

# Rare Digestive Tumours

## 1. Epithelial Tumours of Oesophagus

### 1.1 General Results

**Table 1. Epithelial Tumours of Oesophagus: 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 TUMOURS OF OESOPHAGUS		C	4,938	8.12	4.29	67	0.3	0.711	4,249	23.6
Squamous cell carcinoma with variants of oesophagus		R	2,350	3.87	2.17	65	-1.3	0.080	1,965	19.7
Adenocarcinoma with variants of oesophagus		R	2,442	4.02	2.00	68	1.9	0.202	2,152	27.3
Salivary gland type tumours of oesophagus		R	9	0.01	0.01	64	*	*	8	*
Undifferentiated carcinoma of oesophagus		R	34	0.06	0.03	68	*	*	31	*
Males		R/C	N	CR	WSR	Avg Age	EAPC %	p-value	Relative survival N at risk	5yr (%)
EPITHELIAL TUMOURS OF OESOPHAGUS		C	3,799	12.67	7.18	66	-0.9	0.202	3,253	23.6
Squamous cell carcinoma with variants of oesophagus		R	1,682	5.61	3.35	64	-2.7	0.005	1,398	18.1
Adenocarcinoma with variants of oesophagus		C	2,010	6.70	3.65	67	1.5	0.235	1,757	28.1
Salivary gland type tumours of oesophagus		R	8	0.03	0.02	65	*	*	8	*
Undifferentiated carcinoma of oesophagus		R	22	0.07	0.04	68	*	*	22	*
Females		R/C	N	CR	WSR	Avg Age	EAPC %	p-value	Relative survival N at risk	5yr (%)
EPITHELIAL TUMOURS OF OESOPHAGUS		R	1,139	3.70	1.63	71	1.3	0.390	996	23.6
Squamous cell carcinoma with variants of oesophagus		R	668	2.17	1.06	68	1.4	0.273	567	24.0
Adenocarcinoma with variants of oesophagus		R	432	1.40	0.52	74	2.2	0.523	395	23.4
Salivary gland type tumours of oesophagus		R	1	0.00	0.00	55	*	*	0	-
Undifferentiated carcinoma of oesophagus		R	12	0.04	0.02	69	*	*	9	*

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

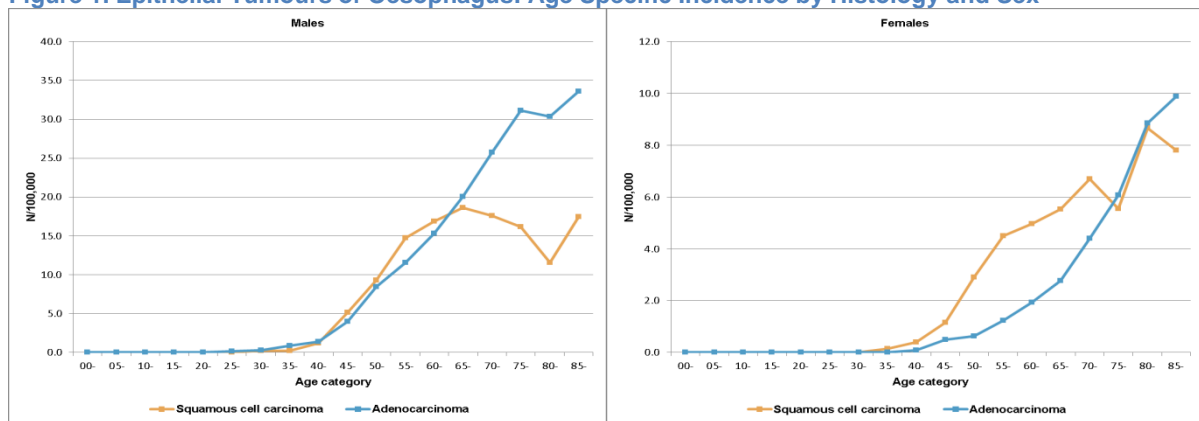
- 4,938 new epithelial tumours of oesophagus are diagnosed in the Flemish Region between 2001 and 2010.
- The male/female ratio is 4.4.

**Table 2. Epithelial Tumours of Oesophagus: Morphological Distribution by Localisation.**

Primary site	Squamous cell	Adenocarcinoma	Salivary gland type	Undifferentiated
Cervical oesophagus	64 2.7%	8 0.3%	1 11.1%	0 0.0%
Thoracic oesophagus	29 1.2%	11 0.5%	0 0.0%	0 0.0%
Abdominal oesophagus	2 0.1%	15 0.6%	0 0.0%	0 0.0%
Upper third of oesophagus	221 9.4%	24 1.0%	0 0.0%	1 2.9%
Middle third of oesophagus	425 18.1%	78 3.2%	2 22.2%	1 2.9%
Lower third of oesophagus	392 16.7%	1114 45.6%	1 11.1%	5 14.7%
Oesophagus, NOS	1217 51.8%	1192 48.8%	5 55.6%	27 79.4%

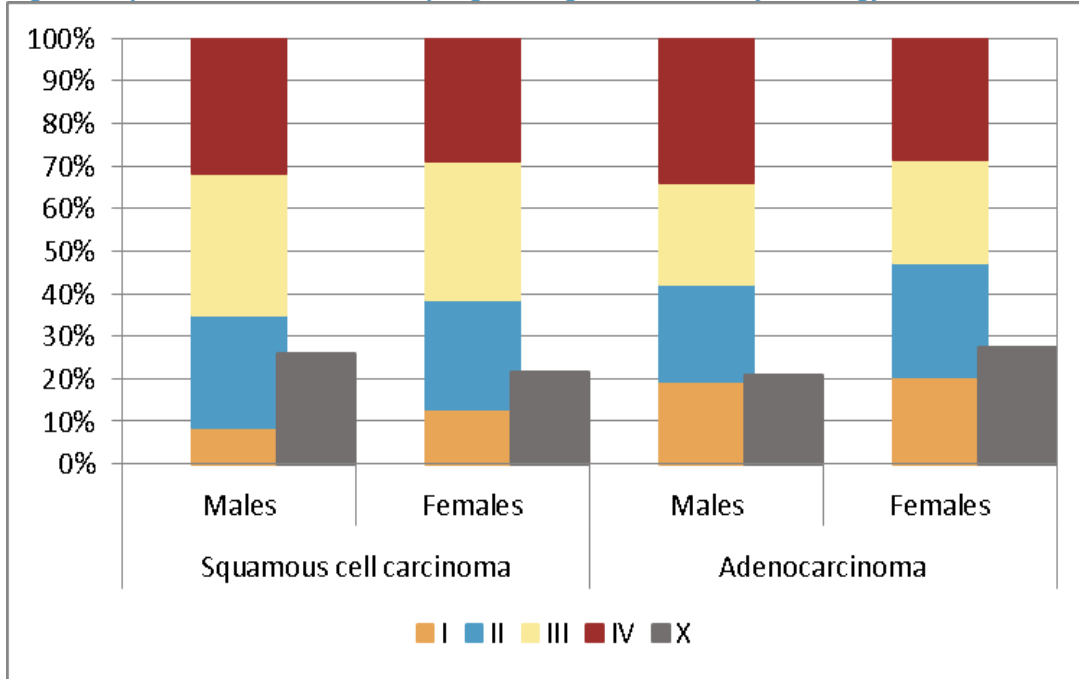
- Four histological entities are considered in the RARECARE list:
  - Squamous cell carcinoma is the most frequent entity in females. The majority of cases in both sexes originate from the middle and lower third of the oesophagus.
  - In males more adenocarcinoma are diagnosed than squamous cell carcinoma. 90% of the adenocarcinoma with a specified primary site is found at the lower third of the oesophagus.
  - Only 9 diagnoses of salivary gland type tumours of oesophagus are registered in the Flemish Region.
  - Undifferentiated carcinomas account for 34 diagnoses.

Figure 1. Epithelial Tumours of Oesophagus: Age Specific Incidence by Histology and Sex



- Incidence rates in males increase from the age of 40 years old. This increase is similar for both major subtypes until the age of 65 years old, when the rates for adenocarcinoma increase further with age while squamous cell carcinoma rates decrease.
- Female adenocarcinomas start to increase at a later age than squamous cell carcinoma. From the age of 75 years, the rates for both histology groups are more comparable.

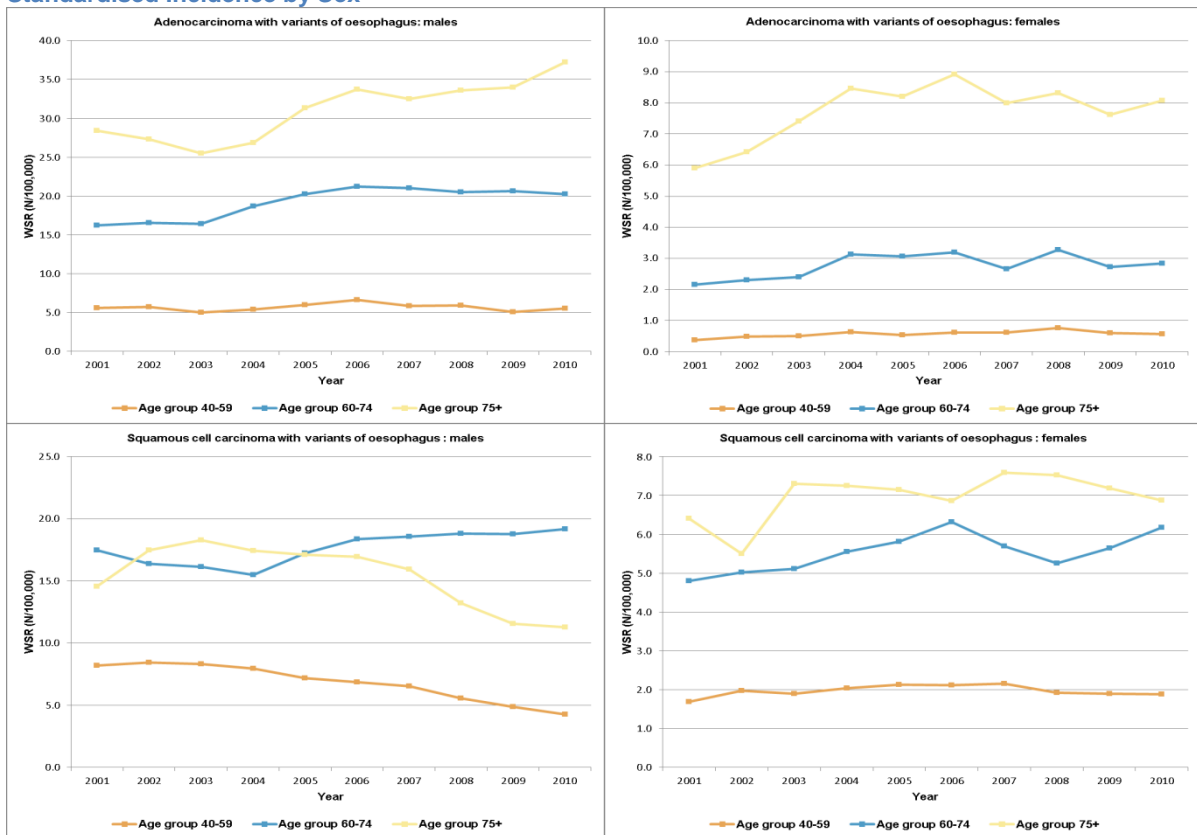
Figure 2. Epithelial Tumours of Oesophagus: Stage Distribution by Histology and Sex



- Stage distribution is similar for both sexes and both histological entities.

### 1.3 Trends

Figure 3. Adenocarcinoma and Squamous Cell Carcinoma with Variants of Oesophagus: Age-Standardised Incidence by Sex



- Incidence rates for males show a significant decrease for squamous cell carcinoma in the period 2001-2010. This significant decrease can only be observed for patients between 40 and 59 years old (EAPC = -7.7% [p = 0.004]) and for patients of 75 years and older (EAPC = -4.7% [p = 0.024]). For the age group 60-74 years, the incidence rates increase annually with 2.2% (p = 0.096).
- Adenocarcinoma incidence rates increase for males of 60 years and older (age group 60-74: EAPC = 3.2% [p = 0.036]; age group 75+: EAPC = 3.2% [p = 0.091]). For the age group 40-59 years the rates seems to decrease (EAPC = -1.4% [p = 0.545]).
- In females, squamous cell carcinomas show no trend for patients between 40 and 59 years of age (EAPC = 0.1% [p = 0.949]) and a non-significant increase for the older age groups (age group 60-74: EAPC = 2.7% [p = 0.181]); age group 75+: EAPC = 3.6% [p = 0.186]).
- The incidence rates for adenocarcinoma in females increase primarily in the age group 75+ (EAPC = 3.8% [p = 0.126]). The rates for the other age groups remain more stable (age group 40-59: EAPC = 0.5% [p = 0.948]; age group 60-74: EAPC = 0.7% [p = 0.844]).

