

# Rare Thoracic Tumours

## 1. Epithelial Tumour of Trachea<sup>1</sup>

### 1.1 General Results

**Table 1. Epithelial Tumours of Trachea: Incidence, Trends, Survival**

Flemish Region 2001-2010		Incidence				Trend		Survival	
Both Sexes		R/C	N	CR	WSR	Avg Age	EAPC % p-value	Relative Survival N at risk 5yr (%)	
EPITHELIAL TUMOUR OF TRACHEA		R	73	0.12	0.12	67	-5.9 0.299	54	23.7
Squamous cell carcinoma with variants of trachea		R	53	0.09	0.09	68	-3.1 0.661	34	*
Adenocarcinoma with variants of trachea		R	9	0.01	0.01	62	* *	9	*
Salivary gland type tumours of trachea		R	5	0.01	0.01	51	* *	5	*
Males		R/C	N	CR	WSR	Avg Age	EAPC % p-value	Relative survival N at risk 5yr (%)	
EPITHELIAL TUMOUR OF TRACHEA		R	52	0.17	0.10	66	-3.9 0.647	37	17.2
Squamous cell carcinoma with variants of trachea		R	42	0.14	0.08	67	-6.4 0.405	27	*
Adenocarcinoma with variants of trachea		R	6	0.02	0.01	62	* *	6	*
Salivary gland type tumours of trachea		R	1	0.00	0.00	70	* *	1	*
Females		R/C	N	CR	WSR	Avg Age	EAPC % p-value	Relative survival N at risk 5yr (%)	
EPITHELIAL TUMOUR OF TRACHEA		R	21	0.07	0.03	68	-5.5 0.653	17	*
Squamous cell carcinoma with variants of trachea		R	11	0.04	0.01	73	* *	7	*
Adenocarcinoma with variants of trachea		R	3	0.01	0.01	62	* *	3	*
Salivary gland type tumours of trachea		R	4	0.01	0.01	46	* *	4	*

R/C: Rare or common

CR: Crude rate (N/100,000 person years)

WSR: age-standardised rate, using the world population (N/100,000 person years)

EAPC: estimated annual percentage change

RS: relative survival

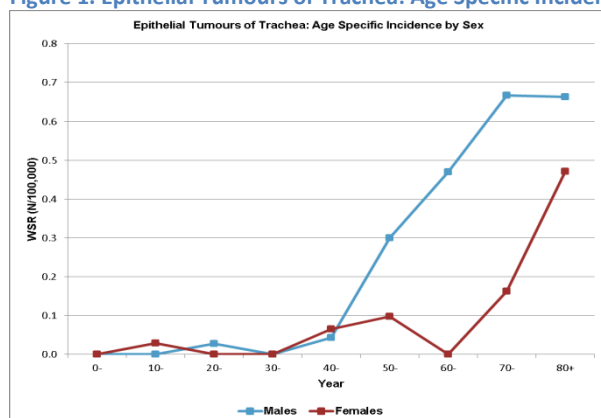
AvgAge: average age at diagnosis

### 1.2 Incidence

- 73 new epithelial tumours of the trachea are diagnosed in the Flemish Region between 2001 and 2010.
- The male/female ratio is 3.0.
- RARECARE defines three rare entities:
  - Squamous cell carcinoma represents the majority of the tracheal tumours (73%).
  - Nine adenocarcinoma of trachea are observed in the Flemish Region between 2001 and 2010.
  - Only one male and 4 females are diagnosed with a salivary gland type tumour of trachea.

<sup>1</sup> Because of the low number at risk for patients with an epithelial tumour of the trachea, only the overall survival is reported.

**Figure 1. Epithelial Tumours of Trachea: Age Specific Incidence by Sex**



- From the age of 50 years, incidence rates for epithelial tumours of trachea increase in males and females.

## 1.3 Survival

### 1.3.1 Overall Survival

**Table 2. Epithelial Tumours of Trachea – Overall Survival**

	N at risk	Observed Survival					Relative Survival				
		1 year	3 year	5 year	10 year	5 year CI	1 year	3 year	5 year	10 year	5 year CI
EPITHELIAL TUMOUR OF TRACHEA	54	44.4	27.7	21.6	-	[11.6 ; 33.4]	45.5	29.4	23.7	-	[12.9 ; 36.7]
Squamous cell carcinoma with variants	34	*	*	*	*	*	*	*	*	*	*
Adenocarcinoma with variants	9	*	*	*	*	*	*	*	*	*	*
Salivary gland type tumours	5	*	*	*	*	*	*	*	*	*	*

- Prognosis of patients diagnosed with an epithelial tumour of the trachea is bad with less than half of the patients surviving the first year, and a 5-year relative survival of only 23.7%.

### 1.3.2 Survival by Sex

**Table 3. Epithelial Tumours of Trachea – Survival by Sex**

Males	N at risk	Observed Survival				Relative Survival			
		1 year	3 year	5 year	5 year CI	1 year	3 year	5 year	5 year CI
EPITHELIAL TUMOUR OF TRACHEA	37	40.5	18.5	15.4	[5.9 ; 29.1]	41.6	19.8	17.2	[6.6 ; 32.3]
Squamous cell carcinoma with variants	27	*	*	*	*	*	*	*	*
Adenocarcinoma with variants	6	*	*	*	*	*	*	*	*
Salivary gland type tumours	1	*	*	*	*	*	*	*	*

Females	N at risk	Observed Survival				Relative Survival			
		1 year	3 year	5 year	5 year CI	1 year	3 year	5 year	5 year CI
EPITHELIAL TUMOUR OF TRACHEA	17	*	*	*	*	*	*	*	*
Squamous cell carcinoma with variants	7	*	*	*	*	*	*	*	*
Adenocarcinoma with variants	3	*	*	*	*	*	*	*	*
Salivary gland type tumours	4	*	*	*	*	*	*	*	*

## 2. Epithelial Tumours of Lung

### 2.1 General Results

Table 4. Epithelial Tumours of Lung: Incidence, Trends, Survival

Flemish Region 2001-2010						Trend		Survival	
Both Sexes						EAPC		Relative Survival	
	R/C	N	CR	WSR	Avg Age	%	p-value	N at risk	5yr (%)
EPITHELIAL TUMOUR OF LUNG	C	40,631	66.83	48.91	68	0.7	0.032	35,619	15.3
Squamous cell carcinoma with variants of lung	C	10,484	17.24	12.22	70	0.7	0.256	8,953	19.1
Adenocarcinoma with variants of lung	C	12,775	21.01	16.19	66	4.9	<0.001	11,228	15.3
Large cell carcinoma of lung	R	2,394	3.94	2.88	68	-3.4	0.006	3,240	14.5
Well differentiated endocrine carcinoma of lung	R	550	0.90	0.72	62	4.2	0.049	481	61.4
Poorly differentiated endocrine carcinoma of lung	C	7,991	13.14	9.80	68	3.1	0.008	7,100	6.7
Bronchiolo-alveolar carcinoma of lung	R	587	0.97	0.72	67	0.6	0.784	483	38.4
Salivary gland type tumours of lung	R	62	0.10	0.08	62	-1.6	0.609	57	35.5
Sarcomatoid carcinoma of lung	R	129	0.21	0.16	65	-2.1	0.551	116	24.1
Undifferentiated carcinoma of lung	R	659	1.08	0.80	68	-22.4	<0.001	591	9.4
Males						EAPC		Relative survival	
	R/C	N	CR	WSR	Avg Age	%	p-value	N at risk	5yr (%)
EPITHELIAL TUMOUR OF LUNG	C	31,868	106.27	55.03	69	-0.9	0.015	27,799	14.1
Squamous cell carcinoma with variants of lung	C	9,266	30.90	15.54	70	-0.3	0.660	7,928	18.8
Adenocarcinoma with variants of lung	C	9,072	30.25	16.59	67	3.5	0.002	7,903	14.0
Large cell carcinoma of lung	C	3,066	10.22	5.25	69	-9.8	<0.001	2,715	14.6
Well differentiated endocrine carcinoma of lung	R	281	0.94	0.55	63	4.8	0.239	251	48.3
Poorly differentiated endocrine carcinoma of lung	C	6,191	20.65	10.87	68	1.4	0.157	5,455	5.8
Bronchiolo-alveolar carcinoma of lung	R	350	1.17	0.64	67	-2.0	0.343	287	33.0
Salivary gland type tumours of lung	R	43	0.14	0.09	64	-5.9	0.311	39	32.7
Sarcomatoid carcinoma of lung	R	96	0.32	0.18	66	-4.7	0.234	86	24.5
Undifferentiated carcinoma of lung	R	525	1.75	0.91	69	-24.5	<0.001	466	8.9
Females						EAPC		Relative survival	
	R/C	N	CR	WSR	Avg Age	%	p-value	N at risk	5yr (%)
EPITHELIAL TUMOUR OF LUNG	C	8,763	28.44	15.17	65	5.3	<0.001	7,820	19.3
Squamous cell carcinoma with variants of lung	R	1,218	3.95	2.01	67	5.7	<0.001	1,025	20.8
Adenocarcinoma with variants of lung	C	3,703	12.02	6.74	64	7.8	<0.001	3,325	18.4
Large cell carcinoma of lung	R	580	1.88	1.01	65	-4.3	0.105	525	14.3
Well differentiated endocrine carcinoma of lung	R	269	0.87	0.54	60	4.4	0.110	230	75.7
Poorly differentiated endocrine carcinoma of lung	R	1,800	5.84	3.20	65	8.1	0.001	1,645	9.7
Bronchiolo-alveolar carcinoma of lung	R	237	0.77	0.39	66	4.9	0.085	196	46.6
Salivary gland type tumours of lung	R	19	0.06	0.04	57	2.8	0.571	18	*
Sarcomatoid carcinoma of lung	R	33	0.11	0.06	64	7.9	0.370	30	*
Undifferentiated carcinoma of lung	R	134	0.43	0.24	65	-20.4	<0.001	125	11.0

