9870, 9871, 9872-9874, 9891, 9895, 9896, 9897, 9910, 9920, 9930-9931, 9984, 9987
	3	AML with t(8:21)(q22;q22); AML 1/ETO)						9896
	3	AML with inv(16)(p13.1q22)or t( 16; 16) (p13.1q22),(CBFbeta/MYH)						9871
	3	Acute myeloid leukemia 11q23 abnormalities						9897
	3	AML NOS (FAB or WHO type)						9861
	3	AML with myelodysplasia-related changes						9895
	3	Refractory Anemia with excess blasts in trasformation (RAEB-T)						9984
	3	Therapy related myelodysplastic syndrome, NOS						9987
	3	Therapy-related myeloid neoplasms						9920
	3	Acute myeloid leukemia minimally differentiation						9872
	3	Acute myeloid leukemia without maturation						9873
	3	Acute myeloid leukemia with maturation						9874
	3	Acute myelomonocytic leukemia						9867
	3	Acute myeloid leukemia M6 type						9840
	3	Acute monocytic leukemia						9891
	3	Acute basophilic leukemia						9870
	3	Acute megakaryoblastic leukemia						9910
	3	Acute panmyelosis with myelofibrosis						9931
	3	Acute leukemias with ambiguous lineage						9805
	3	Myeloid sarcoma						9930
	3	Blastic plasmacytoid dendritic cell neoplasm						ICD-O code not available
	1	<b>MYELOPROLIFERATIVE NEOPLASMS</b>	3,31	3,28	3,34	51.888		9740-9742, 9863, 9875, 9950, 9960-9964
R	2	<b>Chronic myeloid leukemia</b>	1,12	1,10	1,13	17.473		9863, 9875
	3	Chronic myelogenous leukemia BCR/ABL positive						9875
	3	Chronic myeloid leukemia NOS						9863
R	2	<b>Other myeloproliferative neoplasms</b>	2,17	2,14	2,19	33.954		9961, 9962, 9950, 9960, 9963-9964
	3	Primary myelofibrosis						9961
	3	Essential thrombocythemia						9962
	3	Polycythemia vera						9950
	3	Chronic myeloproliferative disease NOS						9960
	3	Chronic neutrophilic leukemia						9963
	3	Hypereosinophilic syndrome						9964
R	2	<b>Mast cell tumour</b>	0,03	0,03	0,03	461		9740-9742
	3	Mastocytoma NOS/mast cell sarcoma						9740
	3	Malignant mastocytosis						9741
	3	Mast cell leukaemia						9742
	1	<b>MYELODYSPLASTIC SYNDROME AND MYELODYSPLASTIC/MYELOPROLIFERATIVE DISEASES</b>	2,47	2,45	2,50	38.739		9876, 9945-9946, 9975, 9980, 9982-9983, 9985-9986, 9989
R	2	<b>Myelodysplastic syndrome with 5q syndrome</b>	0,01	0,01	0,01	156		9986

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R	2	<b>Other myelodysplastic syndrome</b>	2,14	2,12	2,16	33.542		9980, 9982, 9983, 9985, 9989
	3	Refractory anemia						9980
	3	Refractory anemia with sideroblasts						9982
	3	Refractory anemia with excess blasts						9983
	3	Refractory cytopenia with multilineage dysplasia						9985
	3	Myelodysplastic syndrome NOS						9989
R	2	<b>Chronic Myelomonocytic leukemia</b>	0,29	0,28	0,30	4.542		9945, 9946
	3	Juvenile myelomonocytic leukemia						9946
	3	Chronic myelomonocytic leukemia NOS						9945
R	2	<b>Atypical chronic myeloid leukemia BCR/ABL negative</b>	0,02	0,01	0,02	239		9876
	1	<b>HISTIOCYTIC AND DENDRITIC CELL NEOPLASMS</b>	0,05	0,05	0,06	828		9750-9758
R	2	<b>Histiocytic malignancies</b>	0,04	0,04	0,05	656		9750-9754, 9756
	3	Langherans cell histiocytosis						9751-9754
	3	Langherans cell sarcoma						9756
R	2	<b>Lymph node accessory cell tumors</b>	0,01	0,01	0,01	172		9755, 9757, 9758
	3	Histiocytic sarcoma						9755
	3	Interdigitating dendritic cell sarcoma						9757
	3	Follicular dendritic cell sarcoma						9758
	3	Fibroblastic reticular cell tumour						9759 according to the ICD-O3 update of the 2011.

NOS = not otherwise specified

le topografie C49.8 e C49.9 non inclusa nei tier 2 spiegano 4000 casi in più del tier 1. **8930, 8931** (nel tier 1 sono in tutte le sedi mentre nel tier 2 solo nel corpus uteri ma differenze minime; 25 casi ), **8964** (nel tier 1 è in tutte le sedi mentre è presente solo in 2 tier 2: superficial trunk e other viscera: eliminare dal superficial trunk MA, nessun caso al di fuori del kidney), **9020** (è tipico del brest quindi inserito nel tier 2 breast. 1900 casi), **9251** non è in nessun tier 2 (127 casi). spiego 6100 casi di differenza tra tier 1 e tier 2. **Ne restano altri 1100**