Rare Tumours of Female Genital Organs

1. Epithelial Tumours of Corpus Uteri

1.1 General Results

Table 1. Epithelial Tumours of Corpus Uteri: Incidence, Trends, Survival

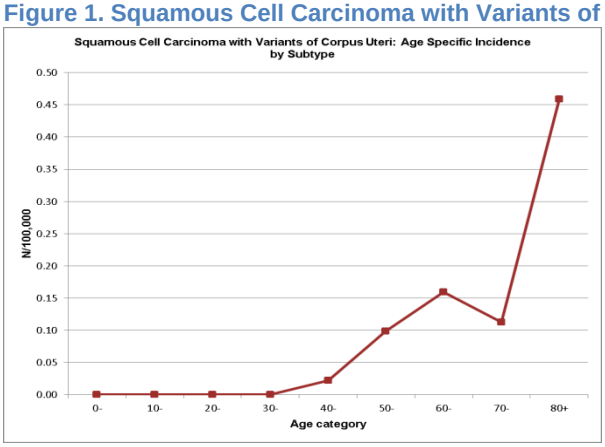
Flemish Region 2001-2010		Incidence				Trend		Survival	
Females						EAPC		Relative survival	
	R/C	N	CR	WSR	Avg Age	%	p-value	N at risk	5yr (%)
EPITHELIAL TUMOURS OF CORPUS UTERI	C	7,629	24.76	12.45	68	-0.8	0.102	6,996	83.9
Adenocarcinoma with variants of corpus uteri	C	7,515	24.39	12.29	68	-0.5	0.277	6,893	84.3
Squamous cell carcinoma with variants of corpus uteri	R	21	0.07	0.03	70	*	*	15	*
Adenoid cystic carcinoma of corpus uteri	R	0	-	-	-	-	-	-	-
Transitional cell carcinoma of corpus uteri	R	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

1.2 Incidence

- 7,629 new epithelial tumours of corpus uteri are diagnosed in the Flemish Region between 2001 and 2010.
- Of the three RARECARE entities, only squamous cell carcinoma is diagnosed. Neither adenoid cystic carcinoma nor transitional cell carcinoma are observed in the Flemish Region between 2001 and 2010.

Figure 1. Squamous Cell Carcinoma with Variants of Corpus Uteri: Age Specific Incidence by Subtype



- From the age of 40 years old, incidence rates for squamous cell carcinoma of corpus uteri start to increase.

Belgian Cancer Registry

1.3 Overall Survival

Table 2. Epithelial Tumours of Corpus Uteri – Overall Survival

	N	Observed Survival					Relative Survival				
	at risk	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 CORPUS UTERI	6,996	92.2	81.5	74.8	62.3	[73.7 ; 75.9]	94.2	87.1	83.9	80.4	[82.7 ; 85.1]
Adenocarcinoma with variants	6,893	92.6	81.9	75.3	62.7	[74.2 ; 76.3]	94.6	87.5	84.3	80.9	[78.8 ; 82.9]
Squamous cell carcinoma with variants	15	*	*	*	*	*	*	*	*	*	*
Adenoid cystic carcinoma	-	-	-	-	-	-	-	-	-	-	-
Transitional cell carcinoma	-	-	-	-	-	-	-	-	-	-	-

- Prognosis of patients with an epithelial tumour of corpus uteri is good, with a 10-year relative survival of more than 80%.

2. Epithelial Tumours of Cervix Uteri

2.1 General Results

Table 3. Epithelial Tumours of Cervix Uteri: Incidence, Trends, Survival

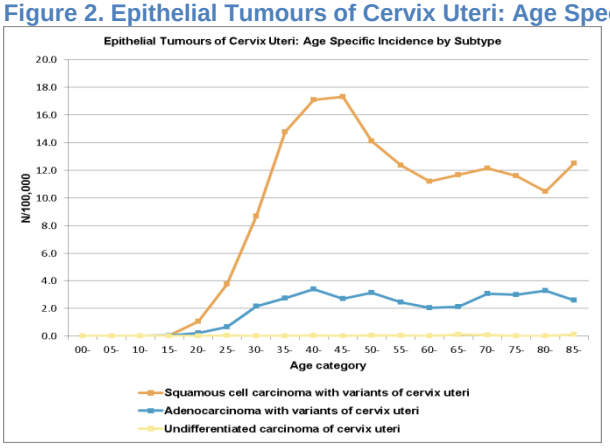
Flemish Region 2001-2010		Incidence				Trend		Survival	
Females						EAPC		Relative survival	
	R/C	N	CR	WSR	Avg Age	%	p-value	N at risk	5yr (%)
EPITHELIAL TUMOURS OF CERVIX UTERI	C	3,635	11.80	8.18	53	-1.6	0.084	3,456	69.6
Squamous cell carcinoma with variants of cervix uteri	C	2,806	9.11	6.33	53	-0.5	0.668	2,674	70.8
Adenocarcinoma with variants of cervix uteri	R	578	1.88	1.26	54	0.4	0.750	543	67.1
Undifferentiated carcinoma of cervix uteri	R	8	0.03	0.02	60	*	*	7	*

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

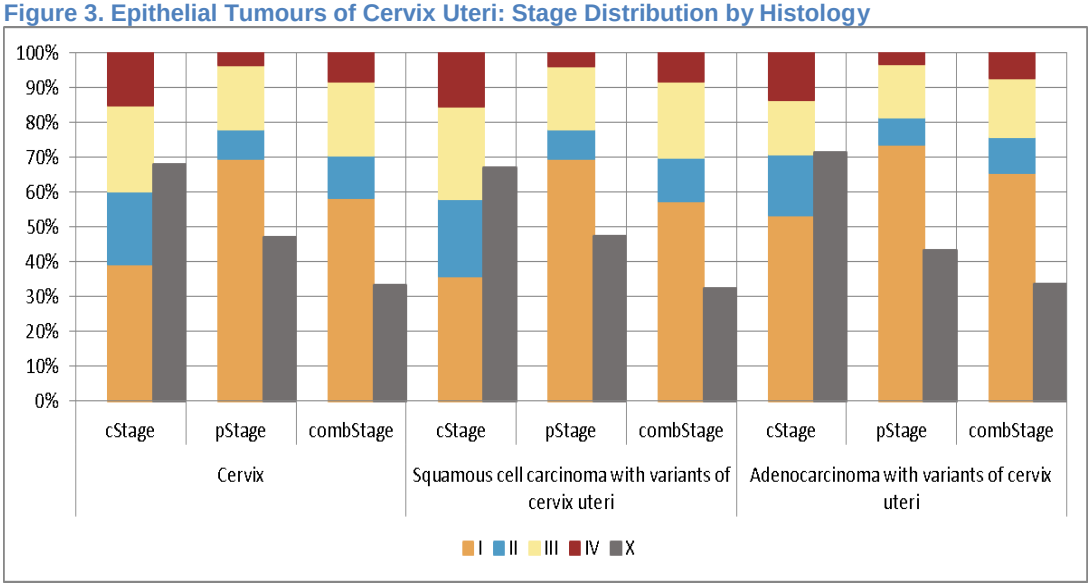
- 3,635 new epithelial tumours of cervix uteri are diagnosed in the Flemish Region between 2001 and 2010.
- RARECARE defines three rare entities:
  - Squamous cell carcinoma should be classified as common in the Flemish Region when following the RARECARE definition for rare tumours (incidence < 6 / 100.000).
  - Adenocarcinoma represents about 16% of the epithelial tumours of cervix uteri. Almost all adenocarcinoma occur in the endocervix.
  - Only 8 cases of undifferentiated carcinoma are registered in the Flemish Region between 2001 and 2010.

Figure 2. Epithelial Tumours of Cervix Uteri: Age Specific Incidence by Subtype



- Incidence rates for squamous cell carcinoma start to increase from an early age. A peak incidence is observed around the age of 40-50 years.
- Adenocarcinoma incidence rates start to increase around the age of 30 years. The rates remain fairly stable with age.
- Overall, the age specific adenocarcinoma incidence rates are three to four times lower than the rates for squamous cell carcinoma.

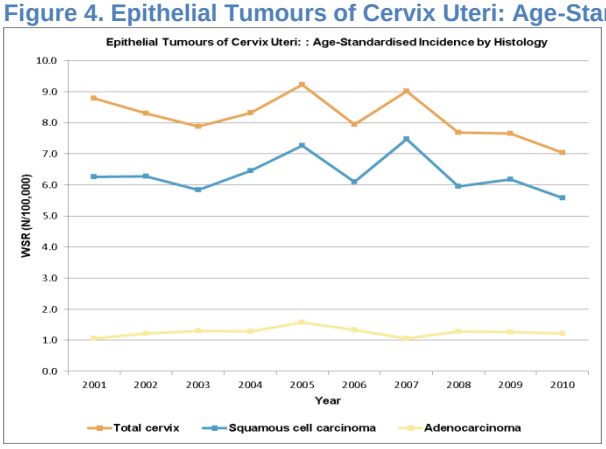
Figure 3. Epithelial Tumours of Cervix Uteri: Stage Distribution by Histology



- Information on stage (combined) is missing in about 30% of all cervical cancers, with a higher percentage of missing information on clinical stage at diagnosis (70%) than on pathological stage (50%).
- Pathological stage is more often stage I than clinically (respectively 70% and 40%).
- Although overall distribution of combined staging for squamous cell carcinoma and adenocarcinoma are similar, the latter histology type has a higher proportion of clinical stage I tumours (>50% and 35% respectively).