R/C: Rare or common

CR: Crude rate (N/100.000 person years)

WSR: age-standardised rate, using the world population (N/100.000 person years)

EAPC: estimated annual percentage change

RS: relative survival

AvgAge: average age at diagnosis

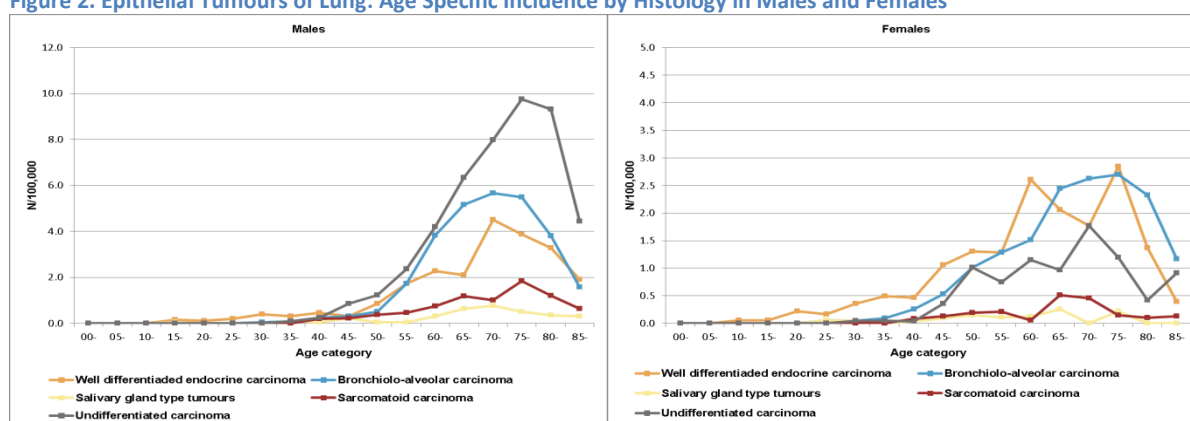
### 2.2 Incidence

- 40,631 new epithelial tumours of the lung are diagnosed in the Flemish Region between 2001 and 2010.
- The male/female ratio is 3.6.
- RARECARE defines nine different entities of which three are common and six rare:
  - The common subtypes represent about 75% of all lung cancers and are: squamous cell carcinoma, adenocarcinoma and poorly differentiated.
  - Large cell carcinoma, although considered rare by RARECARE, is a common malignancy in the Flemish Region. Care must be taken in the interpretation, since the

RARECARE entity includes some codes (8071;8072) that we would classify as squamous cell carcinoma. On the other hand, the code for non-small cell carcinoma (8046) is not included by RARECARE in this entity but is grouped with the endocrine tumours.

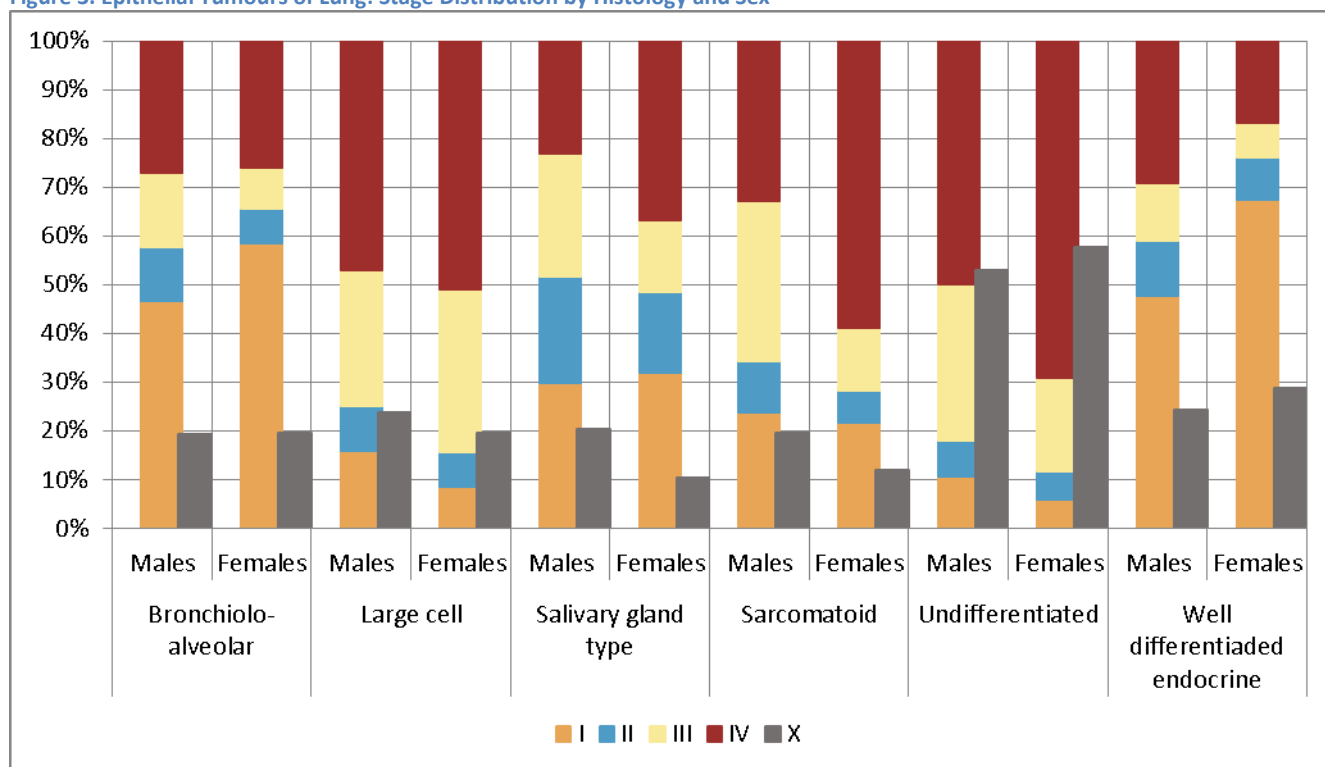
- Well differentiated endocrine lung carcinoma represent 550 new cases between 2001 and 2010.
- Bronchiolo-alveolar carcinoma represent 587 new diagnoses.
- Salivary gland type tumours are the rarest entity with only 62 new diagnoses in the Flemish Region between 2001 and 2010.
- 129 new cases of sarcomatoid carcinoma are observed.
- Undifferentiated carcinoma accounts for 659 new lung cancer cases.

Figure 2. Epithelial Tumours of Lung: Age Specific Incidence by Histology in Males and Females



- Due to its high rates, large cell carcinoma is not represented.
- Undifferentiated carcinoma is the most 'common' rare lung cancer entity in males. From the age of 50 years, incidence rates increase. The highest incidence rates are observed around the age of 75 years.
- For females, well differentiated endocrine carcinoma is the most common rare lung cancer entity. This tumour type already occurs at an early age.
- The age specific incidence rates for bronchiolo-alveolar carcinoma start to increase around the age of 50 years.
- Salivary gland type tumours and sarcomatoid carcinoma are very rare, the highest incidence rates are observed around the age of 70-75 years.

