

Rare Breast Tumours

1. Breast Tumours

1.1 General Results

Table 1. Epithelial Tumours of Breast: Incidence, Trends, Survival

Flemish Region 2001-2010		Incidence				Trend		Survival		
Females		R/C	N	CR	WSR	Avg Age	EAPC		Relative survival	
							%	p-value	N at risk	5yr (%)
EPITHELIAL TUMOURS OF BREAST (FEMALES)		C	54,477	176.81	105.36	61	-0.7	0.010	52,976	87.5
Invasive ductal carcinoma of breast		C	41,893	135.97	82.11	61	0.5	0.057	40,490	88.1
Invasive lobular carcinoma of breast		C	6,945	22.54	12.94	63	-1.3	0.076	6,672	88.7
Mammary Paget's disease of breast		R	209	0.68	0.38	63	-10.5	0.002	205	83.4
Special types of adenocarcinoma of breast		R	1,145	3.72	2.05	63	-6.0	0.007	1,112	96.8
Metaplastic carcinoma of breast		R	166	0.54	0.29	64	10.8	0.059	157	69.7
Salivary gland type tumours of breast		R	66	0.21	0.12	63	2.6	0.749	61	92.5
Epithelial tumour of male breast		R	503	1.68	0.91	67	-1.4	0.415	430	76.0

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

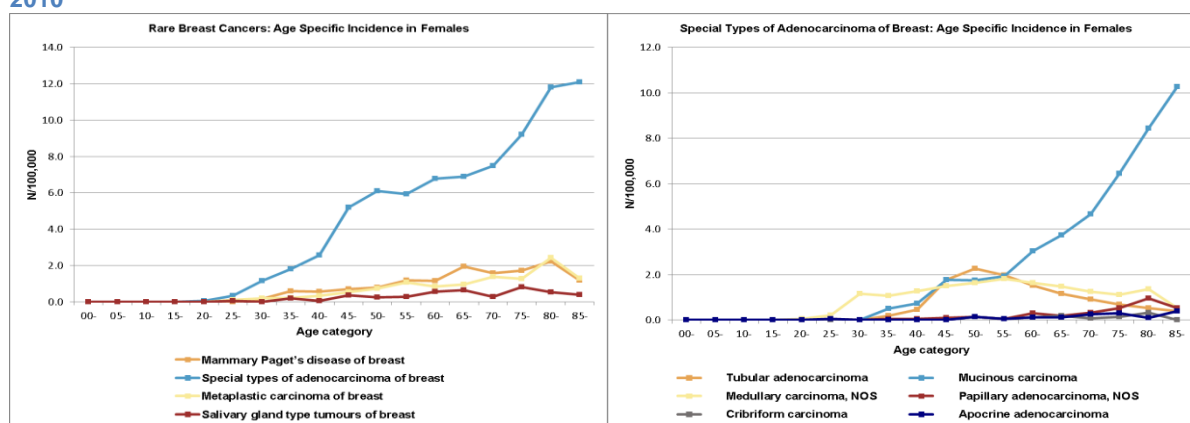
- 54,980 new epithelial tumours of the breast are diagnosed in the Flemish Region between 2001 and 2010.
- The male/female ratio is 0.01.
- Five rare histological entities are considered in the RARECARE list. With 2,089 new diagnoses between 2001 and 2010, they represent less than 4% off all breast cancer cases.
 - Male breast cancer is grouped as one rare entity, regardless of histology, thus including ductal carcinoma. Between 2001 and 2010 there are 503 Flemish males diagnosed with breast cancer.
 - Paget's disease of the breast accounts for 209 new diagnoses in females. Comparing the incidence for Paget's disease is difficult since registration practices for mammary Paget's disease may vary between registries. Some registries, such as the Belgian Cancer Registry, consider Paget's disease without additional information on the invasive behaviour as an in situ malignancy, while other registries may code by default this tumour as a malignant disease.
 - A wide variety of rare adenocarcinoma are grouped together as 'special types of adenocarcinoma'. The most common types are mucinous adenocarcinoma (49%), tubular adenocarcinoma (25%) and medullary carcinoma (19%).
 - Metaplastic carcinomas represent about 10% of all rare female breast cancers.
 - Salivary gland type tumours are the least common rare care entity with only 66 new diagnoses between 2001 and 2010.