2.3 Trends

Figure 4. Epithelial Tumours of Cervix Uteri: Age-Standardised Incidence by Histology



- No significant trends are observed in the Flemish Region between 2001 and 2010.

2.4 Survival

2.4.1 Overall Survival

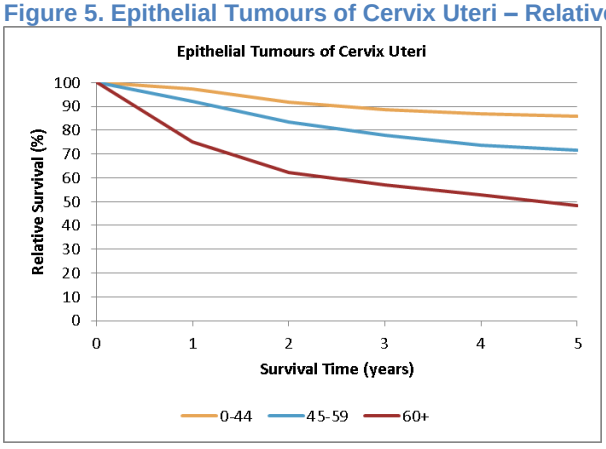
Table 4. Epithelial Tumours of Cervix Uteri – Overall Survival

	N	Observed Survival					Relative Survival				
	at risk	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 CERVIX UTERI	3,456	87.8	73.3	66.8	59.4	[65.2 ; 68.4]	88.6	75.2	69.6	64.8	[67.9 ; 71.3]
Squamous cell carcinoma with variants	2,674	88.4	74.3	67.9	60.9	[66.0 ; 69.8]	89.3	76.2	70.8	66.5	[68.8 ; 72.7]
Adenocarcinoma with variants	543	86.5	71.6	64.2	53.9	[59.7 ; 68.3]	87.5	73.7	67.1	59.7	[62.5 ; 71.4]
Undifferentiated carcinoma	7	*	*	*	*	*	*	*	*	*	*

- Survival of patients with an epithelial tumour of cervix uteri is rather good with a 1-year relative survival of 88.6% and 5-year relative survival of almost 70%.
- Survival is better for squamous cell carcinoma than for adenocarcinoma and this difference becomes larger with a longer follow-up.

2.4.2 Survival by Age Group

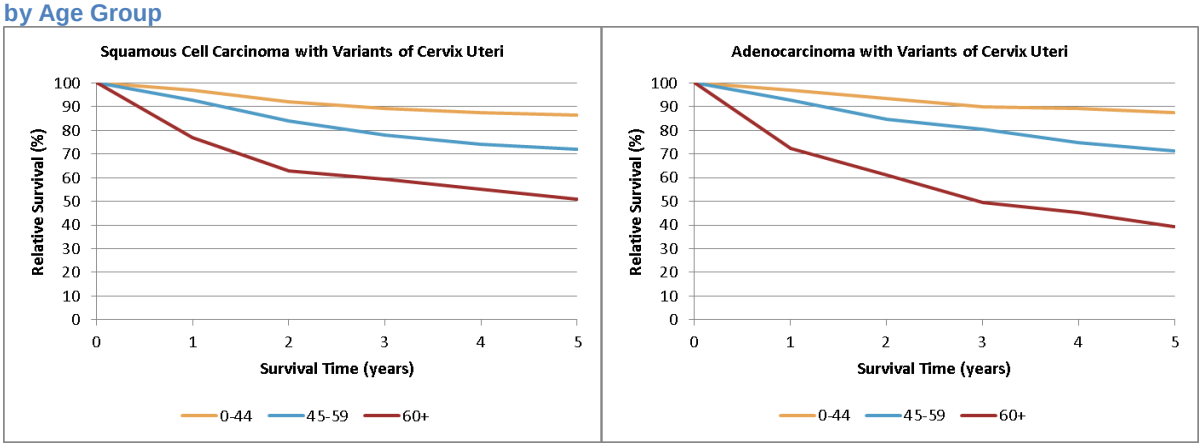
Figure 5. Epithelial Tumours of Cervix Uteri – Relative Survival by Age Group



- Survival is dependent on the age of the patient at diagnosis. Patients younger than 45 have a 5-year relative survival of 85.7%, while this survival rate is lower for patients aged between

45 and 59 years old (71.6%). For patients aged 60 years and above, 5-year relative survival is only 48.3%.

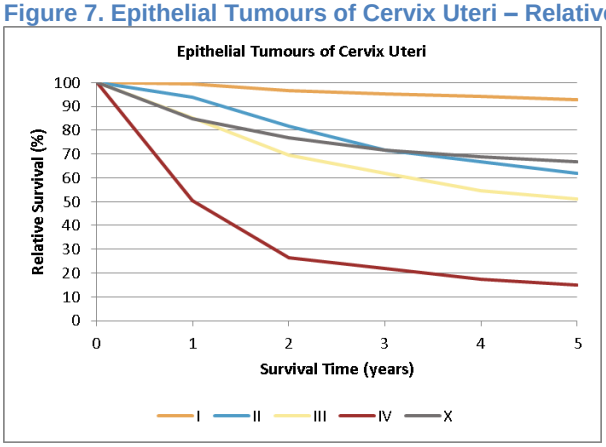
Figure 6. Squamous Cell Carcinoma and Adenocarcinoma with Variants of Cervix Uteri – Relative Survival by Age Group



- Survival is similar between squamous cell carcinoma and adenocarcinoma for the age groups 0-44 years old and 45-59 years old. For patients in the age group 60 years and older, survival is much worse for the adenocarcinoma (5-year relative survival: 39.5% versus 50.9% for squamous cell carcinoma).

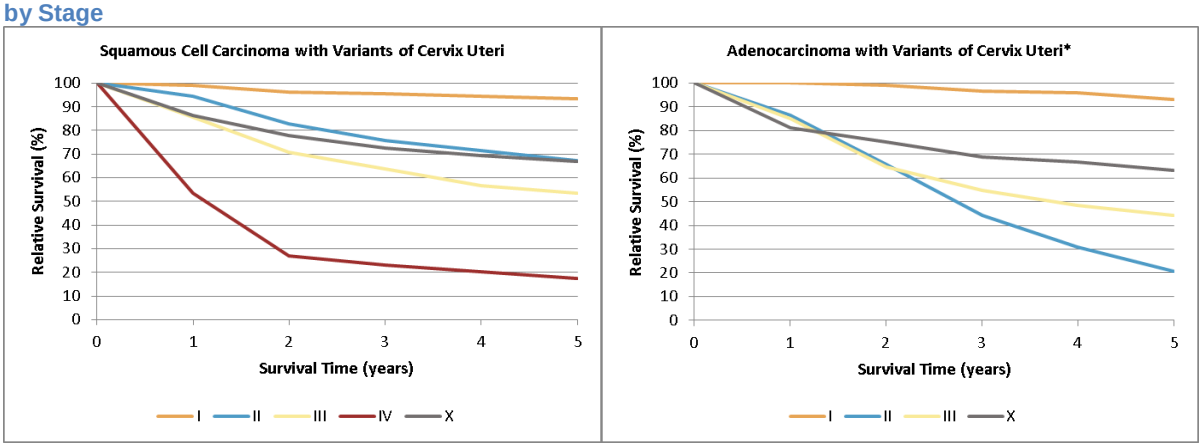
2.4.3 Survival by Stage

Figure 7. Epithelial Tumours of Cervix Uteri – Relative Survival by Stage



- Survival is highly dependent on the stage of the tumour, with a five-year relative survival equal to 92.7% for stage I tumours and of only 15.0% for stage IV tumours.

Figure 8. Squamous Cell Carcinoma and Adenocarcinoma with Variants of Cervix Uteri – Relative Survival by Stage



* Survival of stage IV is not shown because the number at risk is lower than 35.

- Survival is only similar between squamous cell carcinoma and adenocarcinoma for the low stage I tumours.
- The difference between squamous cell carcinoma and adenocarcinoma is enormous for stage II tumours. Survival is much lower than expected for a stage II adenocarcinoma. This is even lower than the survival for stage III adenocarcinoma.
- For stage III tumours, squamous cell carcinomas have a more than 10% better survival than adenocarcinoma (55.6% versus 44.1%).