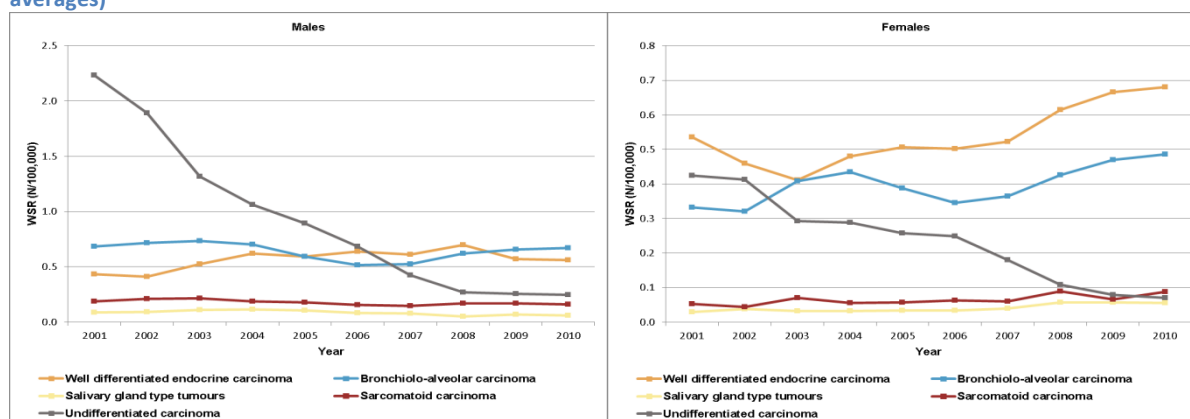
Figure 3. Epithelial Tumours of Lung: Stage Distribution by Histology and Sex



- Stage-information is available in 80% or more of all lung cancer cases. The only exception is the undifferentiated carcinoma, where stage information is missing in more than 50% of all diagnoses.
- Bronchiolo-alveolar and well differentiated endocrine carcinoma have the best prognostic stage distribution. About 50% of all new diagnoses is stage I in males; in females this increases to 60% for bronchiolo-alveolar and almost 70% for well differentiated endocrine carcinoma.
- Large cell and undifferentiated carcinoma in males and females and sarcomatoid carcinomas in females have the worst prognostic stage distribution, with more than 50% stage IV tumours.

## 2.3 Trends

Figure 4. Rare Lung Cancer Entities: Age-Standardised Incidence by Histology in Males and Females (three year moving averages)



- Large cell carcinoma (not represented due to the high incidence rates) and undifferentiated carcinoma show very large decreasing trends in both sexes. This decrease is largely due to more specifications in registration practices.
- Well differentiated carcinoma seems to increase in both sexes.
- Salivary gland type tumours, bronchiolo-alveolar carcinoma and sarcomatoid carcinoma tend to decrease in males and increase in females, however none of these trends are significant.

## 2.4 Survival

### 2.4.1 Overall Survival

Table 5. Epithelial Tumours of Lung – Overall Survival

	N at risk	Observed Survival					Relative Survival				
		1 year	3 year	5 year	10 year	5 year CI	1 year	3 year	5 year	10 year	5 year CI
EPITHELIAL TUMOURS OF LUNG	35,619	42.2	18.6	13.3	7.6	[12.9 ; 13.7]	43.5	20.2	15.3	10.3	[14.8 ; 15.7]
Squamous cell carcinoma with variants	8,953	47.3	22.4	16.2	8.6	[15.4 ; 17.1]	48.9	24.6	19.1	12.2	[18.1 ; 20.0]
Adenocarcinoma with variants	11,228	45.3	20.2	13.8	7.8	[13.1 ; 14.5]	46.3	21.5	15.3	9.9	[14.6 ; 16.1]
Large cell carcinoma	3,240	39.0	16.9	12.4	7.0	[11.2 ; 13.6]	40.3	18.5	14.5	10.1	[13.2 ; 15.9]
Well differentiated endocrine carcinoma	481	73.8	61.3	56.7	50.7	[51.9 ; 61.2]	75.1	64.3	61.4	61.9	[56.2 ; 66.3]
Poorly differentiated endocrine carcinoma	7,100	33.3	8.8	6.0	2.8	[5.4 ; 6.6]	34.1	9.4	6.7	3.6	[6.0 ; 7.4]
Bronchiolo-alveolar carcinoma	483	69.8	44.3	34.2	22.0	[29.6 ; 38.8]	71.4	47.4	38.4	28.8	[33.3 ; 43.6]
Salivary gland type tumours	57	61.4	49.0	32.7	23.5	[19.4 ; 46.6]	62.5	51.2	35.5	26.7	[21.1 ; 50.6]
Sarcomatoid carcinoma	116	41.4	30.1	21.6	9.2	[14.4 ; 29.9]	42.3	32.0	24.1	14.5	[16.0 ; 33.3]
Undifferentiated carcinoma	591	30.3	11.4	8.0	5.5	[6.0 ; 10.4]	31.4	12.6	9.4	7.8	[7.0 ; 12.21]

- Epithelial tumours of the lung can in general be considered as tumours with a very low survival that steeply declines from diagnosis, with a 1-year relative survival of only 43.5% and a 3-year relative survival of 20.2%. Thereafter, survival decreases less steeply to reach a 10-year relative survival of 10.3%.
- Although almost all subtypes have a bad prognosis, differences can be observed. The best survival is observed for well differentiated endocrine carcinoma with a 5-year relative survival of 61.4%. Poorly differentiated endocrine carcinoma and undifferentiated carcinoma on the other hand, have the worst survival. In the group of the poorly differentiated endocrine carcinoma, only one third of the patients survive the first year and less than 10% are still alive at three years after diagnosis.

## 2.4.2 Survival by Sex

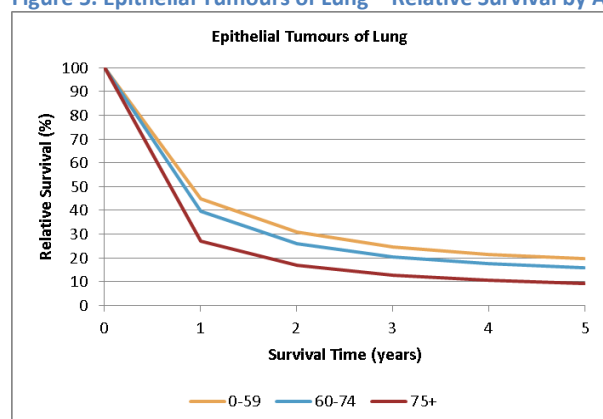
Table 6. Epithelial Tumours of Lung – Survival by Sex

Males	N	Observed Survival					Relative Survival			
	at risk	1 year	3 year	5 year	5 year CI	1 year	3 year	5 year	5 year CI	
EPITHELIAL TUMOURS OF LUNG	27,799	40.5	17.1	12.0	[11.6 ; 12.4]	41.9	18.9	14.1	[13.6 ; 14.6]	
Squamous cell carcinoma with variants	7,928	47.4	22.0	15.9	[15.0 ; 16.8]	49.1	24.3	18.8	[17.8 ; 19.9]	
Adenocarcinoma with variants	7,903	42.4	18.1	12.3	[11.6 ; 13.1]	43.5	19.6	14.0	[13.2 ; 14.9]	
Large cell carcinoma	2,715	39.3	16.9	12.2	[11.0 ; 13.5]	40.6	18.7	14.6	[13.1 ; 16.1]	
Well differentiated endocrine carcinoma	251	64.1	49.6	43.9	[37.4 ; 50.3]	65.6	52.6	48.3	[41.1 ; 55.3]	
Poorly differentiated endocrine carcinoma	5,455	30.8	7.6	5.0	[4.4 ; 5.7]	31.8	8.3	5.8	[5.1 ; 6.5]	
Bronchiolo-alveolar carcinoma	287	64.5	39.5	29.0	[23.5 ; 34.7]	66.2	42.6	33.0	[26.7 ; 39.5]	
Salivary gland type tumours	39	53.9	40.9	29.3	[14.9 ; 45.4]	55.2	43.4	32.7	[16.5 ; 50.7]	
Sarcomatoid carcinoma	86	41.9	31.4	21.7	[13.4 ; 31.3]	42.9	33.6	24.5	[15.1 ; 35.3]	
Undifferentiated carcinoma	466	30.7	11.2	7.4	[5.2 ; 10.0]	31.9	12.6	8.9	[6.3 ; 12.1]	
Females	N	Observed Survival				Relative Survival				
	at risk	1 year	3 year	5 year	5 year CI	1 year	3 year	5 year	5 year CI	
EPITHELIAL TUMOURS OF LUNG	7,820	48.4	23.7	17.8	[16.9 ; 18.7]	49.2	24.8	19.3	[18.3 ; 20.3]	
Squamous cell carcinoma with variants	1,025	46.2	25.2	19.1	[16.6 ; 21.8]	47.0	26.4	20.8	[18.0 ; 23.7]	
Adenocarcinoma with variants	3,325	52.1	25.1	17.2	[15.8 ; 18.6]	52.8	26.1	18.4	[16.9 ; 19.9]	
Large cell carcinoma	525	37.7	16.9	13.0	[10.2 ; 16.2]	38.3	17.8	14.3	[11.2 ; 17.8]	
Well differentiated endocrine carcinoma	230	84.4	74.3	70.7	[64.1 ; 76.4]	85.4	77.0	75.7	[68.6 ; 81.7]	
Poorly differentiated endocrine carcinoma	1,645	41.4	12.6	9.1	[7.6 ; 10.7]	41.9	13.1	9.7	[8.1 ; 11.4]	
Bronchiolo-alveolar carcinoma	196	77.6	51.3	42.1	[34.4 ; 49.6]	79.1	54.4	46.6	[38.1 ; 54.9]	
Salivary gland type tumours	18	*	*	*	*	*	*	*	*	
Sarcomatoid carcinoma	30	*	*	*	*	*	*	*	*	
Undifferentiated carcinoma	125	28.8	11.9	10.2	[5.7 ; 16.3]	29.3	12.4	11.0	[6.1 ; 17.5]	