## 1.4 Survival

### 1.4.1 Overall Survival

**Table 3. Epithelial Tumours of Oesophagus – 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 OESOPHAGUS	4,249	51.6	27.3	20.8	14.0	[19.6 ; 22.2]	53.1	29.5	23.6	18.2	[22.1 ; 25.1]
Squamous cell carcinoma	1,965	49.0	24.3	17.9	11.0	[16.2 ; 19.8]	50.1	25.8	19.7	13.7	[17.8 ; 21.8]
Adenocarcinoma with variants	2,152	54.8	30.3	23.6	17.1	[21.7 ; 25.5]	56.6	33.1	27.3	22.8	[25.1 ; 29.5]
Salivary gland type tumours	8	*	*	*	*	*	*	*	*	*	*
Undifferentiated carcinoma	31	*	*	*	*	*	*	*	*	*	*

- Survival of oesophageal cancer rapidly decreases after diagnosis, with nearly half of the patients dying within one year after diagnosis (1-year observed survival: 51.6%, 1-year relative survival: 53.1%).
- Survival rates further diminish to a 10-year observed survival of 14.0% and a 10-year relative survival of 18.2%.
- Although prognosis is low for all types of epithelial tumours of the oesophagus, adenocarcinoma have a remarkably better prognosis than squamous cell carcinoma with 10-year relative survival rates equal to 22.8% and 13.7%, respectively.

### 1.4.2 Survival by Sex

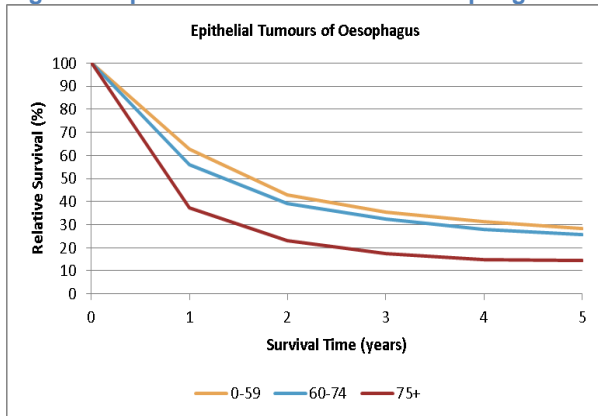
**Table 4. Epithelial Tumours of Oesophagus – Survival by Sex**

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 OESOPHAGUS	3,253	52.6	27.2	20.9	[19.4 ; 22.4]	54.1	29.3	23.6	[21.9 ; 25.3]
Squamous cell carcinoma	1,398	49.0	22.6	16.4	[14.4 ; 18.5]	50.1	24.0	18.1	[15.9 ; 20.4]
Adenocarcinoma with variants	1,757	56.4	31.1	24.5	[22.4 ; 26.7]	58.1	33.8	28.1	[25.7 ; 30.6]
Salivary gland type tumours	8	*	*	*	*	*	*	*	*
Undifferentiated carcinoma	22	*	*	*	*	*	*	*	*
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 OESOPHAGUS	996	48.4	28.0	20.8	[18.2 ; 23.5]	50.0	30.3	23.6	[20.7 ; 26.8]
Squamous cell carcinoma	567	49.0	28.7	21.9	[18.4 ; 25.6]	50.2	30.3	24.0	[20.1 ; 28.0]
Adenocarcinoma with variants	395	47.9	27.0	19.5	[15.5 ; 23.9]	49.9	30.2	23.4	[18.6 ; 28.6]
Salivary gland type tumours	0	-	-	-	-	-	-	-	-
Undifferentiated carcinoma	9	*	*	*	*	*	*	*	*

- Survival differs between males and females for the different histological subtypes. In line with other publications [1], survival benefit for adenocarcinoma over squamous cell carcinoma can be observed in males (5-year relative survival rates of 28.1% versus 18.1%). In females however, a slightly higher survival for squamous cell carcinoma compared with adenocarcinoma is noted (5-year relative survival of 24.0% for squamous cell carcinoma versus 23.4% for adenocarcinoma).

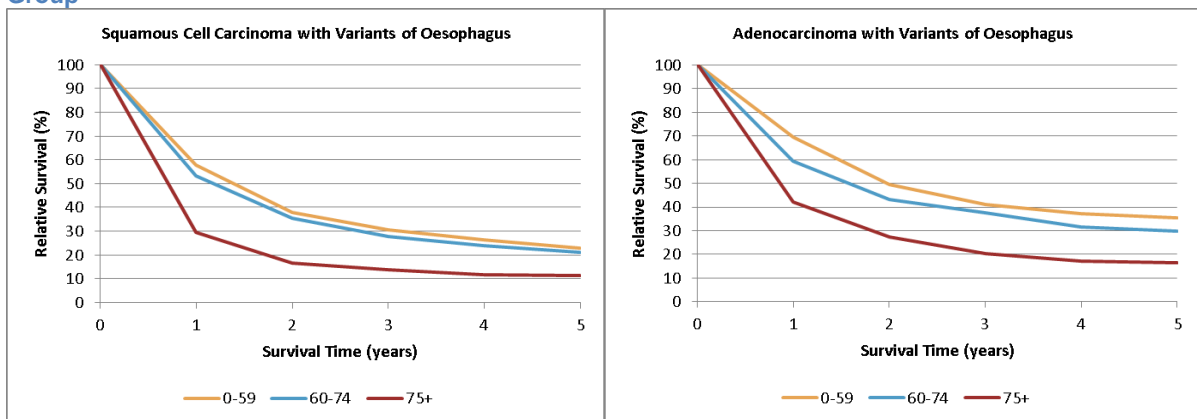
### 1.4.3 Survival by Age Group

Figure 4. Epithelial Tumours of the Oesophagus – Relative Survival by Age Group



- Survival is comparable between the age groups 0-59 years and 60-74 years old, but lower for the patients of 75 years and older.

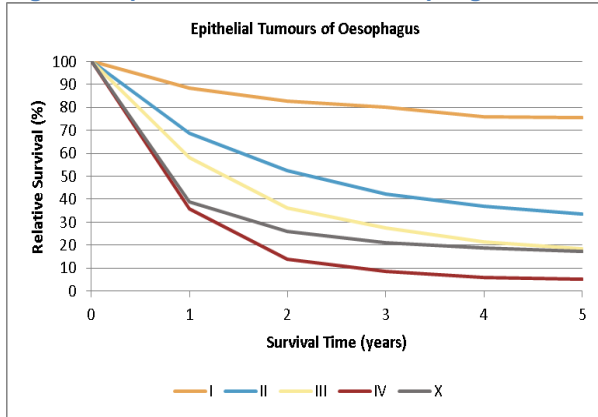
Figure 5. Squamous Cell Carcinoma and Adenocarcinoma of Oesophagus – Relative Survival by Age Group



- Survival is lower for squamous cell carcinoma than for adenocarcinoma in all age groups.

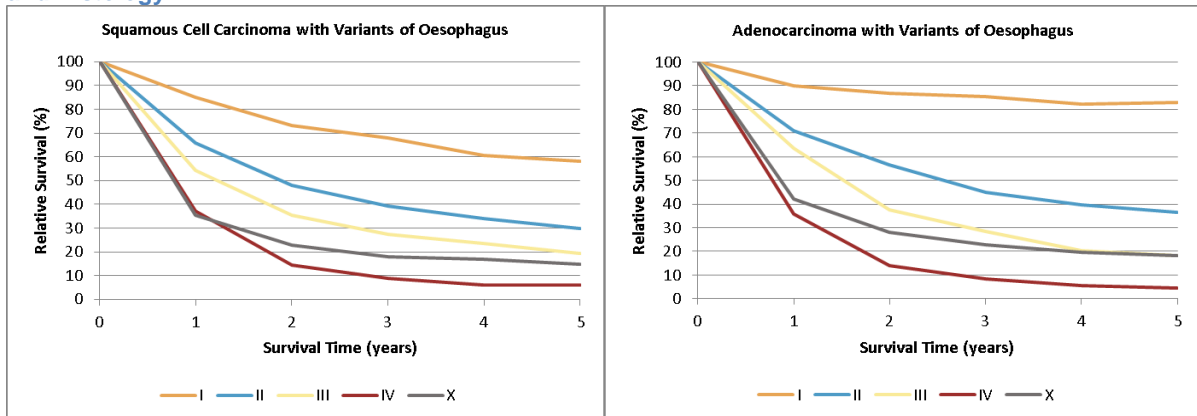
## 1.4.4 Survival by Stage

Figure 6. Epithelial Tumours of Oesophagus – Relative Survival by Stage



- Survival highly depends on the stage at diagnosis.
- Patients diagnosed with a stage I tumour have a 5-year relative survival of about 75%.
- Prognosis is worst for patients diagnosed with a stage IV tumour, for whom survival declines from 35.9% one year after diagnosis to a 5-year relative survival of only 5.1%.

Figure 7. Squamous Cell Carcinoma and Adenocarcinoma of Oesophagus – Relative Survival by Stage and Histology



- Prognosis of Stage I tumours is remarkably worse for squamous cell carcinoma compared to adenocarcinoma of the oesophagus.
- Survival for patients diagnosed with a Stage III and IV tumour is almost similar for squamous cell carcinoma and adenocarcinoma.

## 2. Epithelial Tumours of Stomach

### 2.1 General Results

**Table 5. Epithelial Tumours of Stomach: 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 TUMOURS OF STOMACH		C	7,886	12.97	5.71	72	-1.7	0.004	6,999	27.4
Adenocarcinoma with variants of stomach		C	7,630	12.55	5.53	72	-1.7	0.008	6,773	27.5
Squamous cell carcinoma with variants of stomach		R	30	0.05	0.03	66	*	*	27	*
Salivary gland-type tumours of stomach		R	14	0.02	0.01	75	*	*	12	*
Undifferentiated carcinoma of stomach		R	40	0.07	0.03	70	-10.6	0.199	37	17.7
Males		R/C	N	CR	WSR	Avg Age	EAPC %	p-value	Relative survival N at risk	5yr (%)
EPITHELIAL TUMOURS OF STOMACH		C	4,998	16.67	8.33	71	-2.1	<0.001	4,371	26.4
Adenocarcinoma with variants of stomach		C	4,847	16.16	8.08	71	-1.7	<0.001	4,239	26.6
Squamous cell carcinoma with variants of stomach		R	18	0.06	0.04	62	*	*	17	*
Salivary gland-type tumours of stomach		R	8	0.03	0.01	78	*	*	7	*
Undifferentiated carcinoma of stomach		R	27	0.09	0.05	68	*	*	25	*
Females		R/C	N	CR	WSR	Avg Age	EAPC %	p-value	Relative survival N at risk	5yr (%)
EPITHELIAL TUMOURS OF STOMACH		C	2,888	9.37	3.52	74	-2.3	0.082	2,628	29.1
Adenocarcinoma with variants of stomach		C	2,783	9.03	3.40	74	-2.3	0.109	2,534	29.0
Squamous cell carcinoma with variants of stomach		R	12	0.04	0.01	73	*	*	10	*
Salivary gland-type tumours of stomach		R	6	0.02	0.01	70	*	*	5	*
Undifferentiated carcinoma of stomach		R	13	0.04	0.02	75	*	*	12	*

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

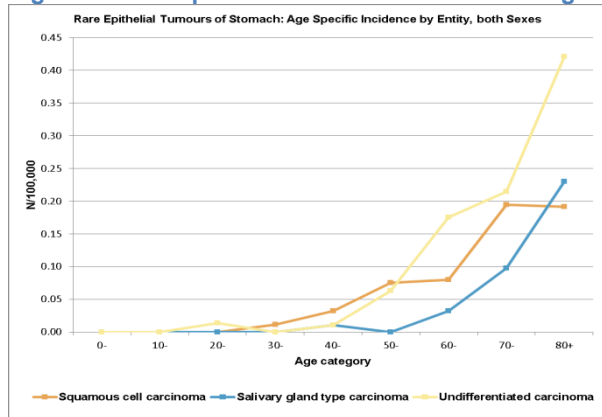
RS: relative survival

AvgAge: average age at diagnosis

### 2.2 Incidence

- 7,886 new epithelial tumours of stomach are diagnosed in the Flemish Region between 2001 and 2010.
- The male/female ratio is 2.4.
- RARECARE defines four different entities:
  - The very common adenocarcinomas represent 97% of the epithelial stomach tumours and are not considered as rare cancers.
  - Squamous cell carcinoma is less common and primarily diagnosed at the cardia.
  - 14 diagnoses of salivary gland type tumours are registered between 2001 and 2010.
  - Undifferentiated carcinoma represents 40 new cases

**Figure 8. Rare Epithelial Tumours of Stomach: Age Specific Incidence by Histological Subtype**



- Under the age of 60 years, the squamous cell carcinomas are the most common of the rare stomach cancer entities.
- The age specific incidence rates for undifferentiated carcinomas increase from the age of 50 years old. The increase continues with age and in the elderly (age group 80+) this histology type is twice as common as the other 2 rare entities.
- Salivary gland type tumours increase from the age of 60 years.

## 2.3 Survival

### 2.3.1 Overall Survival

**Table 6. Epithelial Tumours of Stomach – 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 STOMACH	6,999	49.5	27.1	21.8	14.4	[20.8 ; 22.8]	51.8	31.1	27.4	23.6	[26.1 ; 28.7]
Adenocarcinoma with variants	6,773	50.0	27.3	21.9	14.2	[20.8 ; 22.9]	52.4	31.3	27.5	23.2	[26.2 ; 28.8]
Squamous cell carcinoma with variants	27	*	*	*	*	*	*	*	*	*	*
Salivary gland-type tumours	12	*	*	*	*	*	*	*	*	*	*
Undifferentiated carcinoma	37	24.3	15.1	15.1	-	[5.6 ; 29.0]	25.3	17.4	17.7	-	[6.8 ; 33.4]

- Survival is low for patients diagnosed with an epithelial tumour of the stomach. One year after diagnosis, half of the patients has already died (1-year observed survival: 49.5%, 1-year relative survival: 51.8%).
- Relative survival decreases to 23.6% after ten years of follow-up.