3. Mixed Epithelial and Mesenchymal Tumours of Uterus[1]

3.1 General Results

Table 5. Mixed Epithelial and Mesenchymal Tumours of Uterus: Incidence, Trends, Survival

Flemish Region 2001-2010	Incidence					Trend		Survival	
Females						EAPC		Relative survival	
	R/C	N	CR	WSR	Avg Age	%	p-value	N at risk	5yr (%)
MIXED EPITHELIAL AND MESENCHYMAL TUMOURS OF UTERUS	R	421	1.37	0.63	69	-0.1	0.965	270	38.5

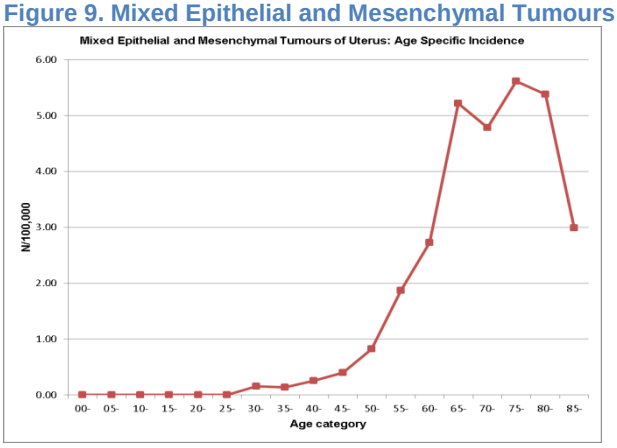
R/C: Rare or common
CR: Crude rate (N/100,000 person years)
WSR: age-standardised rate, using the world population (N/100,000 person years)
EAPC: estimated annual percentage change
RS: relative survival
AvgAge: average age at diagnosis

3.2 Incidence

- 421 new epithelial tumours of mixed epithelial and mesenchymal tumours of uterus are diagnosed in the Flemish Region between 2001 and 2010.
- This layer 1 group is classified as rare by RARECARE, no additional level 2 entities are defined.

[1] Survival by stage is not shown because this type of tumours cannot be staged according to the TNM guidelines.

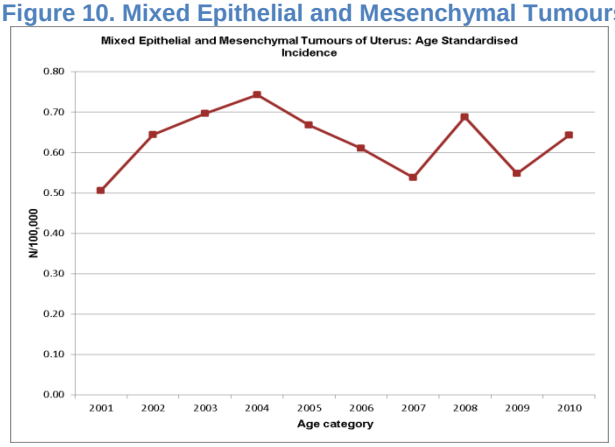
Figure 9. Mixed Epithelial and Mesenchymal Tumours of Uterus: Age Specific Incidence



- Age specific incidence rates increase from the age of 40 years old.

3.3 Trends

Figure 10. Mixed Epithelial and Mesenchymal Tumours of Uterus: Age-Standardised Incidence



- No significant changes are observed over time.

3.4 Survival

3.4.1 Overall Survival

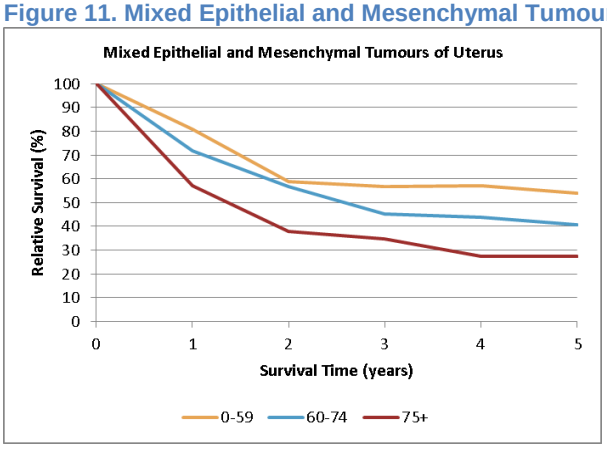
Table 6. Mixed Epithelial and Mesenchymal Tumours of Uterus – Overall Survival

	N	Observed Survival					Relative Survival				
	at risk	1 year	3 year	5 year	10 year	5 year CI	1 year	3 year	5 year	10 year	5 year CI
MIXED EPITHELIAL AND MESENCHYMAL TUMOURS OF UTERUS	270	66.7	40.7	34.2	27.7	[28.3 ; 40.2]	68.2	43.5	38.5	34.4	[31.9 ; 45.2]

- Prognosis is poor for patients diagnosed with a mixed epithelial and mesenchymal tumour of the uterus, with only about two-third of patients surviving the first year and a relative survival at 5 years of only 38.5%.

3.4.2 Survival by Age Group

Figure 11. Mixed Epithelial and Mesenchymal Tumours of Uterus – Relative Survival by Age Group



- Survival is inversely related with age; survival is highest for the youngest age group and lowest for the oldest age group.

4. Epithelial Tumours of Ovary and Fallopian Tube

4.1 General Results

Table 7. Epithelial Tumours of Ovary and Fallopian Tube: Incidence, Trends, Survival

Flemish Region 2001-2010		Incidence				Trend		Survival	
Females						EAPC		Relative survival	
	R/C	N	CR	WSR	Avg Age	%	p-value	N at risk	5yr (%)
EPITHELIAL TUMOURS OF OVARY AND FALLOPIAN TUBE	C	5,280	17.14	9.05	65	-3.6	<0.001	4,588	43.0
Adenocarcinoma with variants of ovary	C	4,055	13.16	6.91	66	-2.9	0.002	3,669	40.7
Mucinous adenocarcinoma of ovary	R	551	1.79	1.10	60	-6.0	0.003	504	63.3
Clear cell adenocarcinoma of ovary	R	218	0.71	0.42	62	0.7	0.794	206	64.9
Adenocarcinoma with variants of fallopian tube	R	112	0.36	0.22	62	13.8	0.007	95	64.3

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

- 5,280 new epithelial tumours of ovary and fallopian tube are diagnosed in the Flemish Region between 2001 and 2010.
- RARECARE defines four rare entities:
  - Adenocarcinoma with variants of ovary in the Flemish Region should be classified as common when following the RARECARE definition for rare tumours (< 6 / 100.000). This group entails a broad histological variety of adenocarcinoma types.
  - Mucinous adenocarcinoma represents 551 new diagnoses.
  - Clear cell adenocarcinoma is diagnosed in 218 patients.

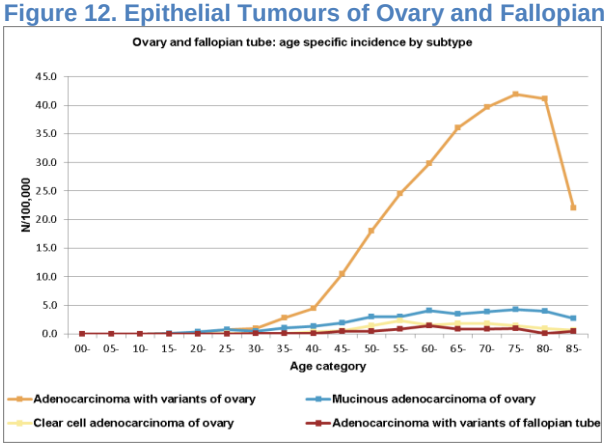
- A total of 112 patients are registered with an adenocarcinoma with variants of fallopian tube.