- For almost all subtypes at all observed time points, survival is better for females than for males.
- The sex difference is most pronounced for the well differentiated endocrine carcinoma (5-year relative survival in males: 48.3% versus 75.7% in females) and bronchiolo-alveolar carcinoma (5-year relative survival in males: 33.0% versus 46.6% in females).

## 2.4.3 Survival by Age Group<sup>2</sup>

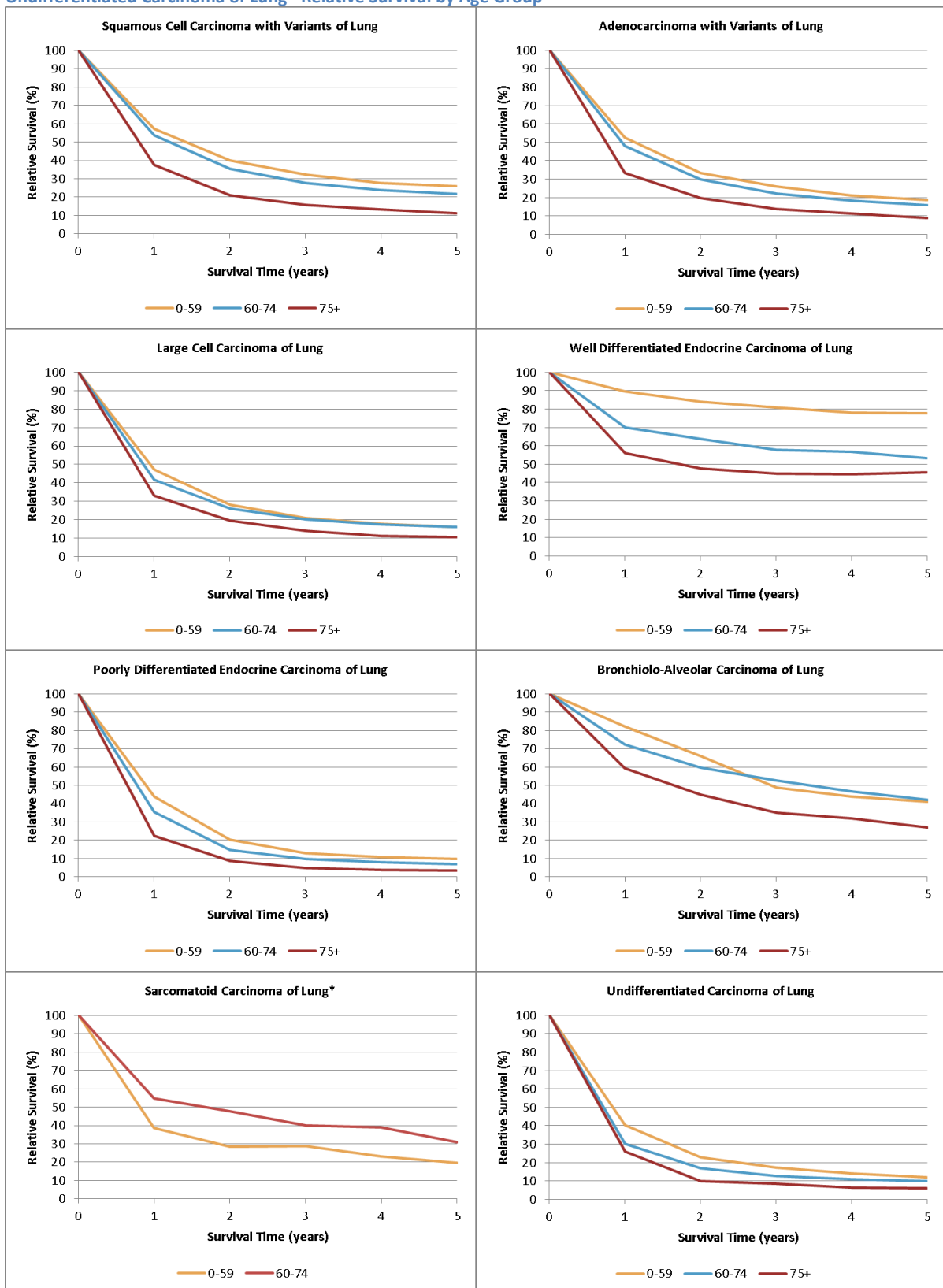
Figure 5. Epithelial Tumours of Lung – Relative Survival by Age Group



- Although prognosis is poor for all age groups, survival is inversely related with the age of the patient at diagnosis, with the highest survival for patients in the age group 0-59 years old (5-year relative survival: 19.8%) and the lowest in the age group 75+ years old (5-year relative survival: 9.1%).

<sup>2</sup> Survival by age group is not displayed for salivary-gland type tumours of lung because none of the age groups have a number at risk higher than 35.

Figure 6. Squamous Cell Carcinoma, Adenocarcinoma, Large Cell Carcinoma, Well Differentiated Endocrine Carcinoma, Poorly Differentiated Endocrine Carcinoma, Bronchiolo-Alveolar Carcinoma, Sarcomatoid Carcinoma and Undifferentiated Carcinoma of Lung—Relative Survival by Age Group

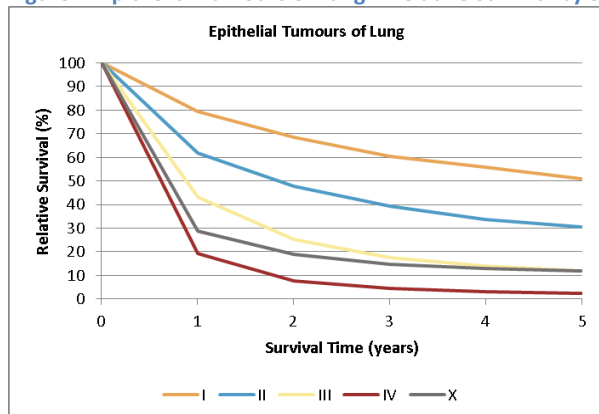


\* Survival for the oldest age group (75+) is not shown because the number at risk is lower than 35.

- for the subtypes squamous cell carcinoma, adenocarcinoma, poorly differentiated endocrine carcinoma and undifferentiated carcinoma, the difference in survival between the different age groups is similar to the results described for all epithelial tumours of the lung together.
- For the large cell carcinoma, no age difference can be observed between the age groups 0-59 years and 60-74 years, although patients aged 75 and above at diagnosis have a worse prognosis.
- Differences in survival by age groups are larger for well differentiated endocrine carcinoma. Especially the youngest age group (0-59 years) has a much better survival rate (5-year relative survival: 77.6%) than the older age groups (5-year relative survival rate: 53.3% for age group 60-74 years and 45.5% for age groups 75+ years).
- For bronchiolo-alveolar carcinoma, survival is similar for the youngest and middle age group from 3 years after diagnosis onwards (5-year relative survival: 42.5% for the age group 0-59 years and 42.7% for the age group 60-74 years), but is much worse for the oldest age group (5-year relative survival: 29.1%).
- Contrary to most cancer types, survival is better for the middle age group than for the youngest age group for sarcomatoid carcinoma at all observation periods.