## 2.3.2 Survival by Sex

Table 7. Epithelial Tumours of Stomach – 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 STOMACH	4,371	50.7	26.7	21.0	[19.8 ; 22.3]	53.0	30.5	26.4	[24.8 ; 28.0]
Adenocarcinoma with variants	4,239	51.3	27.0	21.2	[19.9 ; 22.5]	53.6	30.8	26.6	[25.0 ; 28.3]
Squamous cell carcinoma with variants	17	*	*	*	*	*	*	*	*
Salivary gland-type tumours	7	*	*	*	*	*	*	*	*
Undifferentiated carcinoma	25	*	*	*	*	*	*	*	*
Females	N at risk	Observed Survival				Relative Survival			
EPITHELIAL TUMOURS OF STOMACH	2,628	47.4	27.8	23.0	[21.4 ; 24.7]	49.8	32.0	29.1	[27.0 ; 31.3]
Adenocarcinoma with variants	2,534	48.0	27.9	23.0	[21.3 ; 24.8]	50.3	32.1	29.0	[26.9 ; 31.2]
Squamous cell carcinoma with variants	10	*	*	*	*	*	*	*	*
Salivary gland-type tumours	5	*	*	*	*	*	*	*	*
Undifferentiated carcinoma	12	*	*	*	*	*	*	*	*

- Five-year relative survival is higher in females than males (29.1% and 26.4%).

## 3. Epithelial Tumours of Small Intestine

### 3.1 General Results

Table 8. Epithelial Tumours of Small Intestine: 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 TUMOURS OF SMALL INTESTINE		R	462	0.76	0.34	71	-3.0	0.191	381	36.4
Adenocarcinoma with variants of small intestine		R	426	0.70	0.31	71	-1.8	0.370	349	34.9
Squamous cell carcinoma with variants of small intestine		R	4	0.01	0.00	60	*	*	2	*
Males		R/C	N	CR	WSR	Avg Age	EAPC %	p-value	Relative survival N at risk	5yr (%)
EPITHELIAL TUMOURS OF SMALL INTESTINE		R	260	0.87	0.43	70	-2.4	0.411	205	36.4
Adenocarcinoma with variants of small intestine		R	241	0.80	0.39	71	-1.5	0.595	190	33.9
Squamous cell carcinoma with variants of small intestine		R	3	0.01	0.01	60	*	*	1	*
Females		R/C	N	CR	WSR	Avg Age	EAPC %	p-value	Relative survival N at risk	5yr (%)
EPITHELIAL TUMOURS OF SMALL INTESTINE		R	202	0.66	0.26	72	-4.8	0.234	176	35.9
Adenocarcinoma with variants of small intestine		R	185	0.60	0.24	72	-3.3	0.413	159	35.4
Squamous cell carcinoma with variants of small intestine		R	1	0.00	0.00	61	*	*	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

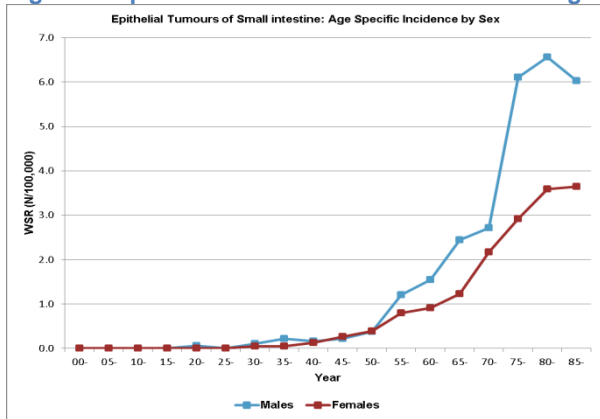
AvgAge: average age at diagnosis

### 3.2 Incidence

- 462 new epithelial tumours of small intestine are diagnosed in the Flemish Region between 2001 and 2010.
- The male/female ratio is 1.6.

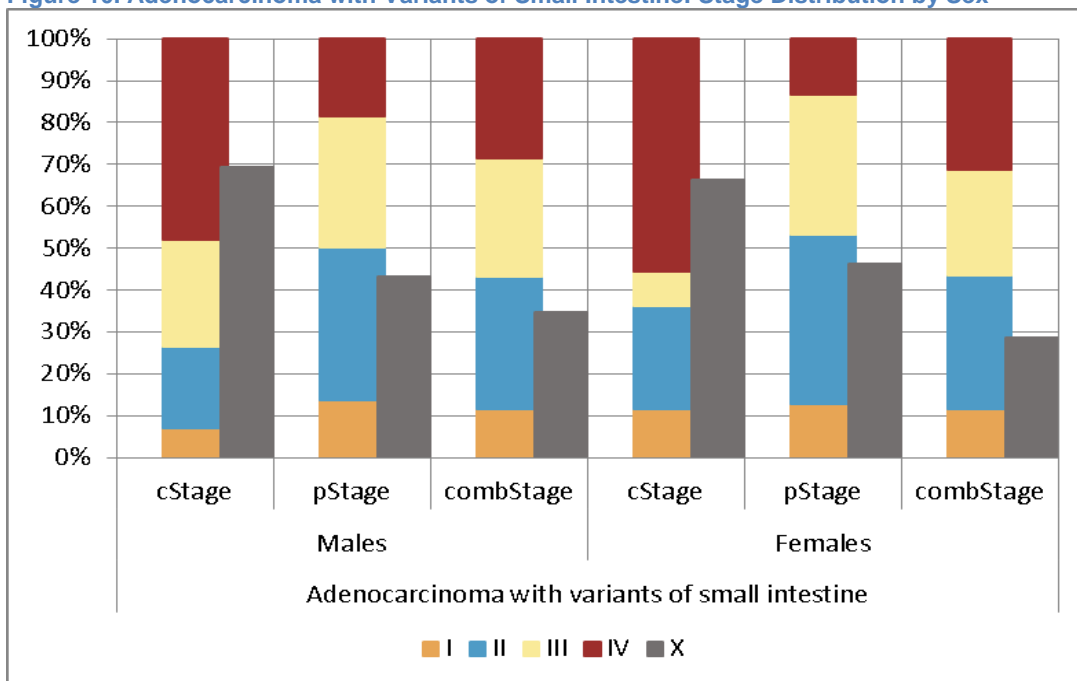
- RARECARE defines two different entities:
  - The very common adenocarcinomas represent 92% of the epithelial small intestine tumours.
  - Squamous cell carcinoma accounts for only 4 cases of small intestine carcinomas, registered at the small intestine between 2001 and 2010 in the Flemish Region.

Figure 9. Epithelial Tumours of Small Intestine: Age Specific Incidence by Sex



- Until the age of 50 years old, the incidence rates between males and females are very comparable.
- After the age of 50 years old, age specific incidence rates increases fast, with higher rates in males than in females.

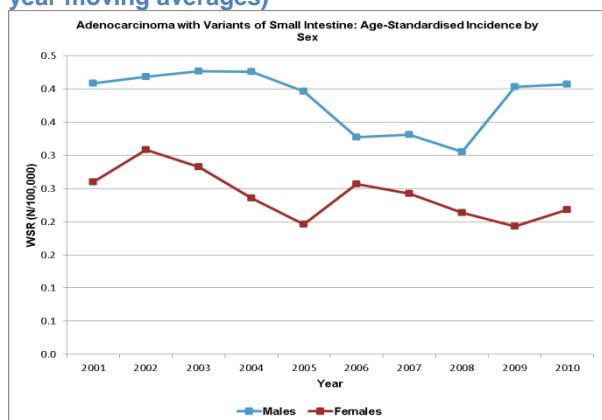
Figure 10. Adenocarcinoma with Variants of Small Intestine: Stage Distribution by Sex



- In about one out of three adenocarcinomas, no information on stage is available. Pathological staging (~45% missing) is more available than clinical staging (~68% missing).
- A high proportion of cancers present in advanced clinical stage, with more than 70% of males and 60% of females diagnosed in clinical stage III or IV.

### 3.3 Trends

Figure 11. Adenocarcinoma with Variants of Small Intestine: Age-Standardised Incidence by Sex (three year moving averages)



- No significant trend is observed in males nor in females

### 3.4 Survival

#### 3.4.1 Overall Survival

Table 9. Epithelial Tumours of Small Intestine – 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 SMALL INTESTINE	381	53.5	35.9	30.9	20.2	[26.1 ; 35.8]	55.6	39.6	36.4	29.8	[30.7 ; 42.2]
Adenocarcinoma with variants	349	53.0	34.4	29.4	18.9	[24.5 ; 34.6]	55.0	37.9	34.9	28.9	[29.0 ; 40.9]
Squamous cell carcinoma with variants	2	*	*	*	*	*	*	*	*	*	*

- Survival decreases rapidly after diagnosis with only slightly more than half of the patients surviving after the first year of diagnosis(1-year relative survival: 55.6%).
- Thereafter, survival decreases more slowly to reach a 10-year relative survival of about 30%.

#### 3.4.2 Survival by Sex

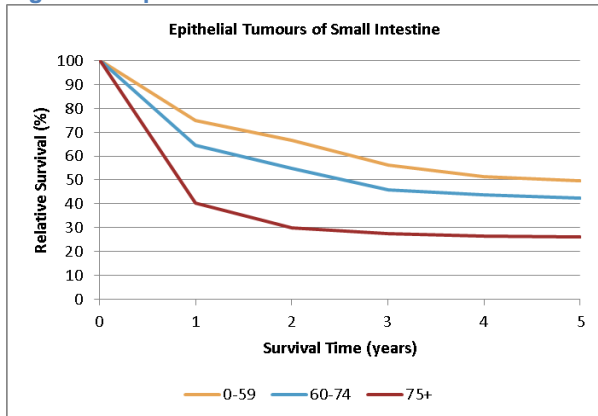
Table 10. Epithelial Tumours of Small Intestine – Survival by Sex

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 SMALL INTESTINE	205	54.2	36.8	30.4	[23.8 ; 37.2]	56.3	41.1	36.4	[28.5 ; 44.6]
Adenocarcinoma with variants	190	52.1	34.6	28.0	[21.3 ; 35.1]	54.3	38.7	33.9	[25.8 ; 42.5]
Squamous cell carcinoma 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 SMALL INTESTINE	176	52.8	34.8	31.2	[24.3 ; 38.4]	54.6	37.9	35.9	[27.9 ; 44.1]
Adenocarcinoma with variants	159	54.1	34.0	30.7	[23.4 ; 38.3]	55.8	36.9	35.4	[27.0 ; 44.1]
Squamous cell carcinoma with variants	1	*	*	*	*	*	*	*	*

- Survival is similar for males and females at all follow-up points.

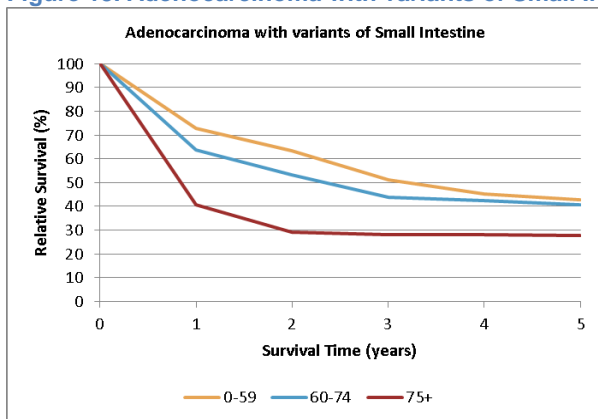
### 3.4.3 Survival by Age Group

Figure 12. Epithelial Tumours of Small Intestine - Relative Survival by Age Group



- Survival is slightly better in the age group 0-59 years compared with the age group 60-74 years, but worse for patients of 75 years and older.

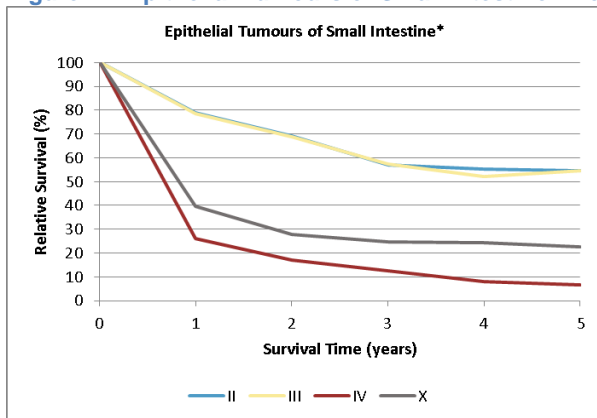
Figure 13. Adenocarcinoma with variants of Small Intestine – Relative Survival by Age Group



- As almost all patients with an epithelial tumour of small intestine are diagnosed with an adenocarcinoma, survival by age group for the adenocarcinomas is very similar to survival of all epithelial tumours of small intestine together.

### 3.4.4 Survival by Stage

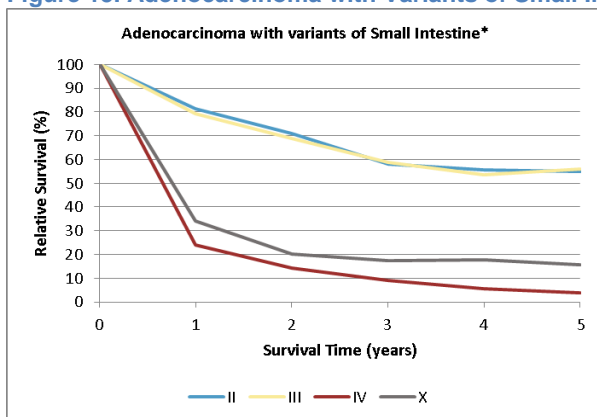
Figure 14. Epithelial Tumours of Small Intestine - Relative Survival by Stage



\* Survival of Stage I is not shown because the number at risk is smaller than 35.