Table 2. Rare breast cancers: Histological Subtypes by Age Groups

Label	Total	0-49	50-69	70+
Mammary Paget's disease of breast	209 13.2%	45 13.4%	87 13.7%	77 12.6%
Special types of adenocarcinoma of breast	1,145 72.2%	247 73.3%	456 71.7%	442 72.1%
Tubular adenocarcinoma	214 13.5%	56 16.6%	127 20.0%	31 5.1%
Mucinous carcinoma	564 35.6%	68 20.2%	180 28.3%	316 51.5%
Medullary carcinoma, NOS	285 18.0%	116 34.4%	118 18.6%	51 8.3%
Papillary adenocarcinoma, NOS	41 2.6%	4 1.2%	12 1.9%	25 4.1%
Cribriform carcinoma	15 0.9%	-	9 1.4%	6 1.0%
Apocrine adenocarcinoma	21 1.3%	1 0.3%	8 1.3%	12 2.0%
Secretory carcinoma	4 0.3%	2 0.6%	1 0.2%	1 0.2%
Glycogen-rich clear cell carcinoma	-	-	-	-
Lipid-rich carcinoma	1 0.1%	-	1 0.2%	-
Oncocytic carcinoma	-	-	-	-
Metaplastic carcinoma of breast	166 10.5%	31 9.2%	64 10.1%	71 11.6%
Squamous carcinoma	14 0.9%	2 0.6%	5 0.8%	7 1.1%
Adenosquamous carcinoma	25 1.6%	4 1.2%	16 2.5%	5 0.8%
Adenocarcinoma with cartilaginous and osseous metaplasia	-	-	-	-
Adenocarcinoma with spindle cell metaplasia	3 0.2%	-	1 0.2%	2 0.3%
Metaplastic carcinoma of breast, NOS	124 7.8%	25 7.4%	42 6.6%	57 9.3%
Salivary gland type tumours of breast	66 4.2%	14 4.2%	29 4.6%	23 3.8%
Mucoepidermoid carcinoma	3 0.2%	-	-	3 0.5%
Adenoid cystic carcinoma	34 2.1%	7 2.1%	17 2.7%	10 1.6%
Myoepithelial carcinoma	4 0.3%	1 0.3%	1 0.2%	2 0.3%
Acinic cell adenocarcinoma	25 1.6%	6 1.8%	11 1.7%	8 1.3%
Rare epithelial tumours of female breast	1,586	337	636	613

- The distribution of rare breast cancer entities varies only slightly with age. The greatest change is observed for the main special types of adenocarcinoma.
 - Medullary carcinomas represent 1 out of 3 rare cancer entities in patients younger than 50 years of age. Their contribution decreases to 19% in patients between the age of 50 and 69 years. In the oldest age group, less than 1 out of every 10 new diagnoses are medullary carcinomas.
 - More than half of all rare breast cancers diagnosed in patients of 70 years and older are mucinous carcinoma. Under the age of 50 years, they represent only 1 out of 5 rare cancers.
 - Tubular carcinomas are more often diagnosed in patients under the age of 70 years, thereafter, only 5% of rare cancers are tubular carcinoma.

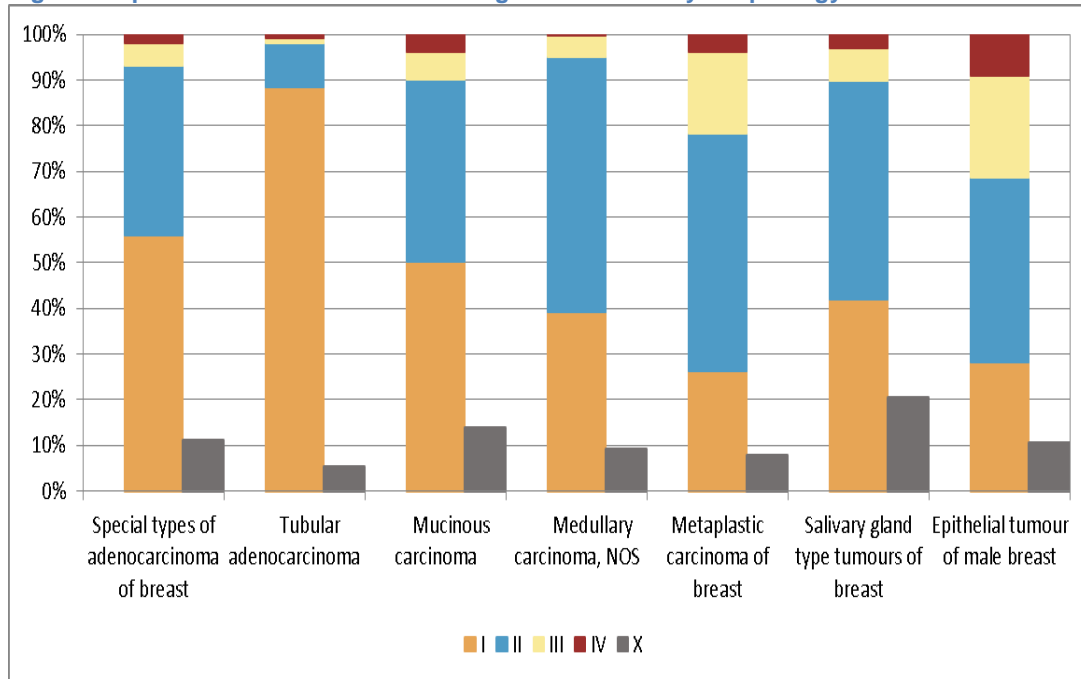
Figure 1. (left) Rare Breast Cancers: Age Specific Incidence in Females, Flemish Region 2001-2010; (right) Special Types of Adenocarcinoma of Breast: Age Specific Incidence in Females, Flemish Region 2001-2010