#### 2.4.4 Survival by Stage<sup>3</sup>

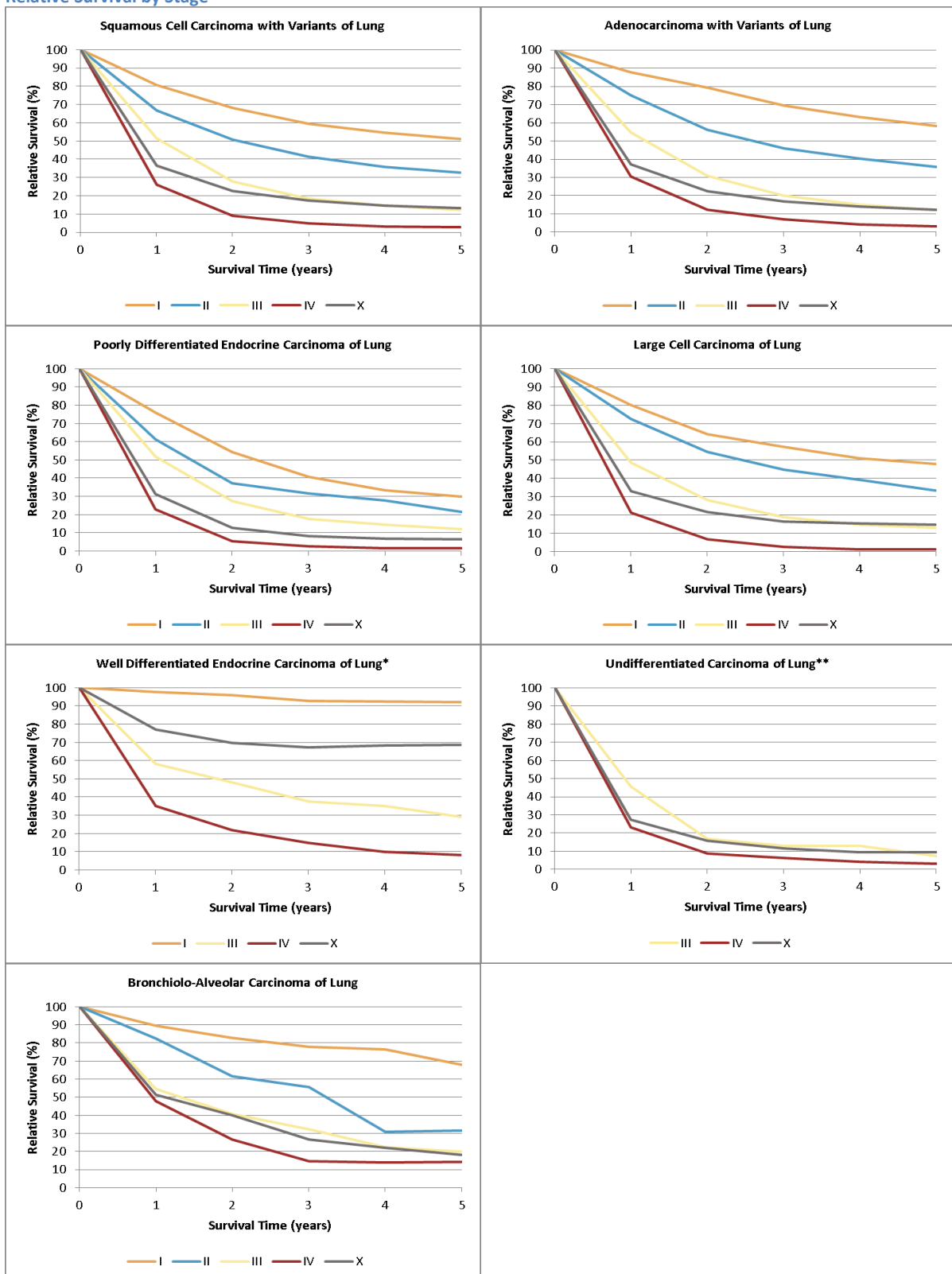
Figure 7. Epithelial Tumours of Lung – Relative Survival by Stage



- Patients with a stage I epithelial tumour of the lung have a clearly better prognosis (5-year relative survival of 51.1%) than patients diagnosed with a more advanced disease stage.
- Prognosis is very poor for patients diagnosed with a stage III or IV tumour. Already one year after diagnosis, relative survival has dropped to 43.3% for stage III and 19.3% for stage IV. Thereafter, survival continues to decrease to reach a 5-year relative survival of 11.8% for stage III and 2.5% for stage IV.

<sup>3</sup> Survival by stage is not displayed for the salivary-gland type tumours (all stages number at risk lower than 35) and the sarcomatoid carcinoma (only stage IV with a number at risk higher than 35).

Figure 8. Squamous Cell Carcinoma, Adenocarcinoma, Poorly Differentiated Endocrine Carcinoma, Large Cell Carcinoma, Well Differentiated Endocrine Carcinoma, Undifferentiated Carcinoma and Bronchiolo-Alveolar Carcinoma of Lung – Relative Survival by Stage



\* Stage II is not shown because the number at risk is lower than 35.

\*\* Stage I and II are not shown because the numbers at risk are lower than 35.

- For adenocarcinoma with variants of the lung, survival is better for the lower stages (5-year relative survival for stage I: 58.3% and stage II: 35.8%) than the earlier described survival of all epithelial tumours of the lung together.
- Well differentiated endocrine carcinoma, especially the stage I (5-year relative survival: 91.7%) and stage III tumours (5-year relative survival: 28.9%) have a better prognosis than all other lung cancers of the same stage.
- On the contrary, poorly differentiated endocrine carcinoma has a poor survival for all stages, with a 5-year relative survival for stage I tumours of only 29.8% and for stage IV tumours under 10% after two years of follow-up.
- The bronchiolo-alveolar carcinoma has a better survival for stage I than is usually seen for lung tumours, with a 5-year relative survival of 67.8%.

### 3. Epithelial Tumours of Thymus<sup>4</sup>

#### 3.1 General Results

**Table 7. Epithelial Tumours of Thymus: Incidence, Trends, Survival**

Flemish Region 2001-2010						Trend		Survival	
Both Sexes	R/C	Incidence				EAPC		Relative Survival	
		N	CR	WSR	Avg Age	%	p-value	N at risk	5yr (%)
EPITHELIAL TUMOURS OF THYMUS	R	144	0.24	0.14	62	-3.8	0.279	126	72.6
Malignant thymoma	R	117	0.19	0.12	62	-3.3	0.425	104	77.8
Squamous cell carcinoma of thymus	R	10	0.02	0.01	63	*	*	9	*
Undifferentiated carcinoma of thymus	R	2	0.00	0.00	74	*	*	2	*
Lymphoepithelial carcinoma of thymus	R	0	-	-	-	-	-	0	-
Adenocarcinoma with variants of thymus	R	5	0.01	0.00	66	*	*	2	*
<b>Males</b>									
	R/C	Incidence				EAPC		Relative survival	
		N	CR	WSR	Avg Age	%	p-value	N at risk	5yr (%)
EPITHELIAL TUMOURS OF THYMUS	R	83	0.28	0.16	62	-0.4	0.948	71	73.3
Malignant thymoma	R	67	0.22	0.13	62	5.5	0.424	58	76.5
Squamous cell carcinoma of thymus	R	5	0.02	0.01	61	*	*	5	*
Undifferentiated carcinoma of thymus	R	1	0.00	0.00	88	*	*	1	*
Lymphoepithelial carcinoma of thymus	R	0	-	-	-	-	-	0	-
Adenocarcinoma with variants of thymus	R	3	0.01	0.01	67	*	*	1	*
<b>Females</b>									
	R/C	Incidence				EAPC		Relative survival	
		N	CR	WSR	Avg Age	%	p-value	N at risk	5yr (%)
EPITHELIAL TUMOURS OF THYMUS	R	61	0.20	0.12	62	-7.3	0.031	55	71.6
Malignant thymoma	R	50	0.16	0.10	62	-9.5	0.059	46	79.1
Squamous cell carcinoma of thymus	R	5	0.02	0.01	65	*	*	4	*
Undifferentiated carcinoma of thymus	R	1	0.00	0.00	60	*	*	1	*
Lymphoepithelial carcinoma of thymus	R	0	-	-	-	-	-	0	-
Adenocarcinoma with variants of thymus	R	2	0.01	0.00	64	*	*	1	*

R/C: Rare or common

CR: Crude rate (N/100,000 person years)

WSR: age-standardised rate, using the world population (N/100,000 person years)

EAPC: estimated annual percentage change

RS: relative survival

AvgAge: average age at diagnosis

<sup>4</sup> Survival by stage is not reported for the epithelial tumours of the thymus because staging is not possible according to the TNM rules.