- Prognosis is almost the same for patients diagnosed with a stage II or III cancer (5-year relative survival about 55%).
- Relative survival for patients diagnosed with a tumour stage IV cancer of the small intestine is very poor at one year after diagnosis (26.0%) and further decreases to only 6.6% after five years.

Figure 15. Adenocarcinoma with Variants of Small Intestine – Relative Survival by Stage



\* Survival of Stage I is not shown because the number at risk is smaller than 35.

- Because almost all patients with an epithelial tumour of small intestine are diagnosed with an adenocarcinoma, survival by stage for the adenocarcinomas is very similar to the survival rates for all epithelial tumours of small intestine together.

## 4. Epithelial Tumours of Colon

### 4.1 General Results

**Table 11. Epithelial Tumours of Colon: 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 TUMOURS OF COLON		C	31,504	51.82	22.90	72	0.5	0.210	27,579	62.7
Adenocarcinoma with variants of colon		C	30,796	50.65	22.46	72	1.0	0.020	26,929	63.6
Squamous cell carcinoma with variants of colon		R	9	0.01	0.01	68	*	*	8	*
Males		R/C	N	CR	WSR	Avg Age	EAPC %	p-value	Relative survival N at risk	5yr (%)
EPITHELIAL TUMOURS OF COLON		C	16,815	56.07	27.59	71	0.6	0.170	14,396	62.0
Adenocarcinoma with variants of colon		C	16,487	54.98	27.08	71	1.0	0.031	14,103	62.8
Squamous cell carcinoma with variants of colon		R	2	0.01	0.00	74	*	*	2	*
Females		R/C	N	CR	WSR	Avg Age	EAPC %	p-value	Relative survival N at risk	5yr (%)
EPITHELIAL TUMOURS OF COLON		C	14,689	47.67	19.08	73	0.5	0.407	13,183	63.5
Adenocarcinoma with variants of colon		C	14,309	46.44	18.70	72	0.9	0.056	12,826	64.4
Squamous cell carcinoma with variants of colon		R	7	0.02	0.01	66	*	*	6	*

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

- 31,504 new epithelial tumours of colon are diagnosed in the Flemish Region between 2001 and 2010.
- The male/female ratio is 1.4.
- RARECARE differentiates between two entities:
  - The very common adenocarcinoma.
  - The very rare squamous cell carcinoma. Only 9 cases are diagnosed in the Flemish Region between 2001 and 2010 of which the majority are females.

### 4.3 Survival

#### 4.3.1 Overall Survival

**Table 12. Epithelial Tumours of Colon – 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 COLON	27,579	78.4	60.3	50.5	36.0	[49.9 ; 51.1]	81.7	68.3	62.7	58.3	[61.9 ; 63.5]
Adenocarcinoma with variants	26,929	79.5	61.2	51.2	36.4	[50.5 ; 51.8]	82.9	69.3	63.6	59.1	[62.8 ; 64.4]
Squamous cell carcinoma with variants	8	*	*	*	*	*	*	*	*	*	*

- Survival is rather good for patients diagnosed with an epithelial tumour of colon (5-year relative survival: 62.7%, 10-year relative survival: 58.3%).

## 4.3.2 Survival by Sex

Table 13. Epithelial Tumours of Colon – 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 COLON	14,396	78.7	59.9	49.3	[48.4 ; 50.2]	82.1	68.4	62.0	[60.9 ; 63.0]
Adenocarcinoma with variants	14,103	79.7	60.7	49.9	[49.0 ; 50.8]	83.2	69.3	62.8	[61.6 ; 63.9]
Squamous cell carcinoma with variants	2	*	*	*	*	*	*	*	*
Females	N at risk	Observed Survival				Relative Survival			
EPITHELIAL TUMOURS OF COLON	13,183	78.2	60.7	51.8	[50.9 ; 52.7]	81.3	68.2	63.5	[62.4 ; 64.6]
Adenocarcinoma with variants	12,826	79.4	61.6	52.6	[51.6 ; 53.5]	82.5	69.2	64.4	[63.3 ; 65.5]
Squamous cell carcinoma with variants	6	*	*	*	*	*	*	*	*

- Prognosis is comparable for males and females diagnosed with an epithelial colon tumour (5-year relative survival of 62.0% and 63.5% respectively).

## 5. Epithelial Tumours of Rectum

### 5.1 General Results

Table 14. Epithelial Tumours of Rectum: Incidence, Trends, Survival

Flemish Region 2001-2010		Incidence				Trend		Survival		
Both Sexes		R/C	N	CR	WSR	Avg Age	EAPC		Relative Survival	
							%	p-value	N at risk	5yr (%)
EPITHELIAL TUMOURS OF RECTUM		C	13,062	21.48	10.49	69	1.2	0.015	11,803	64.9
Adenocarcinoma with variants of rectum		C	12,881	21.19	10.36	69	1.5	0.004	11,636	65.2
Squamous cell carcinoma with variants of rectum		R	21	0.03	0.02	69	5.9	0.493	20	*
Basaloid carcinoma of rectum		R	1	0.00	0.00	68	*	*	1	*
Males		R/C	N	CR	WSR	Avg Age	EAPC		Relative survival	
EPITHELIAL TUMOURS OF RECTUM		C	7,946	26.50	14.00	68	1.1	0.012	7,096	65.1
Adenocarcinoma with variants of rectum		C	7,853	26.19	13.85	68	1.5	0.004	7,010	65.4
Squamous cell carcinoma with variants of rectum		R	9	0.03	0.01	69	*	*	9	*
Basaloid carcinoma of rectum		R	0	-	-	-	-	-	-	-
Females		R/C	N	CR	WSR	Avg Age	EAPC		Relative survival	
EPITHELIAL TUMOURS OF RECTUM		C	5,116	16.60	7.46	70	1.1	0.136	4,707	64.6
Adenocarcinoma with variants of rectum		C	5,028	16.32	7.35	70	1.3	0.062	4,626	65.0
Squamous cell carcinoma with variants of rectum		R	12	0.04	0.02	70	*	*	11	*
Basaloid carcinoma of rectum		R	1	0.00	0.00	68	*	*	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

### 5.2 Incidence

- 13,062 new epithelial tumours of the rectum are diagnosed in the Flemish Region between 2001 and 2010.
- The male/female ratio is 1.9.
- RARECARE differentiates between three entities:
  - The very common adenocarcinoma.

- The very rare squamous cell carcinoma of which only 21 cases are diagnosed in the Flemish Region between 2001 and 2010.
- Only one diagnosis of basaloid rectal carcinoma is registered.

## 5.3 Survival

### 5.3.1 Overall Survival

Table 15. Epithelial Tumours of Rectum – 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 RECTUM	11,803	83.2	65.8	54.7	39.8	[53.7 ; 55.6]	86.1	72.8	64.9	57.9	[63.8 ; 66.1]
Adenocarcinoma with variants	11,636	83.6	66.2	55.0	39.9	[54.0 ; 55.9]	86.5	73.2	65.2	58.1	[64.1 ; 66.4]
Squamous cell carcinoma with variants	20	*	*	*	*	*	*	*	*	*	*
Basaloid carcinoma	1	*	*	*	*	*	*	*	*	*	*

- Survival at one year after diagnosis is rather high, with more than 80% of patients surviving.
- Relative survival decreases to 57.9% at ten years after diagnosis.

### 5.3.2 Survival by Sex

Table 16. Epithelial Tumours of Rectum – 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 RECTUM	7,096	83.3	65.6	54.3	[53.0 ; 55.5]	86.4	73.1	65.1	[63.6 ; 66.6]
Adenocarcinoma with variants	7,010	83.7	65.9	54.5	[53.2 ; 55.8]	86.7	73.4	65.4	[63.9 ; 67.0]
Squamous cell carcinoma with variants	9	*	*	*	*	*	*	*	*
Basaloid carcinoma	0	-	-	-	-	-	-	-	-
Females	N at risk	Observed Survival				Relative Survival			
EPITHELIAL TUMOURS OF RECTUM	4,707	82.9	66.0	55.3	[53.7 ; 56.8]	85.6	72.3	64.6	[62.8 ; 66.4]
Adenocarcinoma with variants	4,626	83.5	66.5	55.6	[54.1 ; 57.2]	86.1	72.8	65.0	[63.1 ; 66.7]
Squamous cell carcinoma with variants	11	*	*	*	*	*	*	*	*
Basaloid carcinoma	1	*	*	*	*	*	*	*	*

- Survival does almost not differ between males and females.



## 6. Epithelial Tumours of Anal Canal

### 6.1 General Results

**Table 17. Epithelial Tumours of Anal Canal: 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 TUMOURS OF ANAL CANAL		R	609	1.00	0.53	66	2.8	0.199	539	68.6
Squamous cell carcinoma with variants of anal canal		R	452	0.74	0.42	64	2.9	0.191	405	73.9
Adenocarcinoma with variants of anal canal		R	141	0.23	0.10	72	1.9	0.673	123	51.4
Paget's disease of anal canal		R	5	0.01	0.00	67	*	*	0	-
Males		R/C	N	CR	WSR	Avg Age	EAPC %	p-value	Relative survival N at risk	5yr (%)
EPITHELIAL TUMOURS OF ANAL CANAL		R	264	0.88	0.50	65	2.4	0.282	230	63.9
Squamous cell carcinoma with variants of anal canal		R	176	0.59	0.35	64	2.4	0.385	156	69.6
Adenocarcinoma with variants of anal canal		R	78	0.26	0.13	70	1.2	0.844	68	50.9
Paget's disease of anal canal		R	4	0.01	0.01	66	*	*	0	-
Females		R/C	N	CR	WSR	Avg Age	EAPC %	p-value	Relative survival N at risk	5yr (%)
EPITHELIAL TUMOURS OF ANAL CANAL		R	345	1.12	0.56	67	2.8	0.325	309	72.0
Squamous cell carcinoma with variants of anal canal		R	276	0.90	0.48	65	3.0	0.248	249	76.6
Adenocarcinoma with variants of anal canal		R	63	0.20	0.07	75	1.8	0.712	55	52.4
Paget's disease of anal canal		R	1	0.00	0.00	70	*	*	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

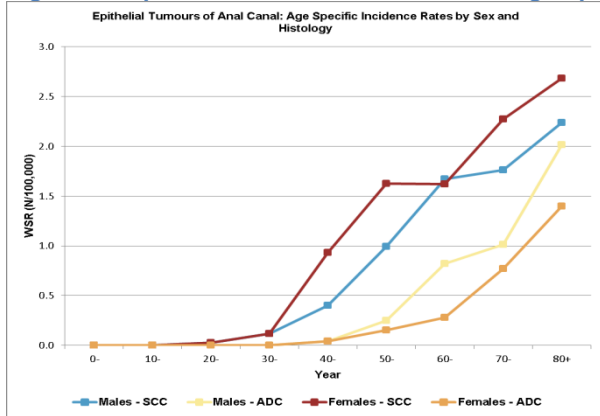
RS: relative survival

AvgAge: average age at diagnosis

### 6.2 Incidence

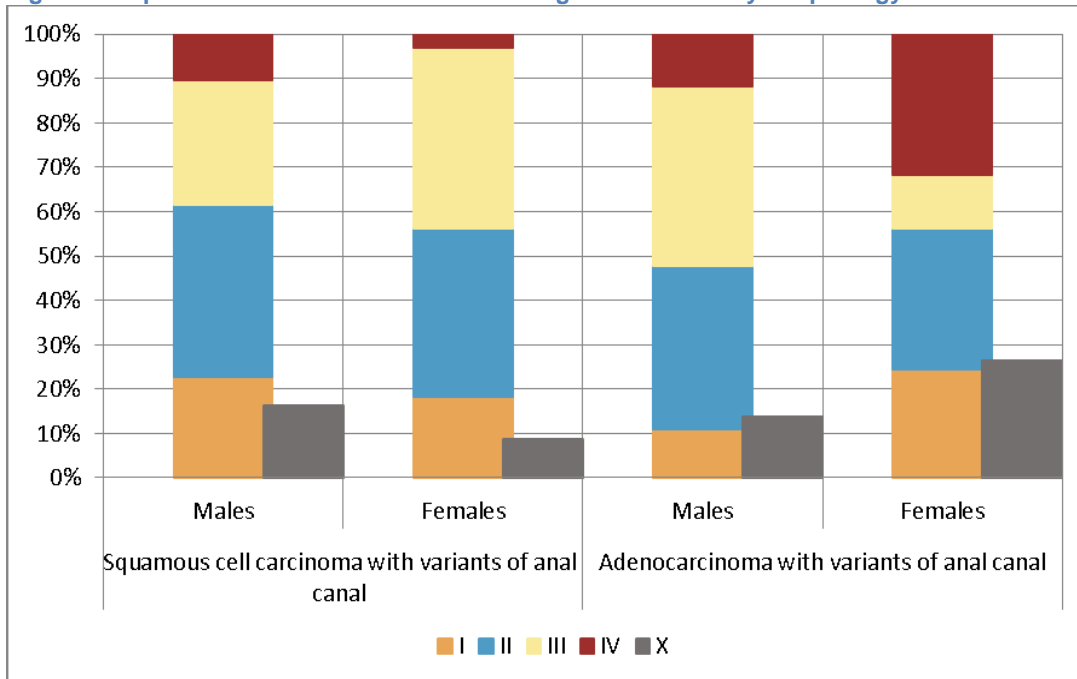
- 609 new epithelial tumours of the anal canal are diagnosed in the Flemish Region between 2001 and 2010.
- The male/female ratio is 0.9.
- RARECARE differentiates between three rare entities:
  - Squamous cell carcinoma represents two out of three diagnoses in males and four out of five in females. More females are diagnosed than males (M/F ratio = 0.7).
  - Adenocarcinomas occur more frequently in males (M/F ratio = 1.8).
  - Only 5 Paget's disease of anal canal are diagnosed in the Flemish Region between 2001 and 2010.