Table 8. Morphology Distribution of Epithelial Tumours of Ovary and Fallopian Tube

Flemish Region 2001-2010	Ovary		Fallopian tube	
Papillary serous cystadenocarcinoma	1,553	32.2%	40	35.1%
Serous cystadenocarcinoma, NOS	849	17.6%	38	33.3%
Adenocarcinoma NOS	687	14.2%	19	16.7%
Mucinous adenocarcinoma	551	11.4%	1	0.9%
Endometrioid adenocarcinoma, NOS	403	8.4%	10	8.8%
Serous surface papillary carcinoma	239	5.0%	0	0.0%
Clear cell adenocarcinoma	218	4.5%	1	0.9%
Papillary adenocarcinoma, NOS	114	2.4%	1	0.9%
Papillary cystadenocarcinoma	82	1.7%	2	1.8%
Adenocarcinoma other specified	61	1.3%	2	1.8%
Cystadenocarcinoma, NOS	46	1.0%	0	0.0%
Clear cell adenocarcinofibroma	9	0.2%	0	0.0%
Serous adenocarcinofibroma	9	0.2%	0	0.0%
Mucinous adenocarcinofibroma	2	0.0%	0	0.0%
Endometrioid adenofibroma malignant	1	0.0%	0	0.0%
Total	**4,824**	**100.0%**	**114**	**100.0%**

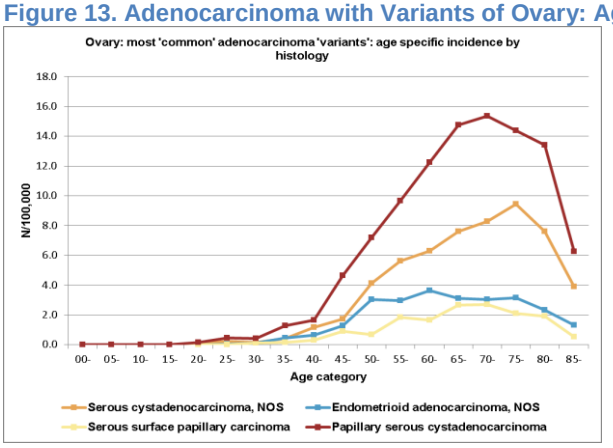
- A wide variety of adenocarcinoma subtypes are observed in the ovary and fallopian tube.
- Papillary serous and serous cystadenocarcinoma are the most frequent histological subtypes.

Figure 12. Epithelial Tumours of Ovary and Fallopian Tube: Age Specific Incidence by Subtype



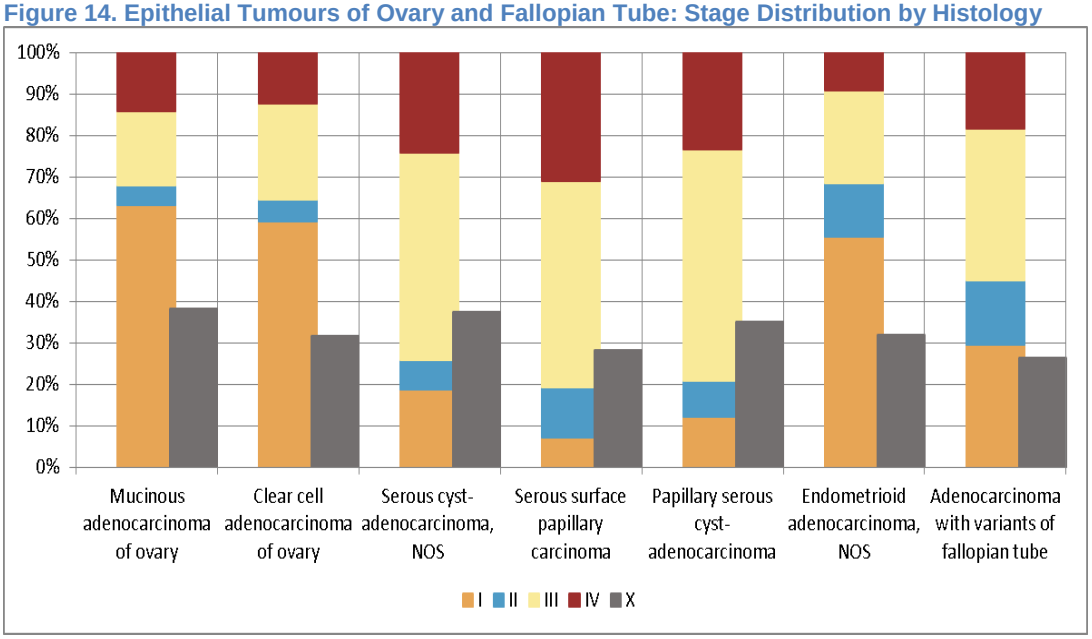
- Age-specific incidences start to increase from 35 years onwards and show a steep rise for adenocarcinoma with variant of ovary, while the incidences for the other subtypes stay low.

Figure 13. Adenocarcinoma with Variants of Ovary: Age Specific Incidence by Subtype



- The different types of adenocarcinoma have a similar pattern in age specific incidence rates. Around the age of 40-50 years age specific incidence rates increase until the age of 65-75 years.

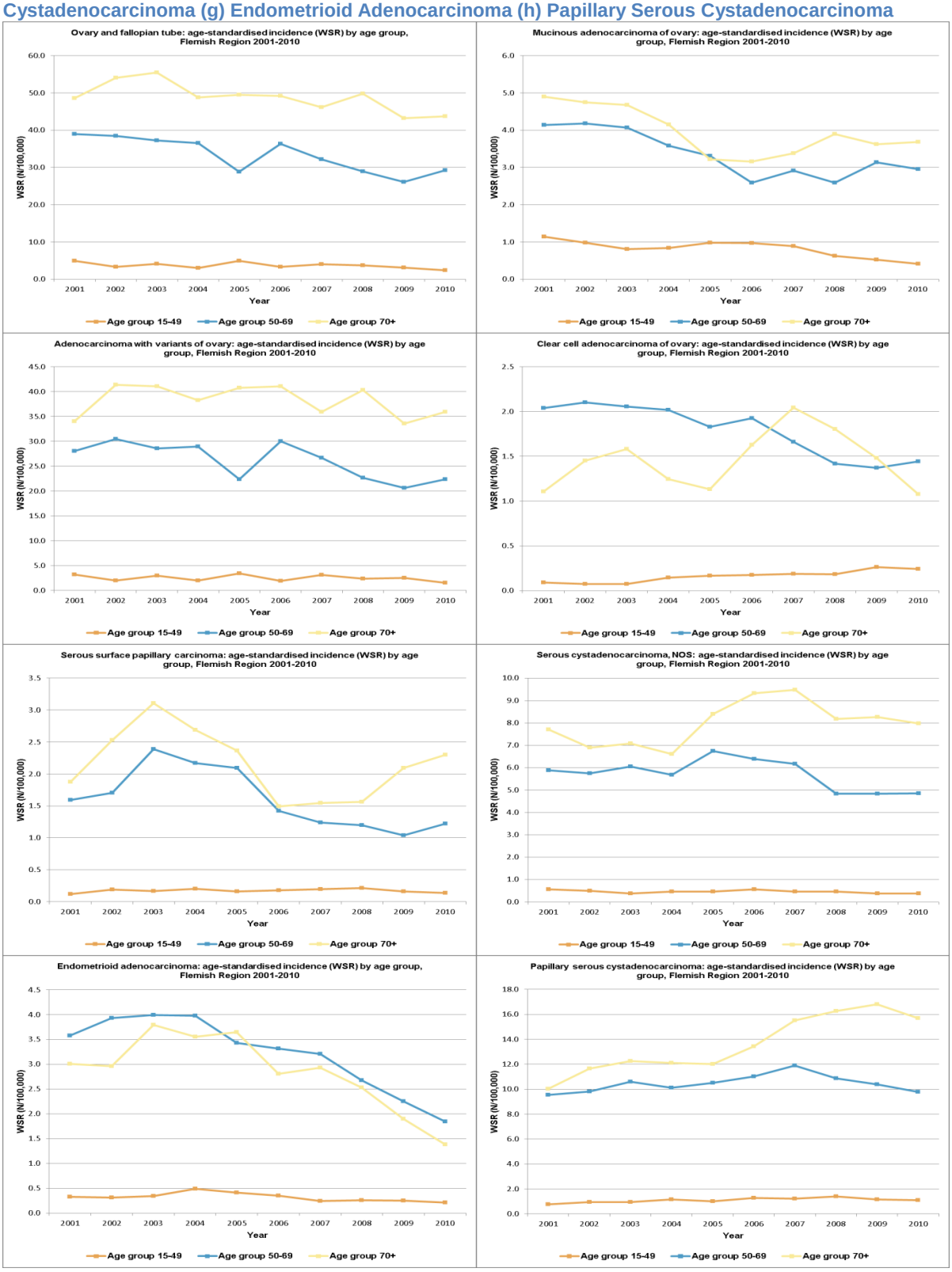
Figure 14. Epithelial Tumours of Ovary and Fallopian Tube: Stage Distribution by Histology



- Information on stage was available in more than 60% of all new diagnoses.
- Mucinous, clear cell and endometrioid carcinoma have a prognostic better stage distribution (30-35% stage III-IV) than the three serous type adenocarcinoma (75-80% stage III-IV).