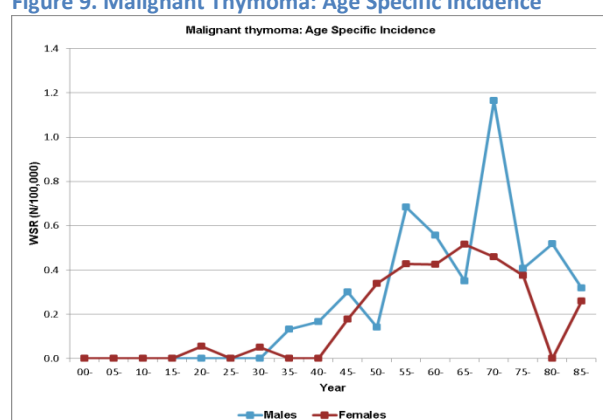
## 3.2 Incidence

- 144 new epithelial tumours of the thymus are diagnosed in the Flemish Region between 2001 and 2010.
- The male/female ratio is 1.4.
- RARECARE defines five rare entities:
  - The majority of thymic tumours are thymoma (81%). Type B thymoma is most commonly observed.
  - Squamous cell carcinoma represents only 10 cases.
  - One undifferentiated carcinoma in males and one in females is observed in the Flemish Region between 2001 and 2010.
  - Lymphoepithelial carcinoma is not observed.
  - Only 5 adenocarcinoma of thymus are registered.

Table 8. Epithelial Tumours of Thymus: Histological Distribution

Flemish Region 2001-2010	Males	Females
EPITHELIAL TUMOURS OF THYMUS	83	61
Malignant thymoma	67	50
Thymoma, malignant, NOS	27	19
Malignant thymoma, type AB	7	4
Malignant thymoma, type A	5	3
Malignant thymoma, type B	17	17
Malignant thymoma, type C	11	7
Squamous cell carcinoma of thymus	5	5
Undifferentiated carcinoma of thymus	1	1
Lymphoepithelial carcinoma of thymus	-	-
Adenocarcinoma with variants of thymus	3	2
Adenocarcinoma NOS	2	1
Papillary adenocarcinoma, NOS	-	1

Figure 9. Malignant Thymoma: Age Specific Incidence



- Incidence rates for malignant thymoma are comparable between males and females.

## 3.3 Survival

### 3.3.1 Overall Survival

Table 9. Epithelial Tumours of Thymus – Overall Survival

	N at risk	Observed Survival					Relative Survival				
		1 year	3 year	5 year	10 year	5 year CI	1 year	3 year	5 year	10 year	5 year CI
EPITHELIAL TUMOURS OF THYMUS	126	88.9	68.8	65.7	52.0	[56.3 ; 73.5]	90.6	72.9	72.6	66.9	[62.2 ; 81.3]
Malignant thymoma	104	92.3	74.8	71.1	58.6	[60.8 ; 79.1]	93.9	79.0	77.8	73.4	[66.5 ; 86.6]
Squamous cell carcinoma	9	*	*	*	*	*	*	*	*	*	*
Undifferentiated carcinoma	2	*	*	*	*	*	*	*	*	*	*
Lymphoepithelial carcinoma	0	-	-	-	-	-	-	-	-	-	-
Adenocarcinoma with variants	2	*	*	*	*	*	*	*	*	*	*

- Survival is rather good for patients diagnosed with an epithelial tumour of thymus, with a 1-year relative survival of 90.6% and a 5-year relative survival of 72.6%.
- Although the majority of patients with an epithelial tumour of the thymus are diagnosed with a malignant thymoma, survival of this subtype is somewhat higher than survival of all epithelial tumours of the thymus together.

### 3.3.2 Survival by Sex

Table 10. Epithelial Tumours of Thymus – Survival by Sex

Males	N at risk	Observed Survival					Relative Survival				
		1 year	3 year	5 year	5 year CI		1 year	3 year	5 year	5 year CI	
EPITHELIAL TUMOURS OF THYMUS	71	87.3	70.0	64.4	[51.5 ; 74.7]		89.5	75.4	73.3	[58.7 ; 85.0]	
Malignant thymoma	58	89.7	75.4	68.4	[53.9 ; 79.2]		91.5	80.4	76.5	[60.2 ; 88.5]	
Squamous cell carcinoma	5	*	*	*	*	*	*	*	*	*	*
Undifferentiated carcinoma	1	*	*	*	*	*	*	*	*	*	*
Lymphoepithelial carcinoma	0	-	-	-	-	-	-	-	-	-	-
Adenocarcinoma with variants	1	*	*	*	*	*	*	*	*	*	*

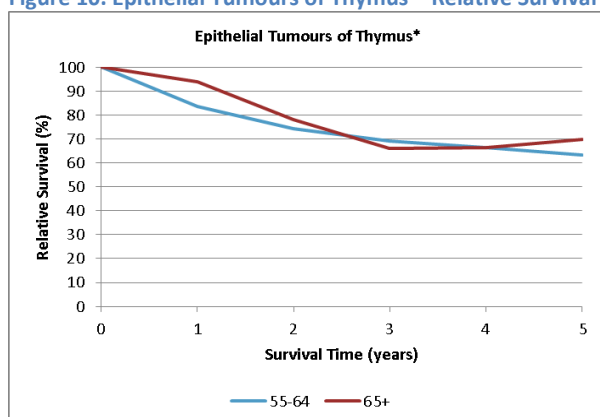
  

Females	N at risk	Observed Survival					Relative Survival				
		1 year	3 year	5 year	5 year CI		1 year	3 year	5 year	5 year CI	
EPITHELIAL TUMOURS OF THYMUS	55	90.9	67.2	67.2	[52.5 ; 78.2]		92.2	69.9	71.6	[55.9 ; 83.5]	
Malignant thymoma	46	95.7	74.1	74.1	[58.0 ; 84.8]		97.0	77.2	79.1	[61.8 ; 90.6]	
Squamous cell carcinoma	4	*	*	*	*	*	*	*	*	*	*
Undifferentiated carcinoma	1	*	*	*	*	*	*	*	*	*	*
Lymphoepithelial carcinoma	0	-	-	-	-	-	-	-	-	-	-
Adenocarcinoma with variants	1	*	*	*	*	*	*	*	*	*	*

- Survival rates for epithelial tumours of thymus are very comparable between both sexes.

### 3.3.3 Survival by Age Group

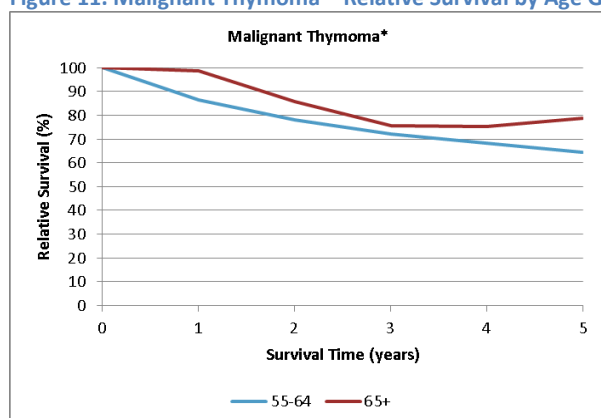
Figure 10. Epithelial Tumours of Thymus – Relative Survival by Age Group



\* Survival is not shown for patients in the age group 0-54 years old because the number at risk is lower than 35.

- Survival is similar for the age groups 55-64 years and the age group 65 years and older.

**Figure 11. Malignant Thymoma – Relative Survival by Age Group**



\* Survival is not shown for patients in the age group 0-54 years old because the number at risk is lower than 35.

- Because almost all patients with an epithelial tumour of the thymus are diagnosed with a malignant thymoma, survival by age group is similar to the earlier described survival of all epithelial tumours of the thymus together. However, the 5-year relative survival is almost 15% higher in the oldest group than in the age group 55-64 years.