Figure 16. Epithelial tumours of Anal Canal: Age Specific Incidence Rates by Sex and Histology



- Incidence rates increase for squamous cell carcinoma from the age of 30 years old. In females, the increase is faster than in males.
- Adenocarcinoma incidence rates increase from the age of 50 years old. The increase in males is more pronounced than in females.

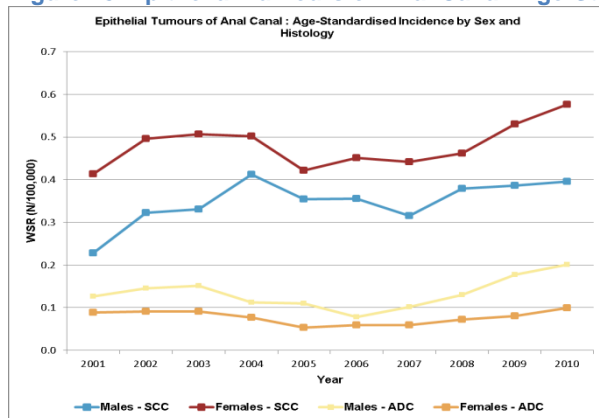
Figure 17. Epithelial Tumours of Anal Canal: Stage Distribution by Morphology and Sex



- Squamous cell carcinomas have a prognostic better stage distribution than adenocarcinomas.
  - In males, stage I and II represent more than 60% of the squamous cell carcinoma cases and less than 50% of the adenocarcinomas.
  - In females, more than 30% of the adenocarcinomas are diagnosed in stage IV. This should be interpreted with caution due to the very limited number of adenocarcinoma cases.

## 6.3 Trends

Figure 18. Epithelial Tumours of Anal Canal: Age-Standardised Incidence by Sex and Histology



- Incidence rates for squamous and adenocarcinoma increase in both sexes, but none of the trends is significant.
- The increase in squamous cell carcinoma incidence is almost twice the increase in adenocarcinoma.

## 6.4 Survival

### 6.4.1 Overall Survival

Table 18. Epithelial Tumours of Anal Canal – 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 ANAL CANAL	539	83.3	66.9	59.3	45.6	[54.7 ; 63.6]	86.0	73.0	68.6	59.7	[63.2 ; 73.6]
Squamous cell carcinoma with variants	405	86.4	71.7	65.3	50.2	[60.1 ; 70.0]	88.9	77.2	73.9	63.0	[68.0 ; 79.2]
Adenocarcinoma with variants	123	75.6	53.8	40.8	31.1	[30.9 ; 50.4]	79.0	61.3	51.4	48.2	[38.9 ; 63.5]
Paget's disease	0	-	-	-	-	-	-	-	-	-	-

- Patients with an epithelial tumour of the anal canal have a rather good prognosis ranging from an relative survival of 86.0% at one year to 59.7% at ten years after diagnosis.
- Squamous cell carcinoma with variants of the anal canal have a better prognosis than adenocarcinoma at all points in time.

### 6.4.2 Survival by Sex

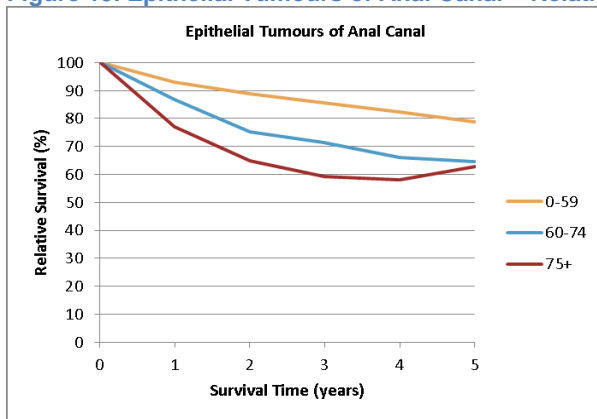
Table 19. Epithelial Tumours of Anal Canal – Survival by Sex

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 ANAL CANAL	230	79.6	63.1	54.3	[47.1 ; 60.9]	82.1	69.4	63.9	[55.4 ; 71.8]
Squamous cell carcinoma with variants	156	82.7	69.3	60.5	[51.7 ; 68.1]	85.0	75.2	69.6	[59.5 ; 78.4]
Adenocarcinoma with variants	68	75.0	49.5	39.8	[26.8 ; 52.5]	78.4	56.9	50.9	[34.1 ; 67.3]
Paget's disease	0	-	-	-	-	-	-	-	-
Females	N at risk	Observed Survival				Relative Survival			
EPITHELIAL TUMOURS OF ANAL CANAL	309	86.1	69.8	63.1	[57.1 ; 68.6]	88.8	75.6	72.0	[65.1 ; 78.3]
Squamous cell carcinoma with variants	249	88.8	73.2	68.5	[61.9 ; 74.2]	91.3	78.5	76.6	[69.2 ; 82.9]
Adenocarcinoma with variants	55	76.4	59.2	42.5	[27.6 ; 56.6]	79.9	66.8	52.4	[34.1 ; 69.7]
Paget's disease	0	-	-	-	-	-	-	-	-

- Survival is better in females than males for both squamous cell carcinomas and adenocarcinomas although the difference is less pronounced in adenocarcinomas.

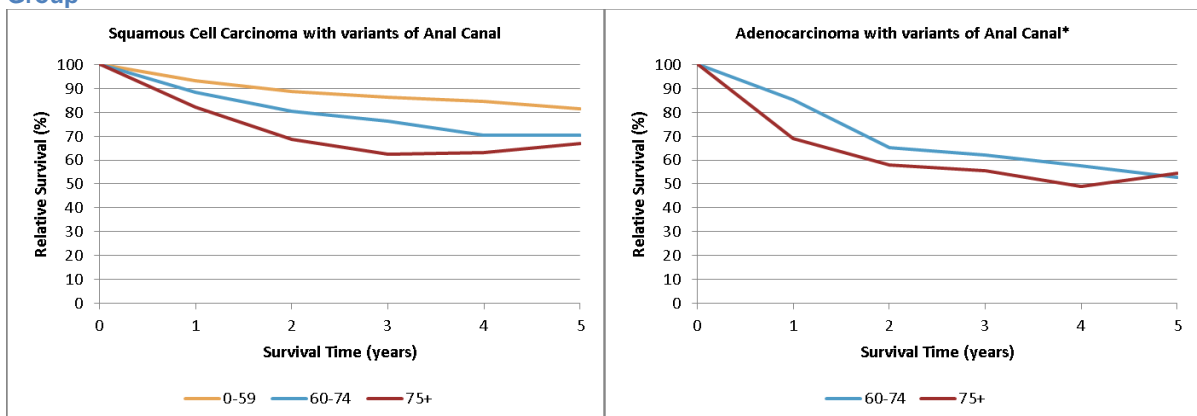
### 6.4.3 Survival by Age Group

Figure 19. Epithelial Tumours of Anal Canal – Relative Survival by Age Group



- Survival is better for the youngest age group (0-59 years) than for the older age groups.
- Relative survival for the oldest patients is only 59.2% after three years of follow-up and stays stable afterwards.

Figure 20. Squamous Cell Carcinoma and Adenocarcinoma of Anal Canal – Relative Survival by Age Group

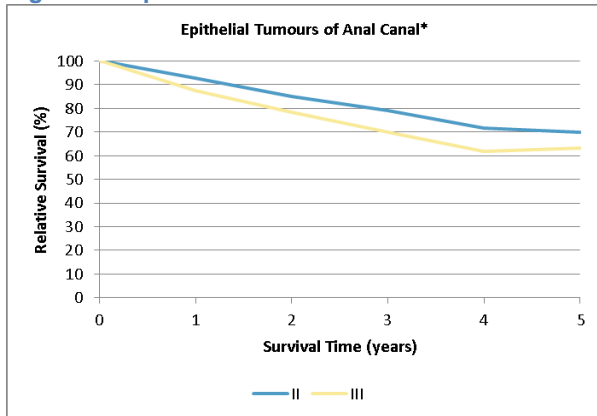


\* Survival of the age group 0-59 years is not displayed because the number at risk is smaller than 35.

- Survival is worse for adenocarcinoma than for squamous cell carcinoma, especially in the age group 60-74 years.

## 6.4.4 Survival by Stage

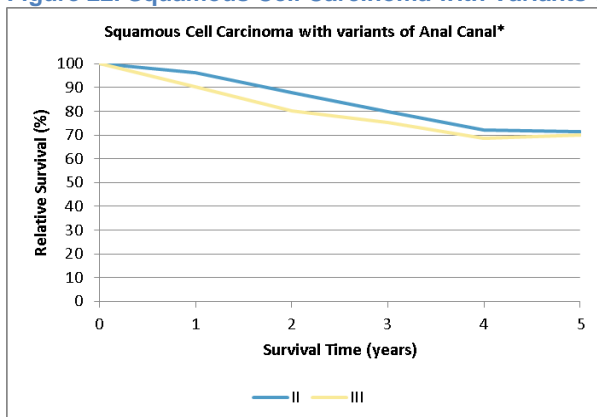
Figure 21. Epithelial Tumours of Anal Canal – Relative Survival by Stage



\* Survival of the stage I and IV is not displayed because the number at risk is smaller than 35.

- Relative survival is rather good, with 70% at 5 years for stage II and more than 60% for stage III.

Figure 22. Squamous Cell Carcinoma with Variants of Anal Canal – Relative Survival by Stage



\* Survival of the stage I and IV is not displayed because the number at risk is smaller than 35.

- For a large part of the epithelial tumours of anal canal, no stage is available because they originate from a location for which staging is not applicable according to TNM.
- For all epithelial tumours of the anal canal together, only a very small survival benefit can be observed for stage II tumours compared with stage III tumours. This difference is even more negligible for squamous cell carcinoma.

## 7. Epithelial Tumours of Pancreas

### 7.1 General results

Table 20. Epithelial Tumours of Pancreas: Incidence, Trends, Survival

Flemish Region 2001-2010		Incidence				Trend		Survival		
Both Sexes		R/C	N	CR	WSR	Avg Age	EAPC		Relative Survival	
							%	p-value	N at risk	5yr (%)
EPITHELIAL TUMOURS OF PANCREAS		C	6,320	10.39	4.93	70	2.1	0.001	5,685	7.1
Adenocarcinoma with variants of pancreas		C	4,914	8.08	4.07	68	3.0	<0.001	4,414	6.1
Squamous cell carcinoma with variants of pancreas		R	3	0.00	0.00	68	*	*	3	*
Acinar cell carcinoma of pancreas		R	27	0.04	0.02	65	*	*	26	*
Mucinous cystadenocarcinoma of pancreas		R	10	0.02	0.01	64	*	*	9	*
Intraductal papillary mucinous carcinoma invasive of pancreas		R	23	0.04	0.02	64	*	*	18	*
Solid pseudopapillary carcinoma of pancreas		R	7	0.01	0.01	47	*	*	6	*
Serous cystadenocarcinoma of pancreas		R	1	0.00	0.00	74	*	*	1	*
Carcinoma with osteoclast-like giant cells of pancreas		R	2	0.00	0.00	72	*	*	2	*
Males		R/C	N	CR	WSR	Avg Age	EAPC		Relative survival	
							%	p-value	N at risk	5yr (%)
EPITHELIAL TUMOURS OF PANCREAS		C	3,296	10.99	5.80	68	1.2	0.148	2,905	7.8
Adenocarcinoma with variants of pancreas		C	2,647	8.83	4.80	67	2.1	0.006	2,333	6.8
Squamous cell carcinoma with variants of pancreas		R	3	0.01	0.01	68	*	*	3	*
Acinar cell carcinoma of pancreas		R	17	0.06	0.03	66	*	*	16	*
Mucinous cystadenocarcinoma of pancreas		R	3	0.01	0.01	71	*	*	2	*
Intraductal papillary mucinous carcinoma invasive of pancreas		R	14	0.05	0.03	62	*	*	11	*
Solid pseudopapillary carcinoma of pancreas		R	3	0.01	0.01	70	*	*	2	*
Serous cystadenocarcinoma of pancreas		R	0	-	-	-	-	-	0	-
Carcinoma with osteoclast-like giant cells of pancreas		R	0	-	-	-	-	-	0	-
Females		R/C	N	CR	WSR	Avg Age	EAPC		Relative survival	
							%	p-value	N at risk	5yr (%)
EPITHELIAL TUMOURS OF PANCREAS		C	3,024	9.81	4.14	71	2.6	0.002	2,780	6.3
Adenocarcinoma with variants of pancreas		C	2,267	7.36	3.39	69	4.1	<0.001	2,081	5.3
Squamous cell carcinoma with variants of pancreas		R	0	-	-	-	-	-	0	-
Acinar cell carcinoma of pancreas		R	10	0.03	0.02	64	*	*	10	*
Mucinous cystadenocarcinoma of pancreas		R	7	0.02	0.01	61	*	*	7	*
Intraductal papillary mucinous carcinoma invasive of pancreas		R	9	0.03	0.01	67	*	*	7	*
Solid pseudopapillary carcinoma of pancreas		R	4	0.01	0.01	30	*	*	4	*
Serous cystadenocarcinoma of pancreas		R	1	0.00	0.00	74	*	*	1	*
Carcinoma with osteoclast-like giant cells of pancreas		R	2	0.01	0.00	72	*	*	2	*

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

### 7.2 Incidence

- 6,320 new epithelial tumours of the pancreas are diagnosed in the Flemish Region between 2001 and 2010.
- The male/female ratio is 1.4.