4.3 Trends

Figure 15. Age-Standardised Incidence by Age Groups for (a) All Epithelial Tumours of Ovary and Fallopian Tube (b) Mucinous Adenocarcinoma of Ovary (c) Adenocarcinoma with Variants of Ovary (d) Clear Cell Adenocarcinoma of Overay (e) Serous Surface Papillary Carcinoma (f) Serous Cystadenocarcinoma (g) Endometrioid Adenocarcinoma (h) Papillary Serous Cystadenocarcinoma



- The incidence rates for tumours of ovary and fallopian tube decrease annually with 4.1% (p = 0.093) in females between 15 and 49 years old. A significant decrease is observed for mucinous adenocarcinoma and endometrioid carcinoma. Clear cell and papillary serous cystadenocarcinoma on the other hand show a significant increase.
- For females between 50 and 69 years of age, a significant decrease is observed for tumours of ovary and fallopian tube (EAPC = -4.0% [p = 0.002]). This decrease is observed in all subtypes except for papillary serous cystadenocarcinoma where no changes in incidence rates are observed.
- The incidence rates for females of 70 years and older decrease with 1.9% annually (p = 0.014). This decrease is mainly observed for mucinous adenocarcinoma, endometrioid carcinoma and serous surface papillary carcinoma. A significant increase is observed for papillary serous cystadenocarcinoma.

4.4 Survival

4.4.1 Overall Survival

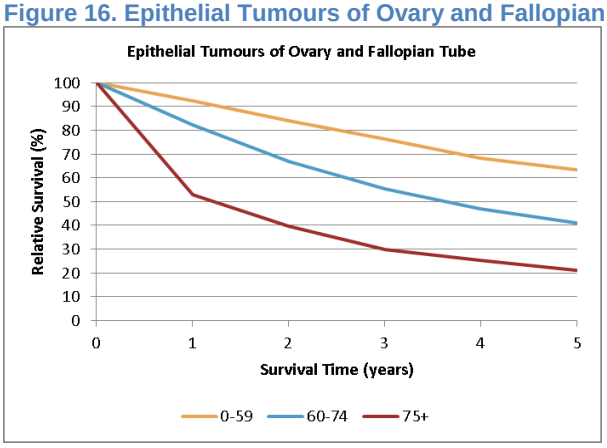
Table 9. Epithelial Tumours of Ovary and Fallopian Tube – Overall Survival

	N	Observed Survival					Relative Survival				
	at risk	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 OVARY AND FALLOPIAN TUBE	4,588	75.8	52.3	39.6	26.6	[38.1 ; 41.1]	77.2	55.1	43.0	31.6	[41.4 ; 44.6]
Adenocarcinoma with variants of ovary	3,669	77.9	52.1	37.6	23.0	[36.0 ; 39.3]	79.3	54.6	40.7	27.1	[38.9 ; 42.5]
Mucinous adenocarcinoma of ovary	504	77.7	64.7	59.0	50.9	[54.4 ; 63.3]	78.9	67.6	63.3	59.3	[58.4 ; 67.9]
Clear cell adenocarcinoma of ovary	206	88.4	67.1	60.5	49.2	[53.0 ; 67.2]	89.5	69.8	64.9	57.5	[56.8 ; 72.0]
Adenocarcinoma with variants of fallopian tube	95	94.7	70.5	60.7	53.8	[48.8 ; 70.6]	96.0	73.1	64.3	60.5	[60.5 ; 44.5]

- Survival is rather poor for patients diagnosed with an epithelial tumour of the ovary or fallopian tube, with a 5-year observed survival of only 39.6% and a 5-year relative survival of only 43.0%.
- Survival patterns differ for the different subtypes: at 1 year after diagnosis, relative survival is similar for the adenocarcinoma with variants and the mucinous adenocarcinoma of ovary (about 79%), but better for clear cell adenocarcinoma of ovary and adenocarcinoma with variants of fallopian tube (89.5% and 96.0%, respectively). This is different for longer follow-up periods (5-year follow-up) where survival is worse for adenocarcinoma with variants of ovary (40.7%) than the other subtypes (all about 64%).

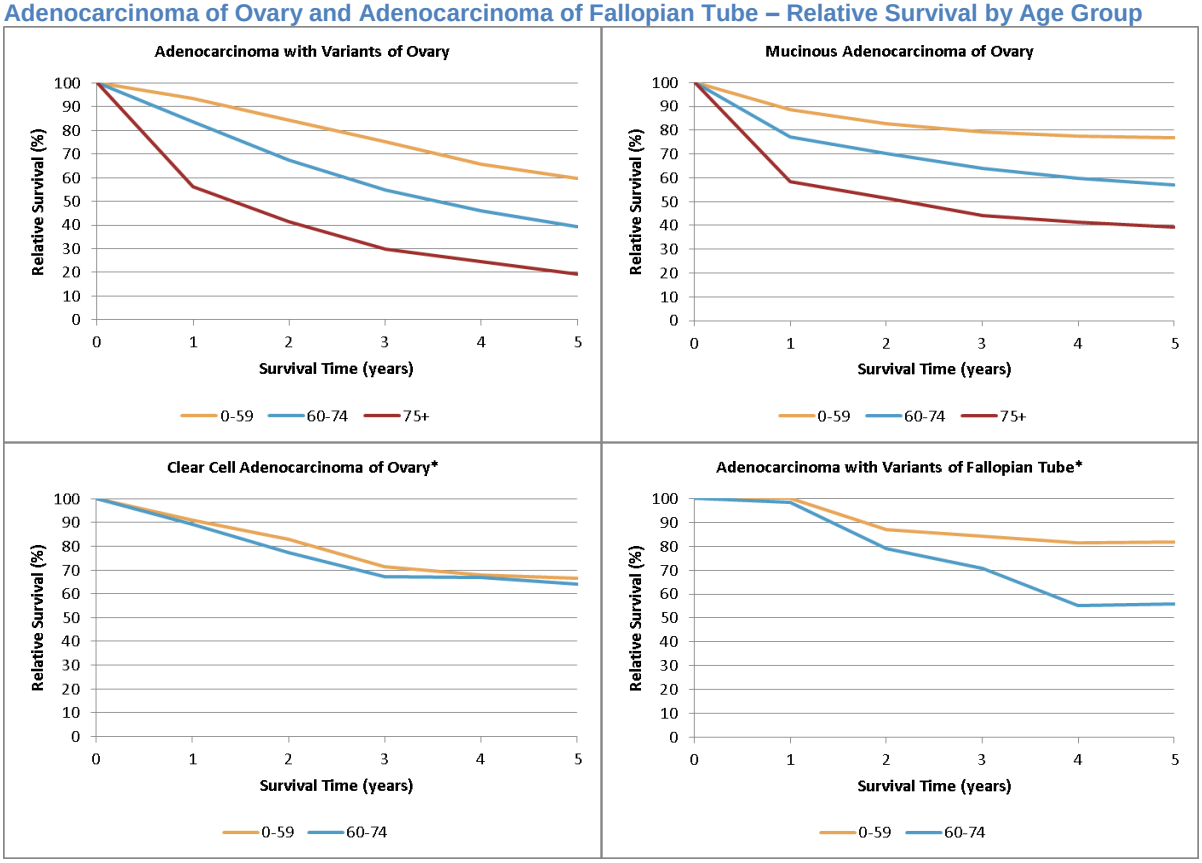
4.4.2 Survival by Age Group

Figure 16. Epithelial Tumours of Ovary and Fallopian Tube – Relative Survival by Age Group



- A large difference in prognosis can be observed between the different age groups. While patients in the youngest age group (0-59 years old) have a 5-year relative survival of 63.6%, this survival rate is more than 20% lower for the middle age group (60-74 years; 5-year relative survival: 41.0%), and only 21.2% for the oldest age group (75+ years).

Figure 17. Adenocarcinoma with Variants of Ovary, Mucinous Adenocarcinoma of Ovary, Clear Cell Adenocarcinoma of Ovary and Adenocarcinoma of Fallopian Tube – Relative Survival by Age Group

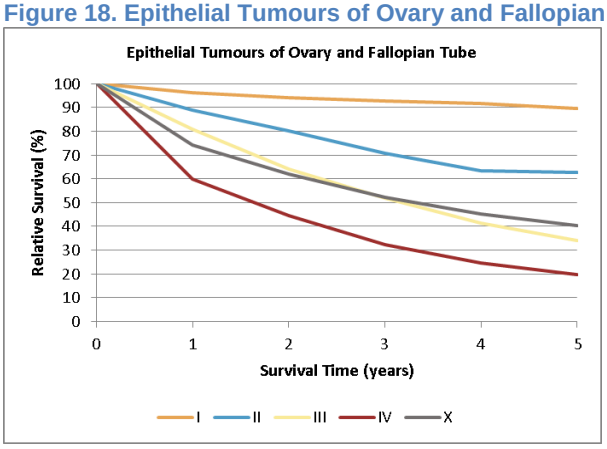


* Survival of patients aged 75 and above is not shown because the number at risk is lower than 35.

- For all age groups, survival is much worse for patients diagnosed with an adenocarcinoma of the ovary compared with the other subtypes.
- Survival of the clear cell adenocarcinoma of the ovary is similar for patients in the different age groups.
- A large difference in survival for longer follow-up periods (after three years) is noted for the adenocarcinoma of the fallopian tube, between the age groups 0-59 years and 60-74 year.