## 4. Malignant Mesothelioma

### 4.1 General Results

**Table 11. Epithelial Tumours of Mesothelioma: Incidence, Trends, Survival**

Flemish Region 2001-2010		Incidence				Trend		Survival	
Both Sexes		R/C	N	CR	WSR	Avg Age	EAPC % p-value	Relative Survival N at risk 5yr (%)	
MALIGNANT MESOTHELIOMA		R	1,608	2.64	1.3	69	1.1 0.433	1,427	5.1
Mesothelioma of pleura and pericardium		R	1,476	2.43	1.2	69	1.3 0.334	1,310	4.5
Mesothelioma of peritoneum and tunica vaginalis		R	116	0.19	0.1	64	1.5 0.453	104	14.0
Males		R/C	N	CR	WSR	Avg Age	EAPC % p-value	Relative survival N at risk 5yr (%)	
MALIGNANT MESOTHELIOMA		R	1,343	4.48	2.35	69	0.3 0.791	1,184	4.5
Mesothelioma of pleura and pericardium		R	1,245	4.15	2.16	69	0.8 0.512	1,098	4.2
Mesothelioma of peritoneum and tunica vaginalis		R	84	0.28	0.17	64	-0.9 0.756	75	9.8
Females		R/C	N	CR	WSR	Avg Age	EAPC % p-value	Relative survival N at risk 5yr (%)	
MALIGNANT MESOTHELIOMA		R	265	0.86	0.40	69	4.4 0.162	243	7.9
Mesothelioma of pleura and pericardium		R	231	0.75	0.34	69	3.4 0.322	212	5.8
Mesothelioma of peritoneum and tunica vaginalis		R	32	0.10	0.06	65	10.1 0.077	29	*

R/C: Rare or common

CR: Crude rate (N/100,000 person years)

WSR: age-standardised rate, using the world population (N/100,000 person years)

EAPC: estimated annual percentage change

RS: relative survival

AvgAge: average age at diagnosis

## 4.2 Incidence

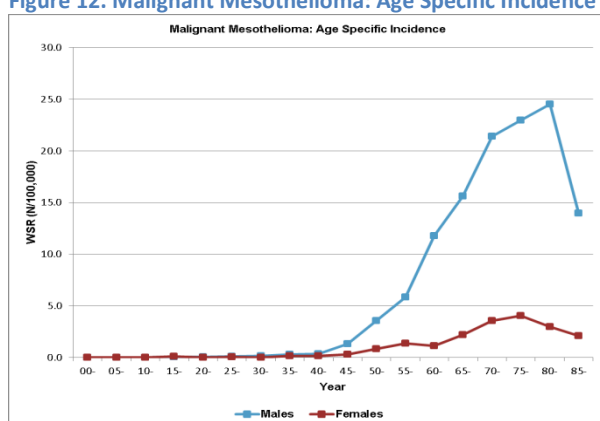
- 1.608 new malignant mesothelioma are diagnosed in the Flemish Region between 2001 and 2010.
- The male/female ratio is 5.9.
- RARECARE defines two rare entities based on the primary localisation:
  - The majority of malignant mesothelioma are of pleural and pericardial origin (92%). The male/female ratio is 6.4.
  - Only 116 mesothelioma originate from the peritoneum or tunica vaginalis. The male/female ratio is 3.0.

Table 12. Epithelial Tumours of Mesothelioma: Morphological Distribution by Sex

Flemish Region 2001-2010	Males	Females	Total
<b>MALIGNANT MESOTHELIOMA</b>	1343	265	1608
Mesothelioma of pleura and pericardium	1245	231	1476
Epithelioid malignant mesothelioma	493	92	585
Sarcomatoid malignant mesothelioma	119	15	134
Mesothelioma of peritoneum and tunica vaginalis	84	32	116
Epithelioid malignant mesothelioma	47	14	61
Sarcomatoid malignant mesothelioma	2	1	3

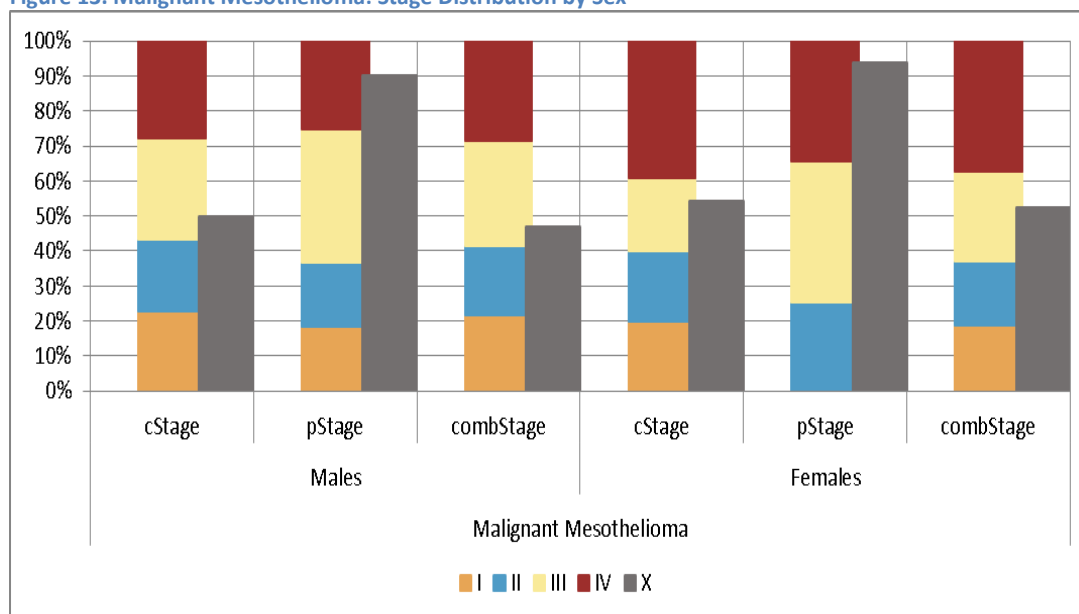
- Less than one out of ten diagnoses of pleural (or pericardial) mesothelioma is of sarcomatoid histology.
- Epithelioid histology is most frequently observed, independent of the site of origin.

Figure 12. Malignant Mesothelioma: Age Specific Incidence



- Age specific incidence rates start to increase around the age of 50 years.
- The increase in males is more pronounced than the increase in females.

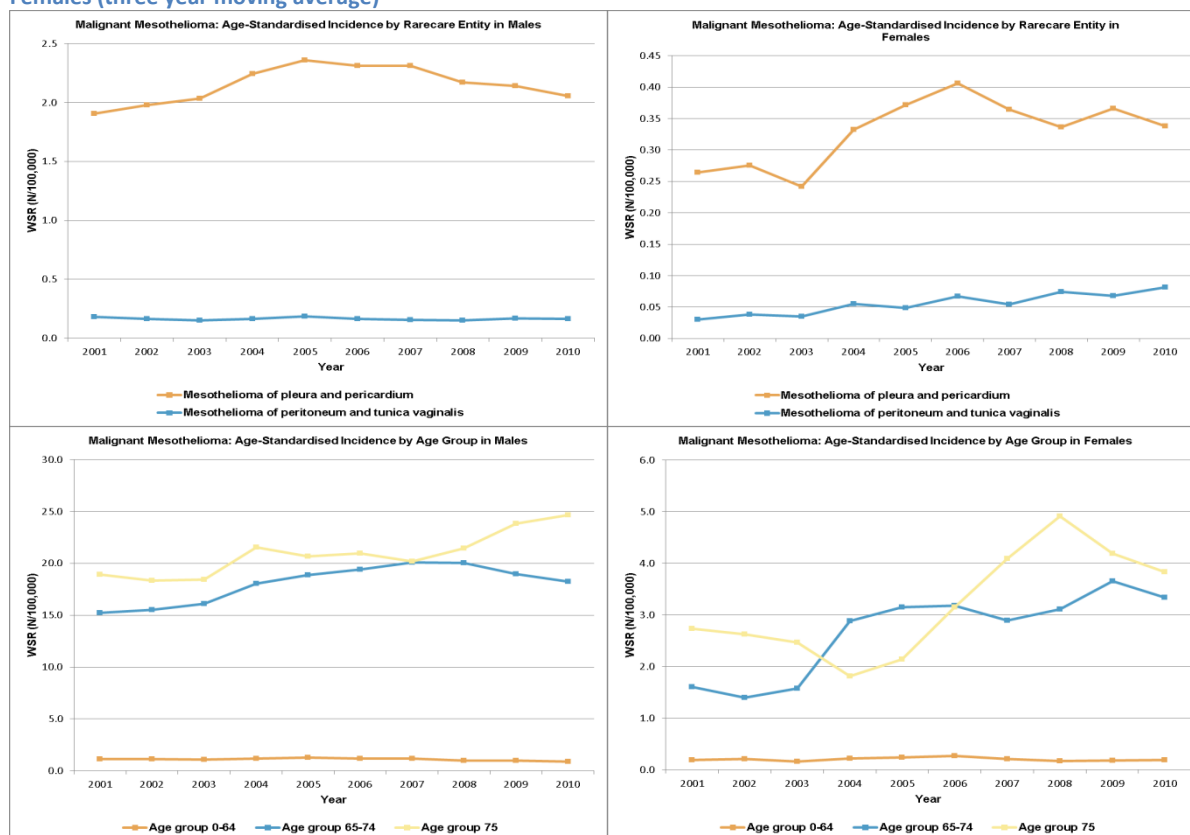
Figure 13. Malignant Mesothelioma: Stage Distribution by Sex



- Information on clinical stage is available in about half of the mesothelioma cases. Pathological staging (~90% missing) is rarely available.