- Incidence rates for 'special types' adenocarcinoma increase from the age of 30 years.

- The increase in very young patients is mainly driven by an increase in medullar carcinoma. The rates for medullar carcinoma increase only slightly until the age of 50 years. After the age of 55, the rates decrease slightly.
- From the age of 40 years, tubular carcinoma incidence rates start to increase to reach a peak at 50 years. Then the incidence rates decrease.
- Mucinous carcinoma incidence rates increase from the age of 40 years and they keep increasing rapidly. With increasing age, mucinous carcinoma becomes more and more the predominant rare breast cancer type.
- The incidence rates for Paget's disease, metaplastic carcinoma and salivary gland type tumours increase slowly with age.

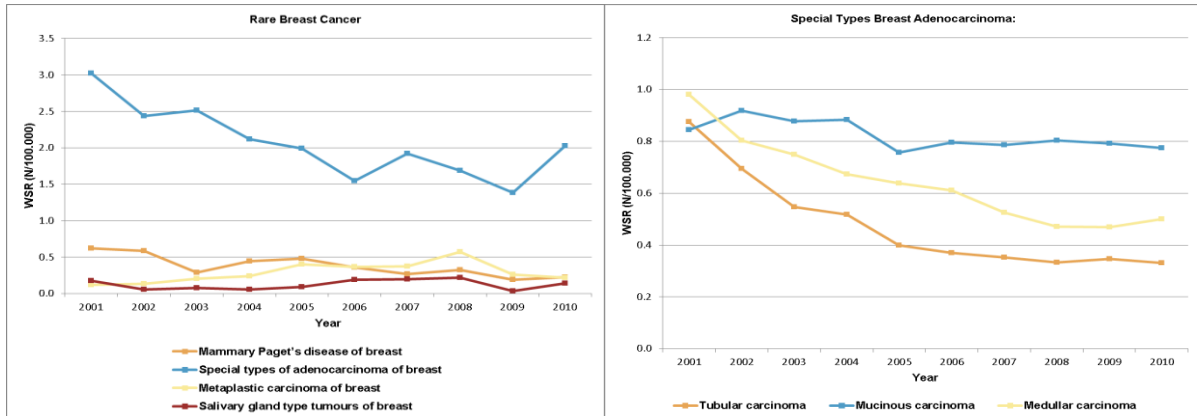
Figure 2. Epithelial Tumours of Breast: Stage Distribution by Morphology



- 'Special types' adenocarcinomas of breast have a good prognostic stage distribution.
 - About 90% of tubular carcinomas are diagnosed in stage I.
 - Half of the mucinous carcinomas are diagnosed in stage I and an additional 40% in stage II.
 - Medullary carcinoma is diagnosed for 40% in stage I and 50% in stage II.
- Metaplastic carcinoma has the most unfavorable stage distribution of the rare female breast cancer entities with more than 20% of cases diagnosed in stage III or IV.
- Salivary gland breast cancer present for 40% in stage I and 50% in stage II.
- Male breast cancer has a fairly poor prognostic stage distribution with 10% stage IV and a little more than 20% stage III.

1.3 Trends

Figure 3. Rare Breast Cancer and Special Types Breast Adenocarcinoma: Age-Standardised Incidence in Females (three year moving average)



- A significant decrease is observed in Paget's disease. An influence of improving and more comparable registration practices (no invasive Paget by default) in the hospitals cannot be excluded.
- Special types adenocarcinomas decrease significantly.
 - Tubular carcinomas decrease annually with 9.3% ($p = 0.010$).
 - Medullar carcinomas decrease significantly with 7.1% each year ($p = 0.009$).
 - Mucinous carcinomas show a slight decrease but the trend is not significant (EAPC = -1.6% [$p = 0.258$]).
- Metaplastic and salivary gland type carcinoma incidence increase, but the trends are not yet significant.