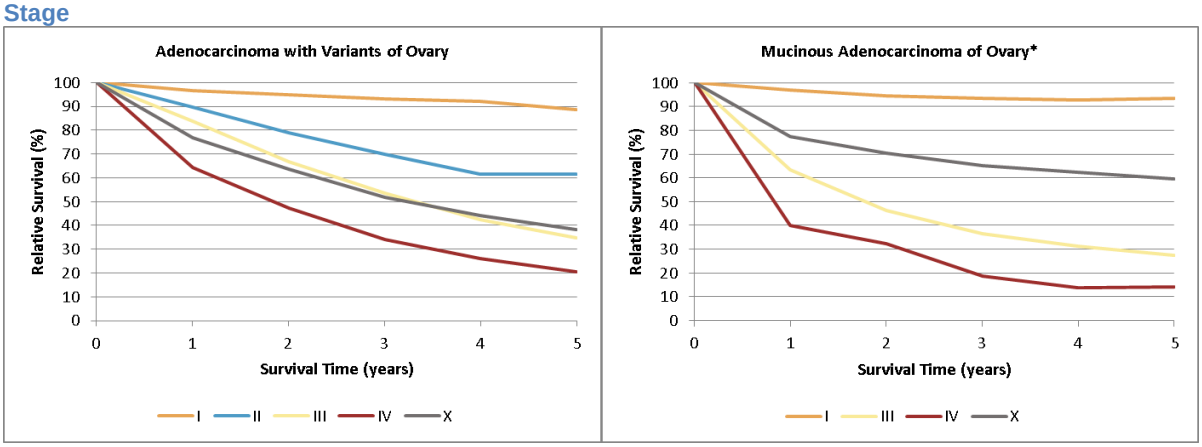
4.4.3 Survival by Stage[2]

Figure 18. Epithelial Tumours of Ovary and Fallopian Tube – Relative Survival by Stage



- The extent of the disease is a strong prognostic factor for survival of epithelial tumours of the ovary and fallopian tube. The 5-year relative survival ranges from 89.5% for stage I tumours to 19.6% for stage IV.

Figure 19. Adenocarcinoma of Ovary and Mucinous Adenocarcinoma of Ovary – Relative Survival by Stage



* Survival results are not displayed for stage II tumours because the number at risk is lower than 35.

- Survival of adenocarcinoma of the ovary is very similar to the survival of all epithelial tumours of the ovary and fallopian tube together.
- Survival of stage IV mucinous adenocarcinoma of the ovary (14.2%) is worse than survival of stage IV of all epithelial tumours of the ovary and fallopian tube together.

[2] Survival by stage is not displayed for clear cell adenocarcinoma of ovary because only stage I and X have a number at risk higher than 35 and for adenocarcinoma of fallopian tube because all stages have a number at risk lower than 35.

5. Non-Epithelial Tumours of Ovary[3]

5.1 General Results

Table 10. Non-Epithelial Tumours of Ovary: Incidence, Trends, Survival

Flemish Region 2001-2010		Incidence				Trend		Survival	
Females						EAPC		Relative survival	
	R/C	N	CR	WSR	Avg Age	%	p-value	N at risk	5yr (%)
NON EPITHELIAL TUMOURS OF OVARY	R	230	0.70	0.60	53	-0.8	0.767	183	66.8
Mixed epithelial/mesenchymal tumours of	R	104	0.30	0.20	68	2.7	0.611	96	19.2
Sex cord tumours of ovary	R	48	0.20	0.10	55	-14.4	0.024	45	86.3
Malignant/Immature teratomas of ovary	R	36	0.10	0.10	35	-4.1	0.462	35	89.4
Germ cell tumour of ovary	R	42	0.10	0.20	30	9.9	0.121	41	95.6

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

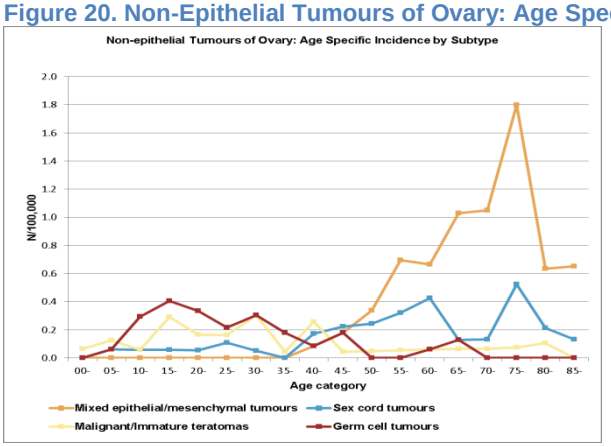
RS: relative survival

AvgAge: average age at diagnosis

5.2 Incidence

- 230 new non-epithelial tumours of ovary are diagnosed in the Flemish Region between 2001 and 2010.
- RARECARE defines four rare entities:
  - About half of all diagnoses are mixed epithelial/mesenchymal tumours of ovary
  - Sex cord ovarian tumours account for 48 new diagnoses.
  - With 36 new cases, malignant/immature teratomes are the least frequently diagnosed entity.
  - 42 new germ cell tumours are observed in the Flemish Region between 2001 and 2010.

Figure 20. Non-Epithelial Tumours of Ovary: Age Specific Incidence by Subtype



[3] Analyses by stage are not reported for the non-epithelial tumours of the ovary because only stage X had a number at risk higher than 35.

- Mixed epithelial/mesenchymal tumours do not occur in females younger than 40 years of age. From the age of 40 year, incidence rates increase rapidly with age until the age of 75 years.
- Sex cord ovarian tumours are occasionally diagnosed under the age of 40 years.
- Malignant/immature teratomas and germ cell tumours of ovary are predominantly diagnosed under the age of 40 years.

5.3 Survival

5.3.1 Overall Survival

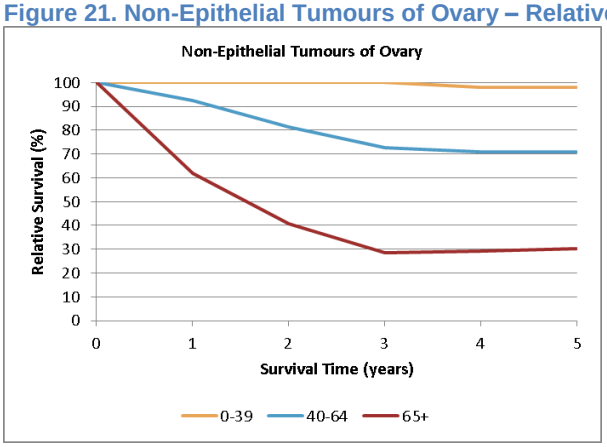
Table 11. Non-Epithelial Tumours of Ovary – Overall Survival

	N	Observed Survival					Relative Survival				
	at risk	1 year	3 year	5 year	10 year	5 year CI	1 year	3 year	5 year	10 year	5 year CI
NON-EPITHELIAL TUMOURS OF OVARY	183	84.2	65.9	64.5	58.9	[56.9 ; 71.1]	85.0	67.4	66.8	63.6	[58.9 ; 73.6]
Mixed epithelial/mesenchymal tumours	96	62.5	23.1	18.2	10.8	[10.5 ; 27.5]	63.7	24.0	19.2	12.0	[11.1 ; 29.1]
Sex cord tumours	45	95.6	81.7	81.7	75.1	[66.6 ; 90.4]	96.8	84.6	86.3	85.8	[70.4 ; 95.5]
Malignant/Immature teratomas	35	94.3	91.3	87.5	87.5	[69.8 ; 95.2]	94.6	92.3	89.4	90.7	[71.3 ; 97.2]
Germ cell tumour	41	95.1	95.1	95.1	95.1	[81.9 ; 98.8]	95.2	95.4	95.6	96.0	[82.3 ; 99.3]

- Survival of the non-epithelial tumours of the ovary decreases after diagnosis to a 5-year observed survival of 64.5% and a 5-year relative survival of 66.8%.
- Survival is very low for the mixed epithelial/mesenchymal tumours with a 5-year observed survival of only 18.2% and a 5-year relative survival of 19.2%.
- Survival of the other subtypes is better than the survival of all non-epithelial tumours of ovary together. Especially germ cell tumours have a high survival rate (5-year relative survival of 95.6%).

