A GUIDE TO THE CANCER-SPECIFIC DATA SHEETS

GUIDA ALLA LETTURA DELLE SCHEDE SPECIFICHE PER TUMORE

HOW TO READ THE CANCER-SPECIFIC DATA SHEETS

This guide is for laymen. For more detailed information please refer to the «Material and methods» chapter (pp. 14-21).

This monograph includes 14 data sheets. Every sheet shows data of a major group of rare tumours identified by combining rare cancers of broad anatomic sites sharing the same clinical referral pattern and expertise.

In this guide the **Rare epithelial tumours of the thoracic cavity** group is used as an example to clarify how rare cancers were identified and described in each group.

HOW THE RARE CANCER GROUPS WERE IDENTIFIED

The rare cancers included in each of the 14 groups come from the RARECARE list of cancers (see Annex 1, supplementary material on-line). Briefly, the RARECARE list of cancers has a hierarchal structure:

- **tier 2** includes several specific cancer histotypes (identified by the ICD-O-3 morphology and topography codes) considered to require similar clinical management and research;
- **tier 1** includes the tier 2 entities plus the Not Otherwise Specified (NOS) morphologies. Tier 1 includes cancers considered to involve the same clinical expertise and patient referral structure.

In this AIRTUM monograph, tier 1 is written in green uppercase, tier 2 entities are written in black below the tier 1 they belong to, and are never in all caps.

In the RARECARE list, a tier 1 can be common or rare, but in this monograph only data on *rare* cancers are presented, therefore:

- **if a tier 1 is common** (incidence >6 per 100,000 at EU level), such as epithelial tumours of lung, the common tier 2 entities of this tier and the NOS histotype are excluded from the tier 1 definition (e.g., for the lung: squamous cell carcinoma, adenocarcinoma, and poorly differentiated endocrine carcinoma are excluded from tier 1 together with the NOS histotypes; see the example below). As its common cancers and the NOS morphologies are excluded, the tier 1 is labelled as **RARE** EPITHELIAL TUMOURS OF LUNG (to clarify that only the rare histotypes are included);
- **if a tier 1 is rare** (incidence rate <6 per 100,000 at EU level), such as epithelial tumours of trachea, all the corresponding tier 2 entities as well as the NOS histotypes are included in the tier 1 definition. The label will not include the "*rare*" specification because all the included cancers are rare.

EPITHELIAL TUMOURS OF TRACHEA
Tier 1 includes all the corresponding tier 2 entities, as well as the NOS histotypes. It is not necessary to add the "rare" specification to the label.

EPITHELIAL TUMOURS OF LUNG
Tier 1 includes only the rare corresponding tier 2 entities, while common tier 2 entities and NOS morphologies are excluded. The label includes the "rare" specification.

AN EXAMPLE

RARE EPITHELIAL TUMOURS OF THE THORACIC CAVITY

MAJOR GROUP	RARE EPITHELIAL TUMOURS OF THE THORACIC CAVITY	TOPOGRAPHY ICD-O-3 CODE	MORPHOLOGY ICD-O-3 CODE
TIER 1	EPITHELIAL TUMOURS OF TRACHEA	C33.9	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
Tier 2	Squamous cell carcinoma with variants of trachea	C33.9	8004, 8020-8022, 8031-8032, 8050-8076, 8078, 8082-8084, 8560, 8980
Tier 2	Adenocarcinoma with variants of trachea	C33.9	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
Tier 2	Salivary gland type tumours of trachea	C33.9	8200, 8430, 8982
TIER 1	RARE EPITHELIAL TUMOURS OF LUNG	C34.0-34.9	8560, 8012, 8014, 8034, 8071, 8072, 8074, 8123, 8200, 8430, 8982, 8004, 8022, 8030-8033, 8074, 8972, 8980
COMMON	~~Squamous cell carcinoma with variants of lung~~	C34.0-34.9	
COMMON	~~Adenocarcinoma with variants of lung~~	C34.0-34.9	
Tier 2	Adenosquamous carcinoma of lung	C34.0-34.9	8560
Tier 2	Large cell carcinoma of lung	C34.0-34.9	8012, 8014, 8034, 8071-8072, 8123
COMMON	~~Poorly differentiated endocrine carcinoma of lung~~	C34.0-34.9	
Tier 2	Salivary gland type tumours of lung	C34.0-34.9	8200, 8430, 8982
Tier 2	Sarcomatoid carcinoma of lung	C34.0-34.9	8004, 8022, 8030-8033, 8074, 8972, 8980
TIER 1	EPITHELIAL TUMOURS OF THYMUS	C37.9	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...