- RARECARE differentiates between the common adenocarcinoma which represents almost all diagnoses and seven rare entities:
  - Acinar cell carcinoma is the most common rare entity with 27 cases between 2001 and 2010.
  - With 23 cases, intraductal papillary mucinous carcinoma invasive of pancreas is the 2<sup>nd</sup> most common rare pancreatic cancer entity.
  - The remaining types each represent 10 cases or less.

## 7.3 Survival

### 7.3.1 Overall Survival

Table 21. Epithelial Tumours of Pancreas – 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 PANCREAS	5,685	29.4	9.1	6.2	4.6	[5.6 ; 6.9]	30.2	9.9	7.1	6.2	[6.4 ; 7.9]
Adenocarcinoma with variants	4,414	31.8	8.7	5.4	3.6	[4.7 ; 6.2]	32.5	9.3	6.1	4.9	[5.3 ; 7.0]
Squamous cell carcinoma with variants	3	*	*	*	*	*	*	*	*	*	*
Acinar cell carcinoma	26	*	*	*	*	*	*	*	*	*	*
Mucinous cystadenocarcinoma	9	*	*	*	*	*	*	*	*	*	*
Intraductal papillary mucinous carcinoma invasive	18	*	*	*	*	*	*	*	*	*	*
Solid pseudopapillary carcinoma	6	*	*	*	*	*	*	*	*	*	*
Serous cystadenocarcinoma	1	*	*	*	*	*	*	*	*	*	*
Carcinoma with osteoclast-like giant cells	2	*	*	*	*	*	*	*	*	*	*

- Survival steeply declines after diagnosis with less than one third of the patients surviving the first year.
- At three years after diagnosis, less than 10% of the patients is still alive.

### 7.3.2 Survival by Sex

Table 22. Epithelial Tumours of Pancreas – 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	
<b>Males</b>										
EPITHELIAL TUMOURS OF PANCREAS	2,905	29.6	9.9	6.8	[5.9 ; 7.9]	30.5	10.7	7.8	[6.7 ; 9.0]	
Adenocarcinoma with variants	2,333	31.5	9.5	6.0	[5.0 ; 7.1]	32.4	10.2	6.8	[5.7 ; 8.0]	
Squamous cell carcinoma with variants	3	*	*	*	*	*	*	*	*	
Acinar cell carcinoma	16	*	*	*	*	*	*	*	*	
Mucinous cystadenocarcinoma	2	*	*	*	*	*	*	*	*	
Intraductal papillary mucinous carcinoma invasive	11	*	*	*	*	*	*	*	*	
Solid pseudopapillary carcinoma	2	*	*	*	*	*	*	*	*	
Serous cystadenocarcinoma	0	-	-	-	-	-	-	-	-	
Carcinoma with osteoclast-like giant cells	0	-	-	-	-	-	-	-	-	
<b>Females</b>										
EPITHELIAL TUMOURS OF PANCREAS	2,780	29.2	8.3	5.6	[4.7 ; 6.6]	30.0	8.9	6.3	[5.3 ; 7.5]	
Adenocarcinoma with variants	2,081	32.1	7.9	4.8	[3.9 ; 5.9]	32.7	8.3	5.3	[4.3 ; 6.5]	
Squamous cell carcinoma with variants	0	-	-	-	-	-	-	-	-	
Acinar cell carcinoma	10	*	*	*	*	*	*	*	*	
Mucinous cystadenocarcinoma	7	*	*	*	*	*	*	*	*	
Intraductal papillary mucinous carcinoma invasive	7	*	*	*	*	*	*	*	*	
Solid pseudopapillary carcinoma	4	*	*	*	*	*	*	*	*	
Serous cystadenocarcinoma	1	*	*	*	*	*	*	*	*	
Carcinoma with osteoclast-like giant cells	2	*	*	*	*	*	*	*	*	

- Survival is comparable for females and males diagnosed with an epithelial tumour of the pancreas.

## 8. Epithelial Tumours of Liver and Intrahepatic Bile Tract (IBT)

### 8.1 General Results

Table 23. Epithelial Tumours of Liver and Intrahepatic Bile Tract: 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 TUMOURS OF LIVER AND INTRAHEPATIC BILE TRACT (IBT)		R	2,490	4.10	2.12	67	7.0	<0.001	2,216	18.9
Hepatocellular carcinoma of Liver and IBT		R	1,690	2.78	1.49	66	8.9	<0.001	1,489	22.8
Cholangiocarcinoma of IBT		R	522	0.86	0.43	68	5.5	0.007	462	11.7
Adenocarcinoma with variants of liver and IBT		R	107	0.18	0.09	68	-4.6	0.585	101	6.6
Undifferentiated carcinoma of liver and IBT		R	0	-	-	-	-	-	0	-
Squamous cell carcinoma with variants of liver and IBT		R	0	-	-	-	-	-	0	-
Bile duct cystadenocarcinoma of IBT		R	1	0.00	0.00	67	*	*	1	*
Males		R/C	N	CR	WSR	Avg Age	EAPC %	p-value	Relative survival N at risk	5yr (%)
EPITHELIAL TUMOURS OF LIVER AND INTRAHEPATIC BILE TRACT (IBT)		R	1,646	5.49	3.04	66	8.7	<0.001	1,438	19.7
Hepatocellular carcinoma of Liver and IBT		R	1,222	4.08	2.30	66	10.1	<0.001	1,065	22.6
Cholangiocarcinoma of IBT		R	268	0.89	0.48	67	7.7	0.016	224	11.6
Adenocarcinoma with variants of liver and IBT		R	65	0.22	0.12	66	-7.2	0.514	61	8.4
Undifferentiated carcinoma of liver and IBT		R	0	-	-	-	-	-	0	-
Squamous cell carcinoma with variants of liver and IBT		R	0	-	-	-	-	-	0	-
Bile duct cystadenocarcinoma of IBT		R	0	-	-	-	-	-	0	-
Females		R/C	N	CR	WSR	Avg Age	EAPC %	p-value	Relative survival N at risk	5yr (%)
EPITHELIAL TUMOURS OF LIVER AND INTRAHEPATIC BILE TRACT (IBT)		R	844	2.74	1.29	69	3.2	0.026	778	17.5
Hepatocellular carcinoma of Liver and IBT		R	468	1.52	0.75	67	5.4	0.027	424	23.3
Cholangiocarcinoma of IBT		R	254	0.82	0.39	68	3.1	0.119	238	11.9
Adenocarcinoma with variants of liver and IBT		R	42	0.14	0.06	70	1.5	0.897	40	3.8
Undifferentiated carcinoma of liver and IBT		R	0	-	-	-	-	-	0	-
Squamous cell carcinoma with variants of liver and IBT		R	0	-	-	-	-	-	0	-
Bile duct cystadenocarcinoma of IBT		R	1	0.00	0.00	67	*	*	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

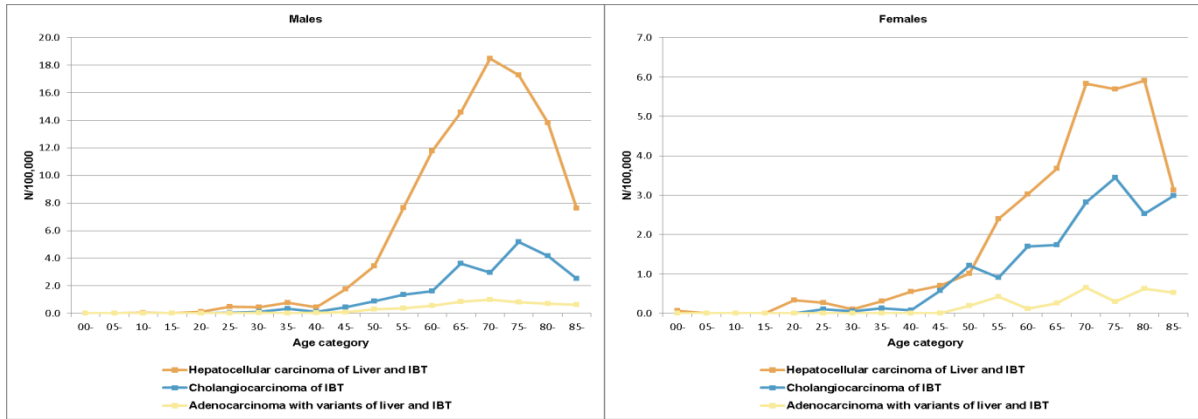
### 8.2 Incidence

- 2,490 new epithelial tumours of the pancreas are diagnosed in the Flemish Region between 2001 and 2010.
- The male/female ratio is 2.4.
- RARECARE differentiates between six rare entities:
  - Hepatocellular carcinoma of liver and IBT is the most common subtype with 1,690 new diagnoses. Incidence rates in males are much higher than females (M/F ratio = 3.0).



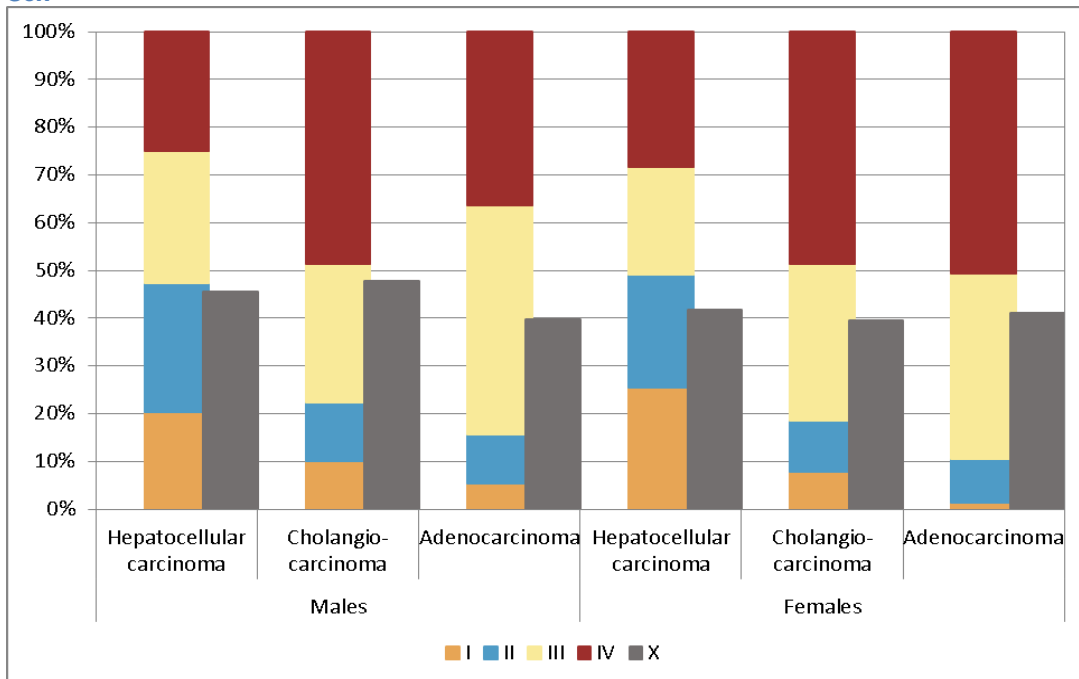
- 522 new cases of cholangiocarcinoma are diagnosed. The incidence rates for cholangiocarcinoma are more comparable between the sexes (M/F ratio = 1.2).
- Adenocarcinoma represents 107 cases (M/F ratio = 2.0).
- The remaining subtypes do not occur in the Flemish Region, with the exception of 1 bile duct cystadenocarcinoma.

**Figure 23. Epithelial Tumours of Liver and Intrahepatic Bile Tract: Age Specific Incidence in Males and Females**



- Incidence rates for hepatocellular carcinoma increase from the age of 50 years.
- In females, the rates for cholangiocarcinoma are comparable with the rates of hepatocellular carcinoma until the age of 50 years.