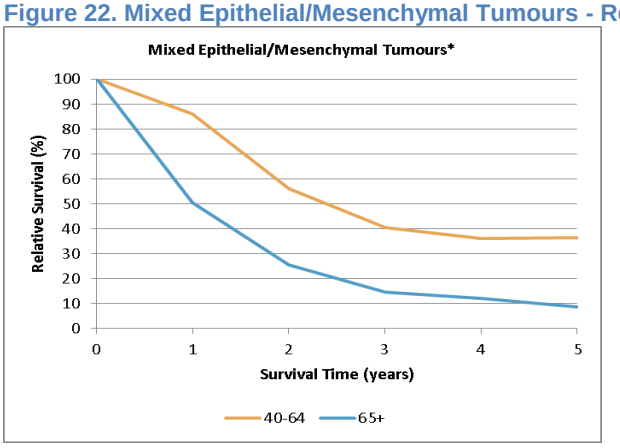
5.3.2 Survival by Age Group

Figure 21. Non-Epithelial Tumours of Ovary – Relative Survival by Age Group



- Survival of all non-epithelial tumours of the ovary together is almost 100% for the youngest age group (0-39 years), but is lower for the older age groups with a 5-year relative survival of 71.1% and 30.3% for the age groups 40-64 years and 65+ years, respectively.

Figure 22. Mixed Epithelial/Mesenchymal Tumours - Relative Survival by Age Group



* Survival results of patients aged between 0 and 39 are not shown because no patients of this age group are diagnosed with an mixed epithelial/mesenchymal tumour.

- Survival for mixed epithelial/mesenchymal tumours is much worse for patients in the age group 65+ (5-year relative survival: 8.8%) than for patients in the age group 40-64 years (5-year relative survival: 36.4%).

6. Epithelial Tumours of Vulva and Vagina

6.1 General Results

Table 12. Epithelial Tumours of Vulva and Vagina: Incidence, Trends, Survival

Flemish Region 2001-2010		Incidence				Trend		Survival	
Females						EAPC		Relative survival	
	R/C	N	CR	WSR	Avg Age	%	p-value	N at risk	5yr (%)
EPITHELIAL TUMOURS OF VULVA AND VAGINA	R	1,124	3.65	1.63	70	2.6	0.044	1,019	61.2
Squamous cell carcinoma with variants of vulva and vagina	R	1,008	3.27	1.46	70	4.1	0.005	921	62.4
Adenocarcinoma with variants of vulva and vagina	R	74	0.24	0.13	65	-5.3	0.147	63	54.8
Paget's disease of vulva and vagina	R	19	0.06	0.03	71	*	*	14	*
Undifferentiated carcinoma of vulva and vagina	R	3	0.01	0.00	78	*	*	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

6.2 Incidence

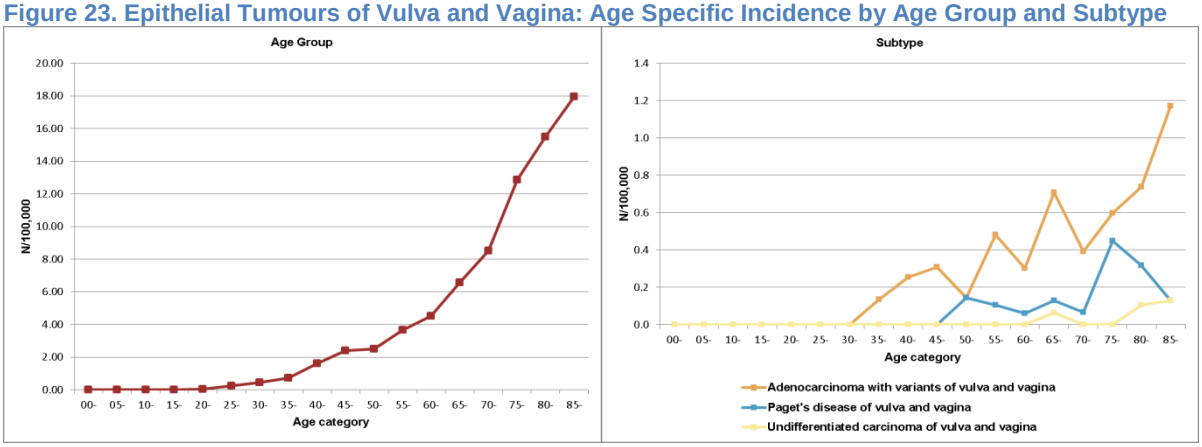
- 1.124 new epithelial tumours of vulva and vagina are diagnosed in the Flemish Region between 2001 and 2010.

Table 13. Morphological Distribution of Epithelial Tumours of Vulva and Vagina

Flemish Region 2001-2010	Total		Vulva		Vagina	
Squamous cell carcinoma with variants of vulva and vagina	1008	91%	837	95%	171	77%
Adenocarcinoma with variants of vulva and vagina	74	7%	24	3%	50	22%
Paget's disease of vulva and vagina	19	2%	19	2%	0	0%
Undifferentiated carcinoma of vulva and vagina	3	0%	1	0%	2	1%
Total	**1104**	**100%**	**881**	**100%**	**223**	**%**

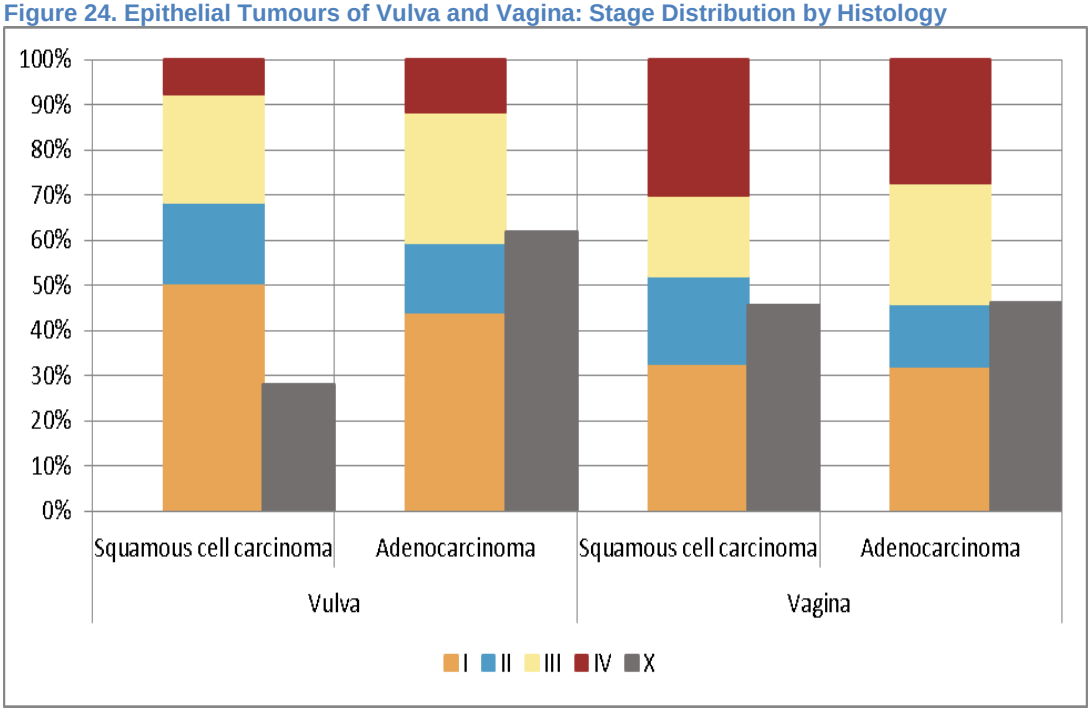
- RARECARE defines four rare entities:
  - Squamous cell carcinoma represents 95% of all tumours of vulva and 77% of the vaginal tumours.
  - 50 vaginal and 24 vulvar adenocarcinoma are registered.
  - 19 cases of vulva Paget's disease are observed.
  - Only 3 new diagnoses of undifferentiated carcinoma of vulva and vagina are observed in the Flemish Region between 2001 and 2010.

Figure 23. Epithelial Tumours of Vulva and Vagina: Age Specific Incidence by Age Group and Subtype



- Age specific incidence rates for squamous cell carcinoma and adenocarcinoma of vulva and vagina start to increase around the age of 35-40 years.
- Paget's disease is not observed in females younger than 50 years of age.

Figure 24. Epithelial Tumours of Vulva and Vagina: Stage Distribution by Histology

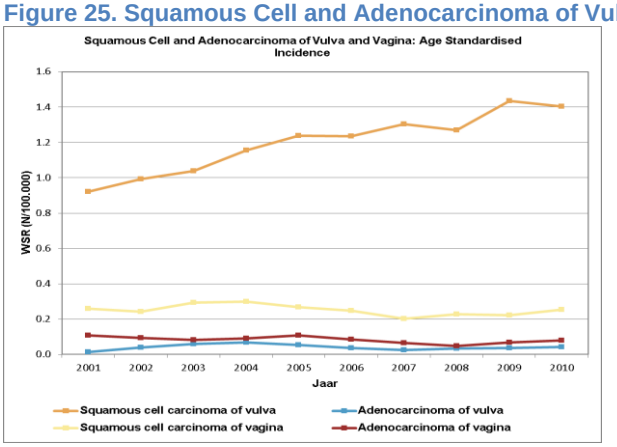


Belgian Cancer Registry

- Epithelial cancers of the vagina are more advanced at diagnosis than epithelial cancers of the vulva.
- For both vaginal and vulvar epithelial cancer, no clear difference in stage distribution between squamous cell carcinoma and adenocarcinoma subtypes is noted.