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AN EXAMPLE
RARE EPITHELIAL TUMOURS OF THE THORACIC CAVITY

INCIDENCE (OF THE MAJOR GROUP)
Estimated number of Italians (M+F) diagnosed with a rare epithelial tumour of the thoracic cavity in 2015.

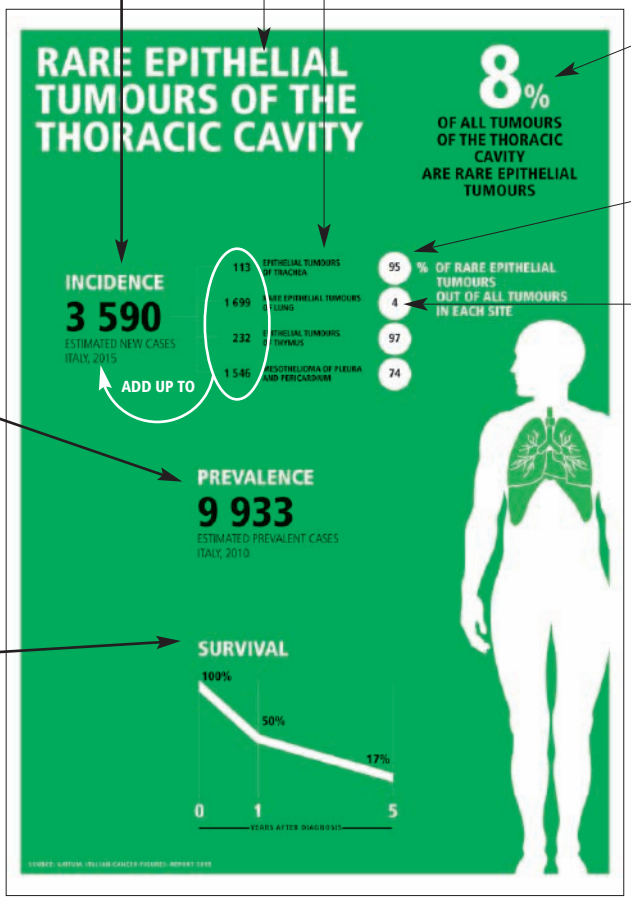
PREVALENCE (OF THE MAJOR GROUP)
Estimated number of Italians (M+F) diagnosed with a rare epithelial tumour of the thoracic cavity who were alive in 2010 (date of the last reliable estimate), regardless of time since diagnosis.

SURVIVAL (OF THE MAJOR GROUP)
Proportion of Italians (M+F) still alive 1 and 5 years after receiving a diagnosis of a rare epithelial tumour of the thoracic cavity. These figures refer to diagnoses delivered in the period 2000-2008 and are calculated net of deaths from other causes.

Are these actually registered cases? No, these are estimated cases. The AIRTUM Network covers 52% of the Italian population, which is why we have to use statistics to estimate the number of cases at the national level.

MAJOR GROUP

TIER 1



Percentage of rare epithelial tumours of the thoracic cavity out of all tumours (common+rare) of the thoracic cavity.

In the white circle: percentage of rare epithelial tumours out of all tumours (common+rare) of a tier 1.

FOR EXAMPLE:
the 1,699 rare epithelial tumours expected in Italy in 2015 represent only 4% of all the tumours of the lung (common+rare) which are diagnosed in the country in the same period.

Note: this percentage was calculated on the AIRTUM pool cases observed in 2000-2010, see incidence table next page.

GLOSSARIO E GUIDA IN ITALIANO SONO DISPONIBILI ON-LINE.

GLOSSARY AVAILABLE ON-LINE

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AN EXAMPLE
RARE EPITHELIAL TUMOURS OF THE THORACIC CAVITY

The **CRUDE INCIDENCE RATE** is the ratio between the number of new cases arising in a specific population and the total number of subjects (males and females) at risk during the same period. Usually it is expressed as annual rate per 100,000 subjects.