### 4.3 Trends

Figure 14. Malignant Mesothelioma: Age-Standardised Incidence by RARECARE Entity and by Age Group in Males and Females (three year moving average)



- The incidence rates for mesothelioma (both subtypes) in females are increasing, however no significant trend is observed.
- In males, the rates for pleural mesothelioma increase until 2005 (EAPC 2001-2005 = 4.0% [p = 0.400]), between 2005 and 2010 they decrease (EAPC 2005-2010 = -2.7% [p = 0.079]).
- Incidence rates for mesothelioma decrease in patients younger than 65 years of age, while in the older age groups the rates are increasing, but the trends are not significant.
  - Age group 0-64 years: males: EAPC = -3.1% [p = 0.149]; females: EAPC = -0.5% [p = 0.914]
  - Age group 65-74 years: males: EAPC = 2.8% [p = 0.116]; females: EAPC = 11.6% [p = 0.077]
  - Age group 75+ years: males: EAPC = 3.4% [p = 0.064]; females: EAPC = 7.5% [p = 0.185]

## 4.4 Survival

### 4.4.1 Overall Survival

Table 13. Malignant Mesothelioma – Overall Survival

	N at risk	Observed Survival					Relative Survival				
		1 year	3 year	5 year	10 year	5 year CI	1 year	3 year	5 year	10 year	5 year CI
MALIGNANT MESOTHELIOMA	1,427	44.4	10.5	4.4	2.3	[3.3 ; 5.7]	45.7	11.4	5.1	3.1	[3.8 ; 6.6]
Mesothelioma of pleura and pericardium	1,310	44.4	9.6	3.8	1.5	[2.8 ; 5.1]	45.7	10.4	4.5	2.2	[3.3 ; 6.0]
Mesothelioma of peritoneum and tunica vaginalis	104	46.6	22.5	13.3	11.4	[6.9 ; 21.8]	47.8	23.7	14.0	12.2	[7.2 ; 23.1]

- Survival is very bad for patients diagnosed with a malignant melanoma. Less than half of the patients survives the first year after diagnosis. Four years later, 5.1% of patients are still alive.
- Although survival is poor for all localisations the malignant mesothelioma, survival is somewhat better for the peritoneum and tunica vaginalis than for the pleural and pericardial localisations.

### 4.4.2 Survival by Sex

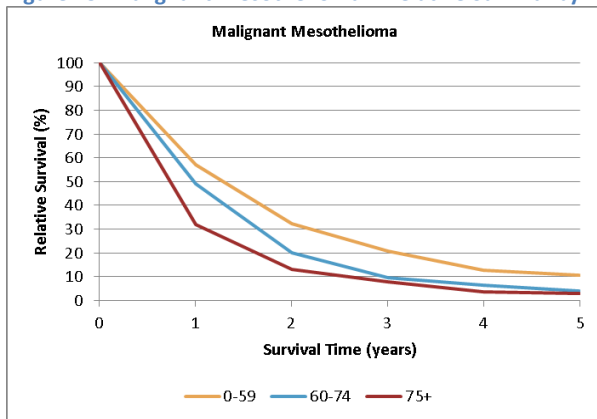
Table 14. Malignant Mesothelioma – Survival by Sex

	N at risk	Observed Survival				Relative Survival			
		1 year	3 year	5 year	5 year CI	1 year	3 year	5 year	5 year CI
Males									
MALIGNANT MESOTHELIOMA	1,184	44.1	9.7	3.8	[2.7 ; 5.2]	45.5	10.6	4.5	[3.2 ; 6.1]
Mesothelioma of pleura and pericardium	1,098	44.3	9.1	3.6	[2.5 ; 5.0]	45.7	9.9	4.2	[2.9 ; 5.8]
Mesothelioma of peritoneum and tunica vaginalis	75	43.2	19.5	9.1	[3.3 ; 18.5]	44.5	20.6	9.8	[3.6 ; 19.9]
Females									
MALIGNANT MESOTHELIOMA	243	45.7	14.3	7.1	[4.1 ; 11.3]	46.6	15.2	7.9	[4.5 ; 12.5]
Mesothelioma of pleura and pericardium	212	44.8	12.3	5.1	[2.5 ; 9.1]	45.7	13.1	5.8	[2.9 ; 10.4]
Mesothelioma of peritoneum and tunica vaginalis	29	*	*	*	*	*	*	*	*

- Survival steeply declines after diagnosis for both males and females, although more long term survivors are observed in the female population.

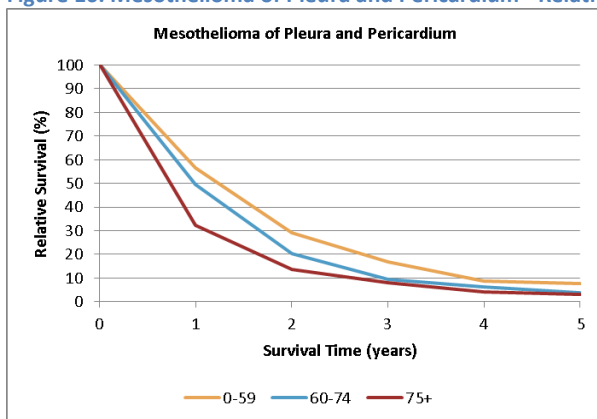
### 4.4.3 Survival by Age Group

Figure 15. Malignant Mesothelioma – Relative Survival by Age Group



- Although very small differences between the age groups can be observed, survival at five years of follow-up is 10% or lower for all age groups.

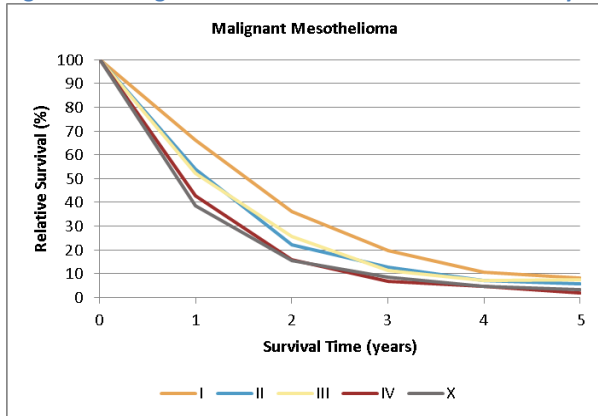
Figure 16. Mesothelioma of Pleura and Pericardium - Relative Survival by Age Group



- Because almost all patients with a malignant mesothelioma are diagnosed with a mesothelioma of the pleura or pericardium, survival by age group is very similar to the results for all malignant mesothelioma together.

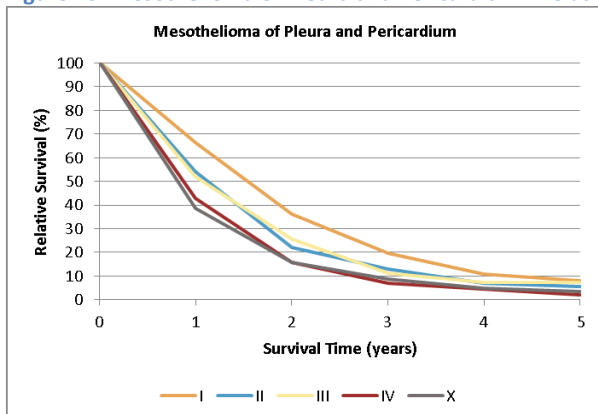
#### 4.4.4 Survival by Stage

Figure 17. Malignant Mesothelioma - Relative Survival by Stage



- At one year after diagnosis, survival differences exist between the stages, ranging between a relative survival of 66.3% for stage I tumours and 42.7% for stage IV tumours. However, at five years after diagnosis, tumours of all stages have a survival rate of less than 10%.

Figure 18. Mesothelioma of Pleura and Pericardium - Relative Survival by Stage



- Because almost all patients with a malignant mesothelioma are diagnosed with a mesothelioma of the pleura or pericardium, survival by stage is very similar to the results for all malignant mesothelioma together.