6.3 Trends

Figure 25. Squamous Cell and Adenocarcinoma of Vulva and Vagina: Age-Standardised Incidence



- There is an increase in the age-standardised incidence of squamous cell carcinoma of the vulva between 2001 and 2010.
- Incidences of squamous cell carcinoma of the vulva and adenocarcinoma of vulva and vagina remain stable in time.

6.4 Survival

6.4.1 Overall Survival

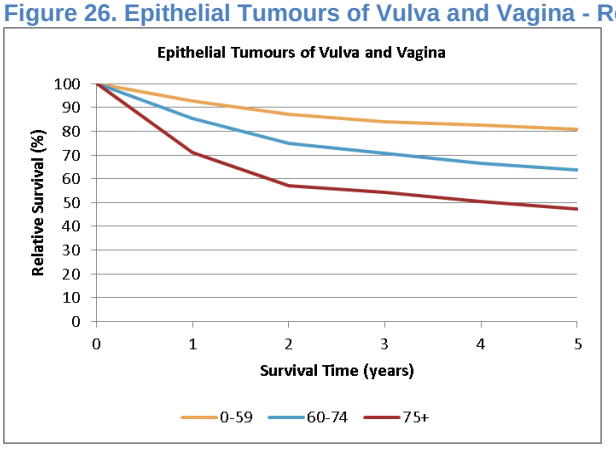
Table 14. Epithelial Tumours of Vulva and Vagina – Overall Survival

	N	Observed Survival					Relative Survival				
	at risk	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 VULVA AND VAGINA	1,019	77.9	60.3	51.8	38.1	[48.4 ; 55.0]	80.9	66.9	61.2	54.4	[57.2 ; 65.0]
Squamous cell carcinoma with variants	921	79.0	61.6	52.7	38.8	[49.1 ; 56.1]	82.0	68.3	62.4	56.3	[58.2 ; 66.5]
Adenocarcinoma with variants	63	69.8	53.3	51.1	37.3	[37.8 ; 62.9]	71.5	56.0	54.8	42.7	[40.6 ; 67.5]
Paget's disease	14	*	*	*	*	*	*	*	*	*	*
Undifferentiated carcinoma	1	*	*	*	*	*	*	*	*	*	*

- Almost half of the patients diagnosed with an epithelial tumour of the vulva or vagina dies within the first five years after diagnosis.
- Survival seems higher for squamous cell carcinoma as for adenocarcinoma although this difference is more pronounced for relative than for observed survival.

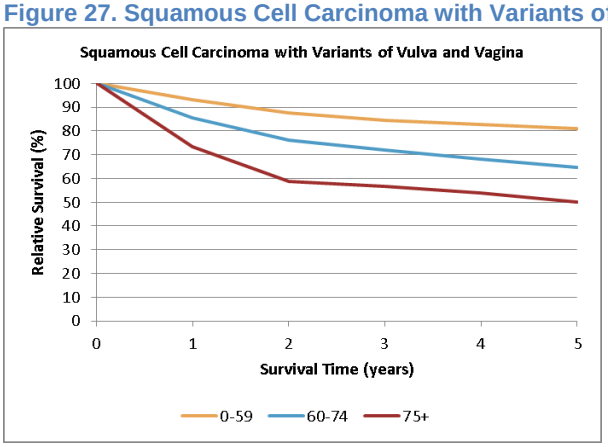
6.4.2 Survival by Age Group[4]

Figure 26. Epithelial Tumours of Vulva and Vagina - Relative Survival by Age Group



- Survival is dependent on age, with a 5-year relative survival of 81.0%, 63.8% and 47.2% for the age groups 0-59 years, 60-74 years and 75+ years respectively.

Figure 27. Squamous Cell Carcinoma with Variants of Vulva and Vagina: Relative Survival by Age Group

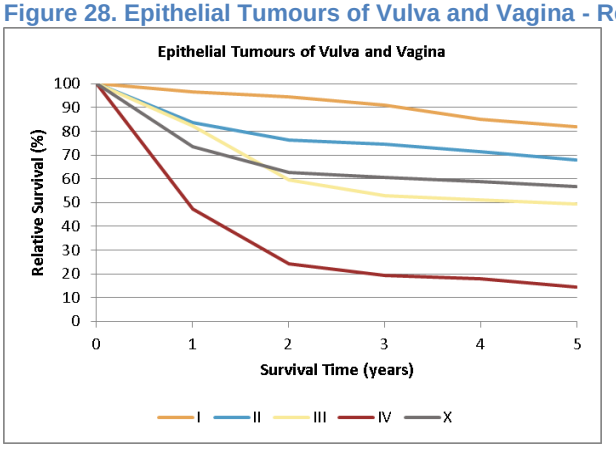


- Because most epithelial tumours of vulva and vagina are squamous cell carcinoma, survival by age group of these squamous cell carcinoma is highly similar to the earlier described survival by age group for all epithelial tumours of vulva and vagina together.

[4] Survival by age group is not displayed for the adenocarcinoma because all age groups have less than 35 patients at risk.

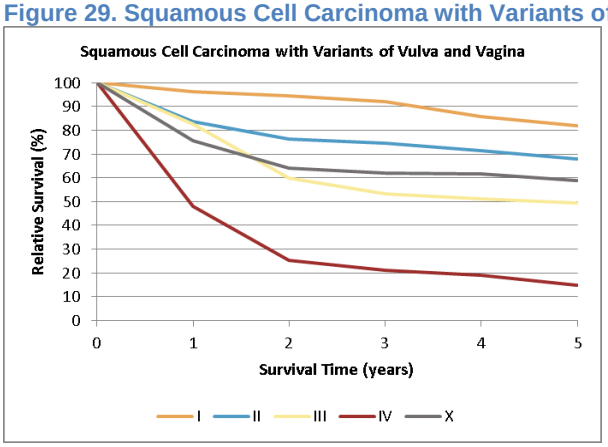
6.4.3 Survival by Stage[5]

Figure 28. Epithelial Tumours of Vulva and Vagina - Relative Survival by Stage



- Survival is dependent on stage, with a much worse survival for stage IV tumours than for the other stages.

Figure 29. Squamous Cell Carcinoma with Variants of Vulva and Vagina - Relative Survival by Stage



- Because most epithelial tumours of vulva and vagina are squamous cell carcinoma, survival by stage of these squamous cell carcinoma is highly similar to the earlier described survival by stage for all epithelial tumours of vulva and vagina together.

[5] Survival by stage is not displayed for the adenocarcinoma because all stages have less than 35 patients at risk.

7. Trophoblastic Tumour of Placenta[6]

7.1 General Results

Table 15. Trophoblastic Tumour of Placenta: Incidence, Trends, Survival

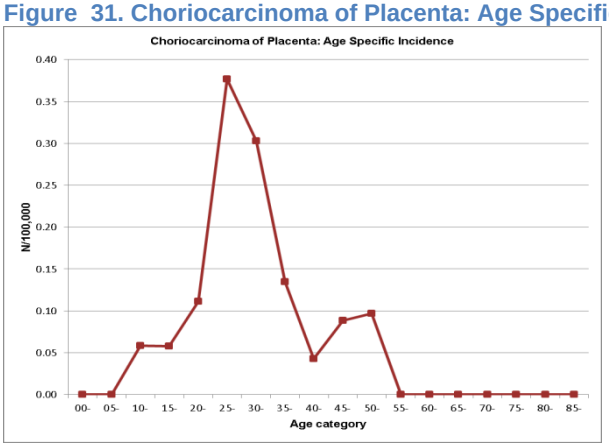
Flemish Region 2001-2010	Incidence					Trend		Survival	
Females						EAPC		Relative survival	
	R/C	N	CR	WSR	Avg Age	%	p-value	N at risk	5yr (%)
TROPHOBLASTIC TUMOUR OF PLACENTA	R	25	0.08	0.09	32	*	*	-	-
Choriocarcinoma of placenta	R	25	0.08	0.09	32	*	*	-	-

R/C: Rare or common
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

- 25 new diagnoses of trophoblastic placental tumours are registered for the Flemish Region between 2001 and 2010.
- All trophoblastic placental tumours are choriocarcinoma.

Figure 31. Choriocarcinoma of Placenta: Age Specific Incidence



- Choriocarcinoma of placeta occur in young women with a peak incidence at the age of 25-30 years.

[6] No survival results are shown for the trophoblastic tumours of the placenta because the number at risk is lower than 35.