* **E.g., large cell carcinoma of the lung: Crude rate = 1.84 per 100,000/year means that in the areas covered by AIRTUM less than 2 persons out of 100,000 develop this kind of cancer.**

Note: even in the case of gender-specific tumours – such as male and female genital tumours – the denominator of the ratio is the resident M+F population.

The **CONFIDENCE INTERVAL (CI)** is the measure of the uncertainty of the figures we present. The larger the interval, the more uncertain the figures. When you read the value of an indicator we suggest you also look at the CI in which the value is included.

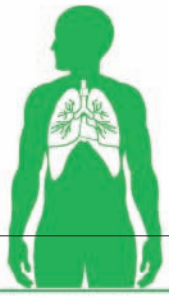
* **E.g., incidence rate 1.84, CI:1.78-1.89. This CI is narrow, showing reliability of the provided estimation.**

Note: confidence intervals equal to 0.00-0.00 mean that both lower and upper boundaries are smaller than 0.01 (e.g., 0.001-0.004), therefore when they are rounded to two decimal places they become 0.00.

The **CRUDE INCIDENCE RATE** was calculated for populations divided into gender groups (M-F) and age groups.

AGE GROUPS are wider than usual because of the rarity of the tumours and are the same for all the major groups of tumours except for embryonal tumours, where special attention was paid to children and young people.

Note: the sex- and age-specific rates for cancers with less than 15 observed cases between 2000 and 2010 were considered not estimable (NE).



INCIDENCE

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

	AIRTUM POOL (period of diagnosis 2000-2010)														ITALY
					SEX				AGE						
					MALE		FEMALE		0-54 yrs		55-64 yrs		65+ yrs		
	RATE	95% CI	OBSERVED CASES (No.)	RARE CANCERS BY SITE (%)	RATE	95% CI	RATE	95% CI	RATE	95% CI	RATE	95% CI	RATE	95% CI	ESTIMATED NEW CASES 2015
RARE EPITHELIAL TUMOURS OF THE THORACIC CAVITY	**5.42**	**5.33-5.52**	**13 027**	**8%**	**8.57**	**8.39-8.74**	**2.48**	**2.39-2.57**	**0.87**	**0.82-0.92**	**10.14**	**9.77-10.53**	**18.08**	**17.69-18.49**	**3 590**
EPITHELIAL TUMOURS OF TRACHEA	0.17	0.15-0.19	374	95%	0.27	0.24-0.30	0.07	0.06-0.09	0.03	0.02-0.04	0.33	0.27-0.41	0.55	0.48-0.62	113
Squamous cell carcinoma with variants of trachea	0.08	0.07-0.09	175		0.14	0.11-0.16	0.03	0.02-0.04	0.01	0.01-0.02	0.16	0.12-0.22	0.26	0.21-0.31	53
Adenocarcinoma with variants of trachea	0.03	0.02-0.04	64		0.05	0.04-0.06	0.01	0.01-0.02	<0.01	0.00-0.01	0.08	0.05-0.12	0.08	0.06-0.11	19
Salivary gland type tumours of trachea	0.01	0.01-0.02	26		0.01	0.01-0.02	0.01	0.01-0.02	<0.01	0.00-0.01	0.02	0.01-0.04	0.03	0.02-0.05	8
RARE EPITHELIAL TUMOURS OF LUNG	2.58	2.51-2.65	5 722	4%	4.37	4.24-4.49	0.91	0.85-0.96	0.40	0.36-0.43	4.97	4.71-5.25	8.57	8.30-8.85	1 699
Adenosquamous carcinoma of lung	0.41	0.38-0.44	909		0.66	0.61-0.71	0.18	0.15-0.20	0.06	0.05-0.08	0.76	0.66-0.87	1.39	1.28-1.50	268
Large cell carcinoma of lung	*1.84	*1.78-1.89	4 071		3.18	3.07-3.29	0.58	0.54-0.62	0.26	0.23-0.28	3.53	3.31-3.76	6.20	5.97-6.43	1 213
Salivary gland type tumours of lung	0.06	0.05-0.07	140		0.09	0.07-0.11	0.04	0.03-0.05	0.03	0.02-0.04	0.12	0.09-0.17	0.15	0.11-0.19	41
Sarcomatoid carcinoma of lung	0.27	0.25-0.29	602		0.44	0.40-0.48	0.11	0.09-0.13	0.05	0.04-0.06	0.56	0.48-0.66	0.85	0.76-0.94	177
EPITHELIAL TUMOURS OF THYMUS	0.36	0.34-0.39	804	97%	0.41	0.38-0.45	0.32	0.28-0.35	0.18	0.16-0.20	0.73	0.64-0.84	0.75	0.67-0.84	232
Malignant thymoma	0.28	0.25-0.30	612		0.31	0.28-0.35	0.24	0.21-0.27	0.15	0.13-0.17	0.56	0.47-0.65	0.54	0.47-0.61	175