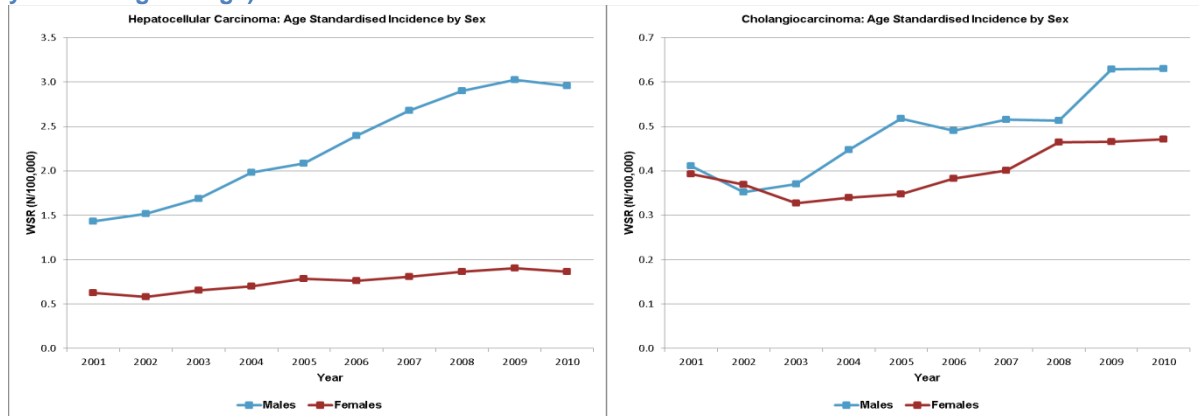
**Figure 24. Epithelial Tumours of Liver and Intrahepatic Bile Tract: Stage Distribution by Morphology and Sex**



- Information on the stage is available for 50-60% of all diagnoses.
- Hepatocellular carcinomas have a prognostic better stage distribution (~50% stage I and II) than the other 2 subtypes (~15-20% stage I and II).

## 8.3 Trends

Figure 25. Hepatocellular Carcinoma and Cholangiocarcinoma: Age-Standardised Incidence by Sex (three year moving average)



- Significant incidence increases for hepatocellular carcinomas are observed in males and females.
- Cholangiocarcinomas increase significantly in males, the increase in females is not significant.
- For both subtypes, the rates increase two times faster in males than in females.

## 8.4 Survival

### 8.4.1 Overall Survival

Table 24. Epithelial Tumours of Liver and Intrahepatic Bile Tract – 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 LIVER AND INTRAHEPATIC BILE TRACT (IBT)	2,216	44.4	24.6	16.9	11.4	[15.2 ; 18.6]	45.5	26.4	18.9	14.2	[17.0 ; 20.9]
Hepatocellular carcinoma of liver and IBT	1,489	48.4	29.3	20.5	14.1	[18.3 ; 22.8]	49.5	31.2	22.8	17.2	[20.3 ; 25.4]
Cholangiocarcinoma of IBT	462	38.5	16.9	10.4	6.4	[7.6 ; 13.7]	39.5	18.2	11.7	8.2	[8.6 ; 15.4]
Adenocarcinoma with variants of liver and IBT	101	38.6	14.4	6.0	4.0	[2.1 ; 12.8]	39.7	15.3	6.6	4.8	[2.3 ; 14.2]
Undifferentiated carcinoma of liver and IBT	0	-	-	-	-	-	-	-	-	-	-
Squamous cell carcinoma with variants of liver and IBT	0	-	-	-	-	-	-	-	-	-	-
Bile duct cystadenocarcinoma of IBT	1	*	*	*	*	*	*	*	*	*	*

- Survival for patients diagnosed with an epithelial tumour of the liver or intrahepatic bile tract is poor with less than half of the patients surviving the first year and less than 20% surviving five years.
- Prognosis is highly influenced by the histological subtype with a better prognosis for hepatocellular carcinomas and a worse prognosis for cholangiocarcinoma or adenocarcinoma.

## 8.4.2 Survival by Sex

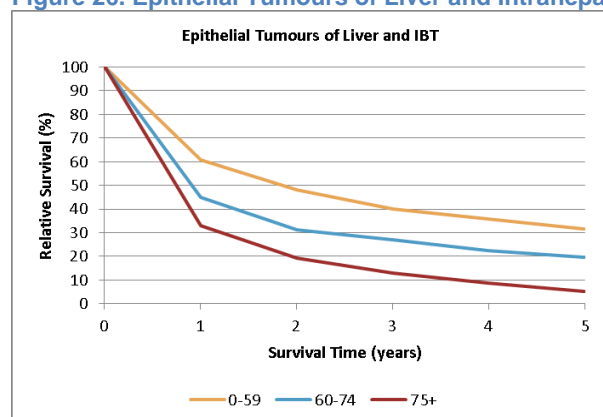
Table 25. Epithelial Tumours of Liver and Intrahepatic Bile Tract – Survival by Sex

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
<b>EPITHELIAL TUMOURS OF LIVER AND INTRAHEPATIC BILE TRACT (IBT)</b>									
	1,438	45.4	25.2	17.3	[15.2 ; 19.6]	46.6	27.2	19.7	[17.3 ; 22.2]
Hepatocellular carcinoma of liver and IBT	1,065	49.0	28.6	20.1	[17.5 ; 22.9]	50.2	30.7	22.6	[19.7 ; 25.7]
Cholangiocarcinoma of IBT	224	36.2	16.4	9.8	[6.1 ; 14.6]	37.2	18.2	11.6	[7.2 ; 17.2]
Adenocarcinoma with variants of liver and IBT	61	42.6	18.5	7.4	[2.2 ; 17.1]	43.9	20.0	8.4	[2.4 ; 19.4]
Undifferentiated carcinoma of liver and IBT	0	-	-	-	-	-	-	-	-
Squamous cell carcinoma with variants of liver and IBT	0	-	-	-	-	-	-	-	-
Bile duct cystadenocarcinoma of IBT	0	-	-	-	-	-	-	-	-
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
<b>EPITHELIAL TUMOURS OF LIVER AND INTRAHEPATIC BILE TRACT (IBT)</b>									
	778	42.4	23.5	16.0	[13.3 ; 18.9]	43.4	24.9	17.5	[14.6 ; 20.7]
Hepatocellular carcinoma of liver and IBT	424	46.7	30.9	21.5	[17.3 ; 25.9]	47.7	32.5	23.3	[18.8 ; 28.1]
Cholangiocarcinoma of IBT	238	40.8	17.3	11.0	[7.1 ; 15.7]	41.6	18.2	11.9	[7.7 ; 17.0]
Adenocarcinoma with variants of liver and IBT	40	32.5	7.1	3.6	[0.3 ; 14.9]	33.2	7.5	3.8	[0.3 ; 15.8]
Undifferentiated carcinoma of liver and IBT	0	-	-	-	-	-	-	-	-
Squamous cell carcinoma with variants of liver and IBT	0	-	-	-	-	-	-	-	-
Bile duct cystadenocarcinoma of liver and IBT	1	*	*	*	*	*	*	*	*

- Prognosis is almost the same for males and females for all studied types of epithelial liver and intrahepatic bile duct cancer.

## 8.4.3 Survival by Age Group<sup>1</sup>

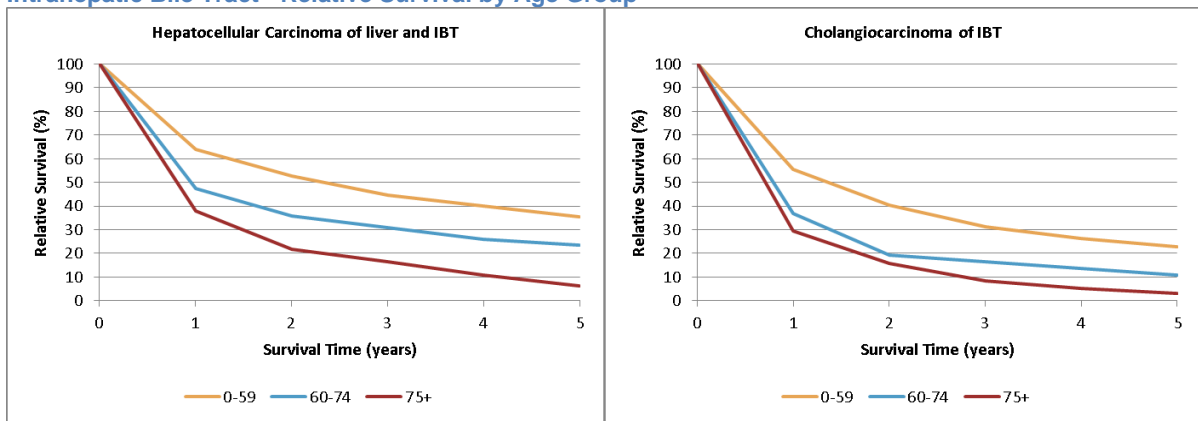
Figure 26. Epithelial Tumours of Liver and Intrahepatic Bile Tract – Relative Survival by Age Group



- Relative survival is inversely related with age. Five-year relative survival is 31.5% for patients in the youngest age group (0-59 years), 19.6% for the middle age group (60-74 years) and only 5.2% for patients of 75 years and older.

<sup>1</sup> Survival by age group is not displayed for the adenocarcinoma because only the age group 60-74 years old has a number at risk higher than 35.

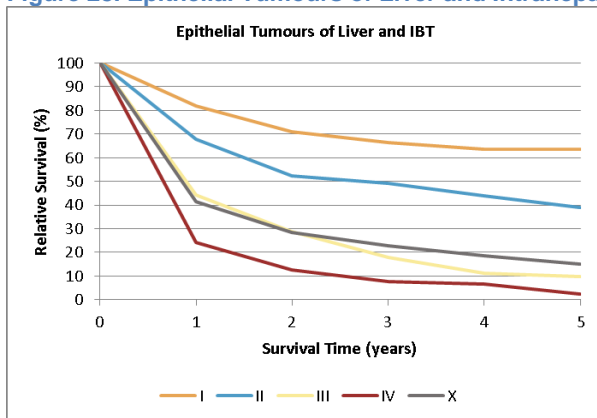
Figure 27. Hepatocellular Carcinoma of Liver and Intrahepatic Bile Tract and Cholangiocarcinoma of Intrahepatic Bile Tract - Relative Survival by Age Group



- For all age groups, survival is worse for cholangiocarcinoma than for hepatocellular carcinoma.

### 8.4.4 Survival by Stage<sup>2</sup>

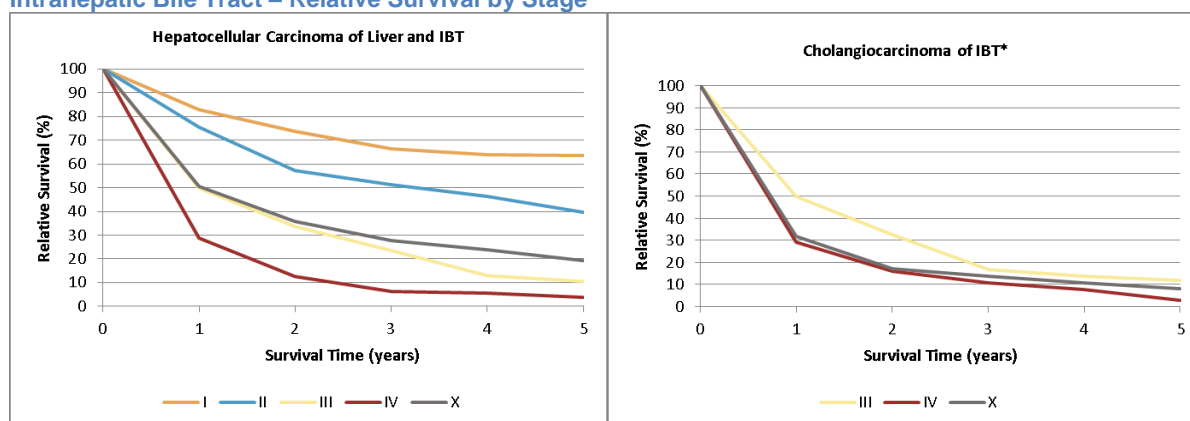
Figure 28. Epithelial Tumours of Liver and Intrahepatic Bile Tract – Survival by Stage



- Survival is relatively good for stage I but remarkably worse for more advanced stages. Long-term survivors with tumour-invaded regional lymph nodes or distant metastases are very rare.

<sup>2</sup> Survival by stage is not displayed for adenocarcinoma because of the low number at risk for all stages except for stage X.

Figure 29. Hepatocellular Carcinoma of Liver and Intrahepatic Bile Tract and Cholangiocarcinoma of Intrahepatic Bile Tract – Relative Survival by Stage



\* Only survival for the higher stages is shown due to low numbers at risk for the lower stages.

- Because of the large proportion of patients diagnosed with an hepatocellular carcinoma, survival by stage for this subgroup is very similar to the survival rates for all epithelial tumours of liver and intrahepatic bile tract together.

## 9. Epithelial Tumours of Gallbladder and Extrahepatic Biliary Tract (EBT)

### 9.1 General Results

Table 26. Epithelial Tumours of Gallbladder and Extrahepatic Biliary Tract: Incidence, Trends, Survival

Flemish Region 2001-2010		Incidence				Trend		Survival		
Both Sexes		R/C	N	CR	WSR	Avg Age	EAPC %	p-value	N at risk	5yr (%)
EPITHELIAL TUMOURS OF GALLBLADDER AND EXTRAHEPATIC BILIARY TRACT (EBT)		R	1,964	3.23	1.41	72	3.1	0.044	1,762	21.7
Adenocarcinoma with variants of gallbladder and EBT		R	1,715	2.82	1.27	71	3.5	0.066	1,531	23.0
Squamous cell carcinoma of gallbladder and EBT		R	11	0.02	0.01	77	*	*	11	*
Males		R/C	N	CR	WSR	Avg Age	EAPC %	p-value	N at risk	5yr (%)
EPITHELIAL TUMOURS OF GALLBLADDER AND EXTRAHEPATIC BILIARY TRACT (EBT)		R	879	2.93	1.47	70	4.6	0.009	771	24.0
Adenocarcinoma with variants of gallbladder and EBT		R	784	2.61	1.33	70	4.9	0.018	685	25.1
Squamous cell carcinoma of gallbladder and EBT		R	2	0.01	0.00	69	*	*	2	*
Females		R/C	N	CR	WSR	Avg Age	EAPC %	p-value	N at risk	5yr (%)
EPITHELIAL TUMOURS OF GALLBLADDER AND EXTRAHEPATIC BILIARY TRACT (EBT)		R	1,085	3.52	1.35	74	1.8	0.227	991	19.9
Adenocarcinoma with variants of gallbladder and EBT		R	931	3.02	1.20	73	2.3	0.218	846	21.3
Squamous cell carcinoma of gallbladder and EBT		R	9	0.03	0.01	79	*	*	9	*

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

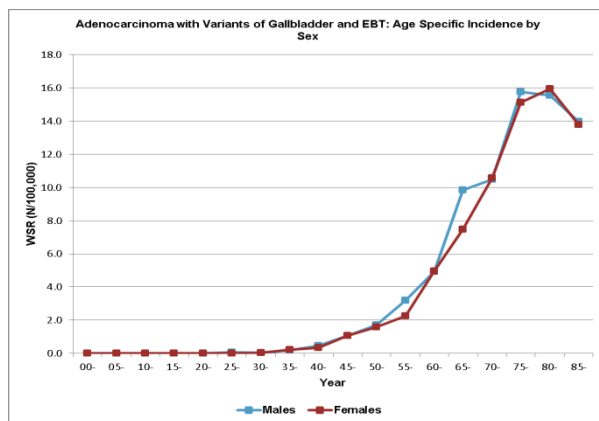
RS: relative survival

AvgAge: average age at diagnosis

## 9.2 Incidence

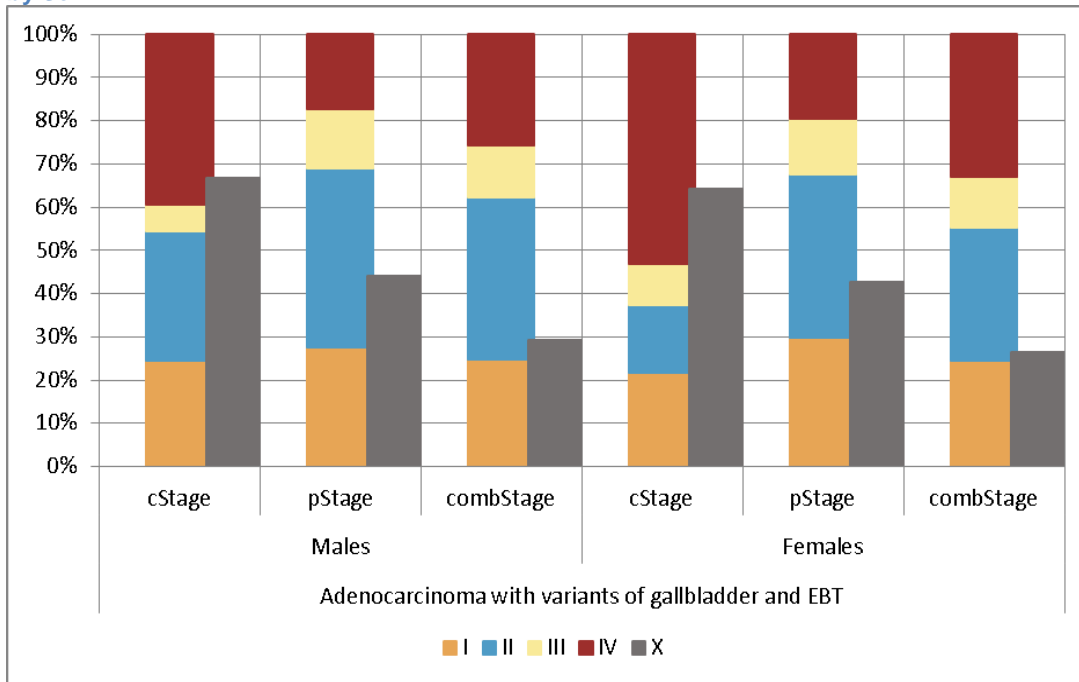
- 1,964 new epithelial tumours of gallbladder and extrahepatic biliary tract are diagnosed in the Flemish Region between 2001 and 2010.
- The male/female ratio is 1.1.
- RARECARE differentiates between two rare entities:
  - Adenocarcinoma (which includes the cholangiocarcinoma) of gallbladder is the most common subtype with 1,715 new diagnoses.
  - Only 11 cases of squamous cell carcinoma are diagnosed between 2001 and 2010 in the Flemish Region.

Figure 30. Adenocarcinoma with Variants of Gallbladder and Extrahepatic Biliary Tract: Age Specific Incidence by Sex



- Incidence rates increase from the age of 45 years in both sexes.
- The age specific incidence rates in males and females are comparable.

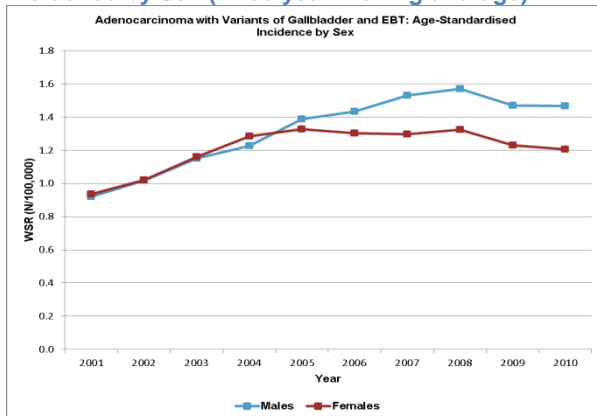
**Figure 31. Adenocarcinoma with Variants of Gallbladder and Extrahepatic Biliary Tract: Stage Distribution by Sex**



- Information on clinical stage is available for 30-40% of all diagnoses, pathological stage information is available in about 55%. Combining clinical and pathological information on stage results in more than 70% of cases with known information on stage.
- In females 15% more clinical stage IV tumours are diagnosed than in males.

### 9.3 Trends

**Figure 32. Adenocarcinoma with Variants of Gallbladder and Extrahepatic Biliary Tract: Age-Standardised Incidence by Sex (three year moving average)**



- Adenocarcinoma of the gallbladder increase significantly in males and non-significantly in females.
- The rates don't show a significant increase over the entire 10 year time period. In males the increase seems to end in 2008, in females the increase ends in 2005 after which the rates remain more stable.

## 9.4 Survival

### 9.4.1 Overall Survival

Table 27. Epithelial Tumours of Gallbladder and Extrahepatic Biliary Tract – 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 GALLBLADDER AND EXTRAHEPATIC BILIARY TRACT (EBT)	1,762	45.9	23.2	18.3	13.7	[16.4 ; 20.2]	47.6	25.8	21.7	19.9	[19.5 ; 24.0]
Adenocarcinoma with variants	1,531	48.6	24.8	19.6	14.6	[17.5 ; 21.8]	50.4	27.4	23.0	20.8	[20.6 ; 25.5]
Squamous cell carcinoma	11	*	*	*	*	*	*	*	*	*	*

- One year after diagnosis, less than half of the patients is still alive.
- Survival decreases rapidly to a 5-year relative survival of only 21.7%.
- Because the majority of patients with an epithelial tumour of the gallbladder and extrahepatic biliary tract are diagnosed with an adenocarcinoma, prognosis of this subtype is very similar to the prognosis of all epithelial tumours together.

### 9.4.2 Survival by Sex

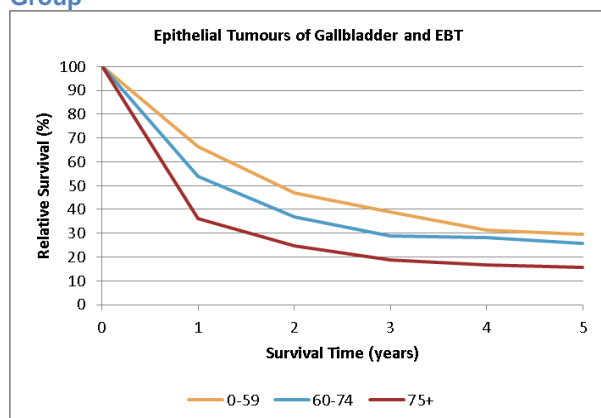
Table 28. Epithelial Tumours of Gallbladder and Extrahepatic Biliary Tract – Survival by Sex

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 GALLBLADDER AND EXTRAHEPATIC BILIARY TRACT (EBT)	771	50.8	25.6	20.3	[17.4 ; 23.4]	52.9	28.3	24.0	[20.6 ; 27.7]
Adenocarcinoma with variants	685	53.6	27.1	21.3	[18.1 ; 24.6]	55.6	30.0	25.1	[21.4 ; 29.0]
Squamous cell carcinoma	2	*	*	*	*	*	*	*	*
Females	N at risk	Observed Survival				Relative Survival			
EPITHELIAL TUMOURS OF GALLBLADDER AND EXTRAHEPATIC BILIARY TRACT (EBT)	991	42.0	21.4	16.7	[14.4 ; 19.3]	43.6	23.8	19.9	[17.1 ; 22.9]
Adenocarcinoma with variants	846	44.5	22.9	18.2	[15.6 ; 21.1]	46.1	25.3	21.3	[18.2 ; 24.6]
Squamous cell carcinoma	9	*	*	*	*	*	*	*	*

- In contrast to most tumours, survival of epithelial tumours of the gallbladder and extrahepatic biliary tract is higher in males than females.
- The difference between males and females is largest at one year after diagnosis and becomes slightly smaller after a longer follow-up period.

### 9.4.3 Survival by Age Group

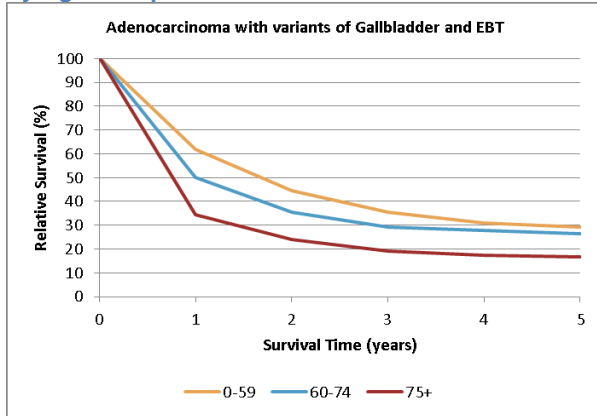
Figure 33. Epithelial Tumours of Gallbladder and Extrahepatic Biliary Tract – Relative Survival by Age Group





- Survival is inversely related with age, although the difference in survival between patients aged under 59 years and between 60 and 74 years decreases after three years of follow-up.
- Prognosis is worse for patients of 75 years and above.

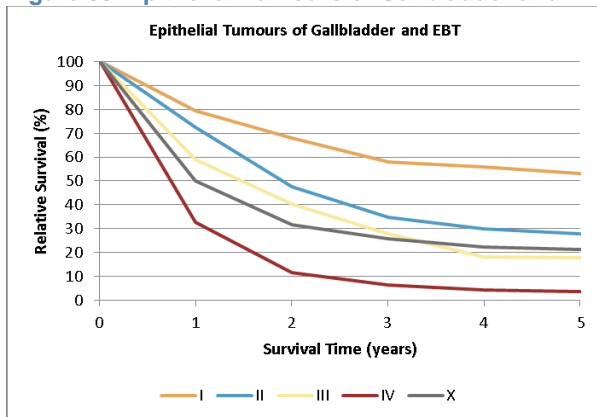
**Figure 34. Adenocarcinoma with Variants of Gallbladder and Extrahepatic Biliary Tract – Relative Survival by Age Group**



- Because almost all patients with an epithelial tumour of the gallbladder and extrahepatic biliary tract are diagnosed with an adenocarcinoma, survival hardly differs between these two.

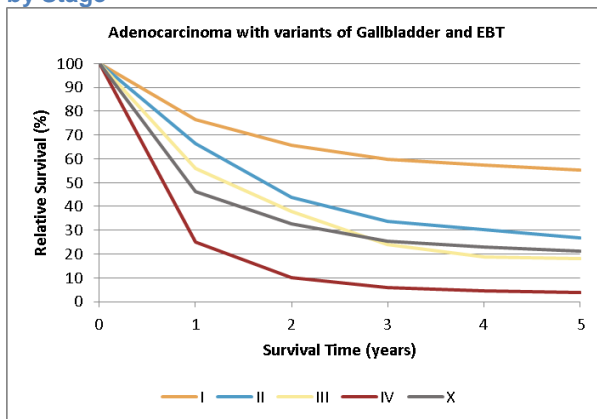
#### 9.4.4 Survival by Stage

**Figure 35. Epithelial Tumours of Gallbladder and Extrahepatic Biliary Tract – Relative Survival by Stage**



- Survival is much better for patients with a stage I tumour (5-year relative survival: 53.0%) than for other stages.
- 5-year relative survival is almost negligible for stage IV tumours (3.9%).

Figure 36. Adenocarcinoma with variants of Gallbladder and Extrahepatic Biliary Tract – Relative Survival by Stage



- Prognosis of patients diagnosed with an adenocarcinoma is almost the same as the above described results of all epithelial tumours of the gallbladder and extrahepatic biliary tract together.

## References

1. Bohanes P, Yang D, Chhibar RS et al. Influence of sex on the survival of patients with esophageal cancer. J Clin Oncol 2012; 30: 2265-2272.