Note:
for **RARE EPITHELIAL TUMOURS OF LUNG** the sum of the observed cases of tier 2 entities (5,722) is exactly the same as the number of observed cases of the corresponding tier 1 (5,722) because tier 1 includes only rare tier 2 entities and exclude common tier 2 entities and NOS morphologies.

For **EPITHELIAL TUMOURS OF TRACHEA** the sum (265) of the observed cases of tier 2 entities is different from the number of observed cases of the corresponding tier 1 (374) because tier 1 includes NOS histotypes. NOS histotypes are never included in tier 2 because tier 2 entities include rare cancers which, by definition, are identified by detailed morphologies.

Note:
the absolute numbers reported in these two columns are not directly comparable because the ones in the **OBSERVED CASES** column are actually registered in the area covered by the CRs, while the others are estimated cases for the whole Italian territory; the former refer to a period of 11 years, while the latter are cases expected in one year only.

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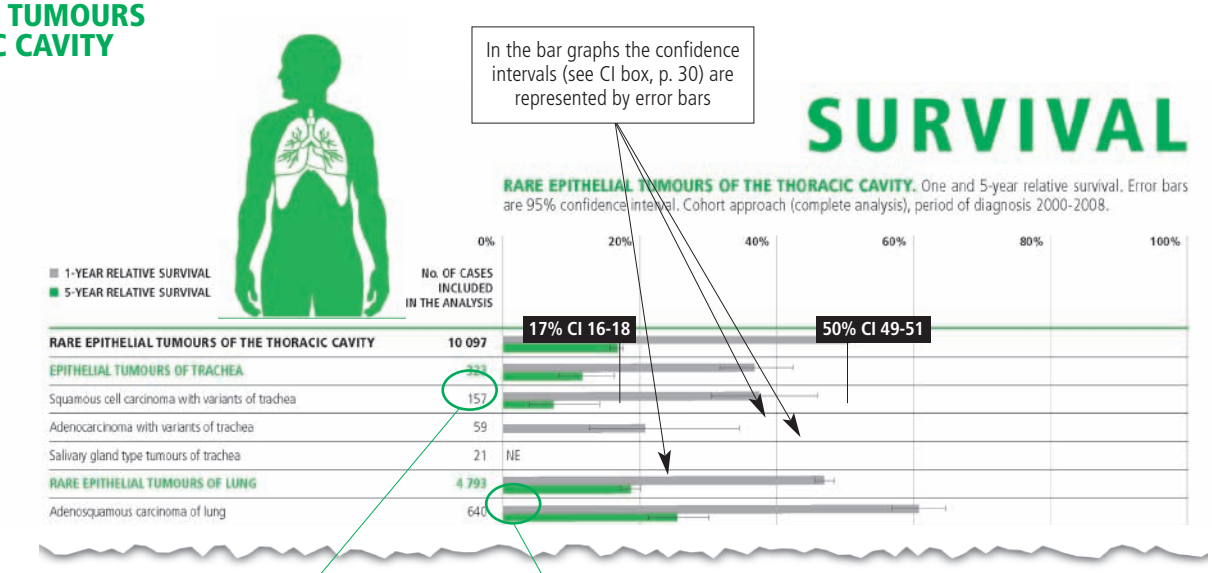
AN EXAMPLE
RARE EPITHELIAL TUMOURS OF THE THORACIC CAVITY



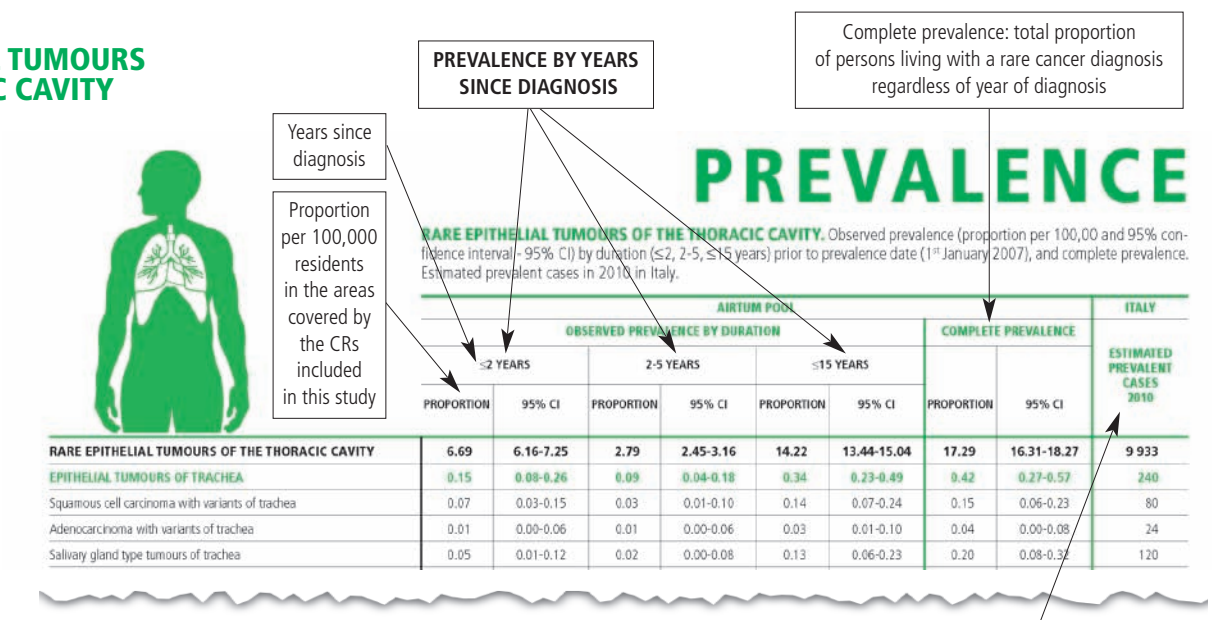
HOW TO READ THIS GRAPH (AN EXAMPLE)

For **RARE EPITHELIAL TUMOURS OF THE THORACIC CAVITY** (major group) we read: in the period 2000-2008 more than ten thousand rare epithelial tumours of the thoracic cavity were registered. One year after diagnosis **50%** of the patients were still alive; only **17%** were alive 5 years after diagnosis.

NOTE: CIs of tier 1 and 2 entities are wider than CIs of the major group because the smaller the number of analysed cases, the larger the uncertainty affecting the estimates.

In this monograph survival estimates were computed only if the number of cases was sufficient to produce reliable indicators, with interpretable CIs. That is the reason why relative survival was not computed when 30 or fewer cases were observed in the AIRTUM pool during 2000-2008. In this case the indicator is marked as not estimable (**NE** in the graph) and we suggest readers consult European survival data (www.rarecarenet.eu).

AN EXAMPLE
RARE EPITHELIAL TUMOURS OF THE THORACIC CAVITY



HOW TO READ THIS TABLE (AN EXAMPLE)

For **RARE EPITHELIAL TUMOURS OF THE THORACIC CAVITY** (major group) we read: slightly less than seven (6.69) persons out of every 100,000 residents (M+F) were alive in 2007 after receiving, in the previous **2 years**, a diagnosis of a rare epithelial tumour of the thoracic cavity. Slightly less than three (2.79) were alive in 2007 after **2-5 years** since diagnosis and fourteen (14.2) were alive in 2007 after being diagnosed in the previous **15 years**. In the **complete prevalence** column, we read how many people diagnosed with a rare epithelial tumour of the thoracic cavity out of 100,000 residents in the area covered by AIRTUM were alive in 2007, regardless of time since diagnosis.