1.4 Survival

1.4.1 Overall Survival

Table 3. Epithelial Tumours of Breast - 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 BREAST	52,976	95.5	87.4	80.2	66.8	[79.8 ; 80.6]	97.1	92.0	87.5	80.1	[87.1 ; 87.9]
Invasive ductal carcinoma	40,490	96.1	88.2	81.2	68.2	[80.8 ; 81.6]	97.7	92.6	88.1	81.1	[87.7 ; 88.6]
Invasive lobular carcinoma	6,672	95.7	88.2	80.6	64.6	[79.6 ; 81.6]	97.5	93.3	88.7	78.6	[87.5 ; 89.8]
Mammary Paget's disease	205	94.2	81.9	76.2	60.8	[69.2 ; 81.4]	95.9	86.3	83.4	74.9	[75.7 ; 88.9]
Special types of adenocarcinoma	1,112	96.0	89.4	84.9	73.2	[82.5 ; 87.0]	98.5	96.8	96.8	95.1	[94.1 ; 99.2]
Metaplastic carcinoma	157	90.5	72.1	62.6	42.8	[54.3 ; 70.5]	92.5	76.8	69.7	54.8	[60.4 ; 78.5]
Salivary gland type tumours	61	91.8	86.4	84.5	77.9	[72.3 ; 91.7]	93.4	91.0	92.5	98.1	[79.2 ; 100.3]
Epithelial tumour of male breast	430	90.2	76.8	63.0	44.8	[57.7 ; 67.8]	93.7	86.0	76.0	65.8	[69.7 ; 81.8]

- Epithelial tumors of the male and metaplastic carcinoma in females have the worst prognosis, with a 5-year relative survival of 76.0% and 69.7% respectively.
- The best prognosis is seen in the salivary gland type tumours and the special types of adenocarcinoma with a 5-year relative survival of more than 90%.

1.4.2 Survival by Sex

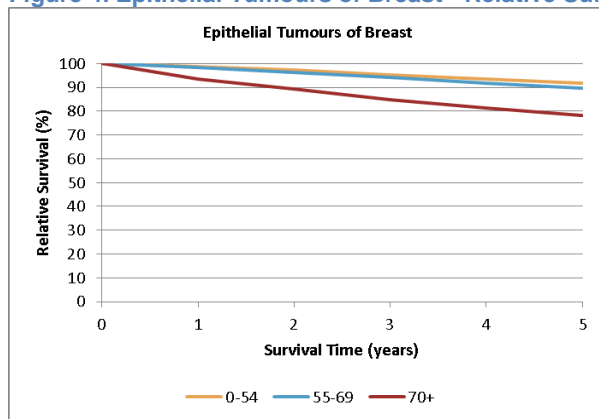
Table 4. Epithelial Tumours of Breast - Survival by Sex

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 BREAST	430	90.2	76.8	63.0	[57.7 ; 67.8]	93.7	86.0	76.0	[69.7 ; 81.8]
Invasive ductal carcinoma	-	-	-	-	-	-	-	-	-
Invasive lobular carcinoma	-	-	-	-	-	-	-	-	-
Mammary Paget's disease	-	-	-	-	-	-	-	-	-
Special types of adenocarcinoma	-	-	-	-	-	-	-	-	-
Metaplastic carcinoma	-	-	-	-	-	-	-	-	-
Salivary gland type tumours	-	-	-	-	-	-	-	-	-
Epithelial tumour of male breast	430	90.2	76.8	63.0	[57.7 ; 67.8]	93.7	86.0	76.0	[69.7 ; 81.8]
Females	N at risk	Observed Survival				Relative Survival			
EPITHELIAL TUMOURS OF BREAST	52,546	95.5	87.5	80.4	[80.0 ; 80.7]	97.2	92.0	87.6	[87.2 ; 88.0]
Invasive ductal carcinoma	40,490	96.1	88.2	81.2	[80.8 ; 81.6]	97.7	92.6	88.1	[87.7 ; 88.6]
Invasive lobular carcinoma	6,672	95.7	88.2	80.6	[79.6 ; 81.6]	97.5	93.3	88.7	[87.5 ; 89.8]
Mammary Paget's disease	205	94.2	81.7	75.9	[69.2 ; 81.4]	95.8	86.1	83.0	[75.7 ; 88.9]
Special types of adenocarcinoma	1,112	96.0	89.4	84.9	[82.5 ; 87.0]	98.5	96.8	96.8	[94.1 ; 99.2]
Metaplastic carcinoma	157	90.5	72.6	63.0	[54.3 ; 70.5]	92.4	77.3	70.1	[60.4 ; 78.5]
Salivary gland type tumours	61	91.8	86.4	84.5	[72.3 ; 91.7]	93.4	91.0	92.5	[79.2 ; 100.3]
Epithelial tumour of male breast	-	-	-	-	-	-	-	-	-

- Epithelial tumours of the breast have a much worse prognosis in males than in females.

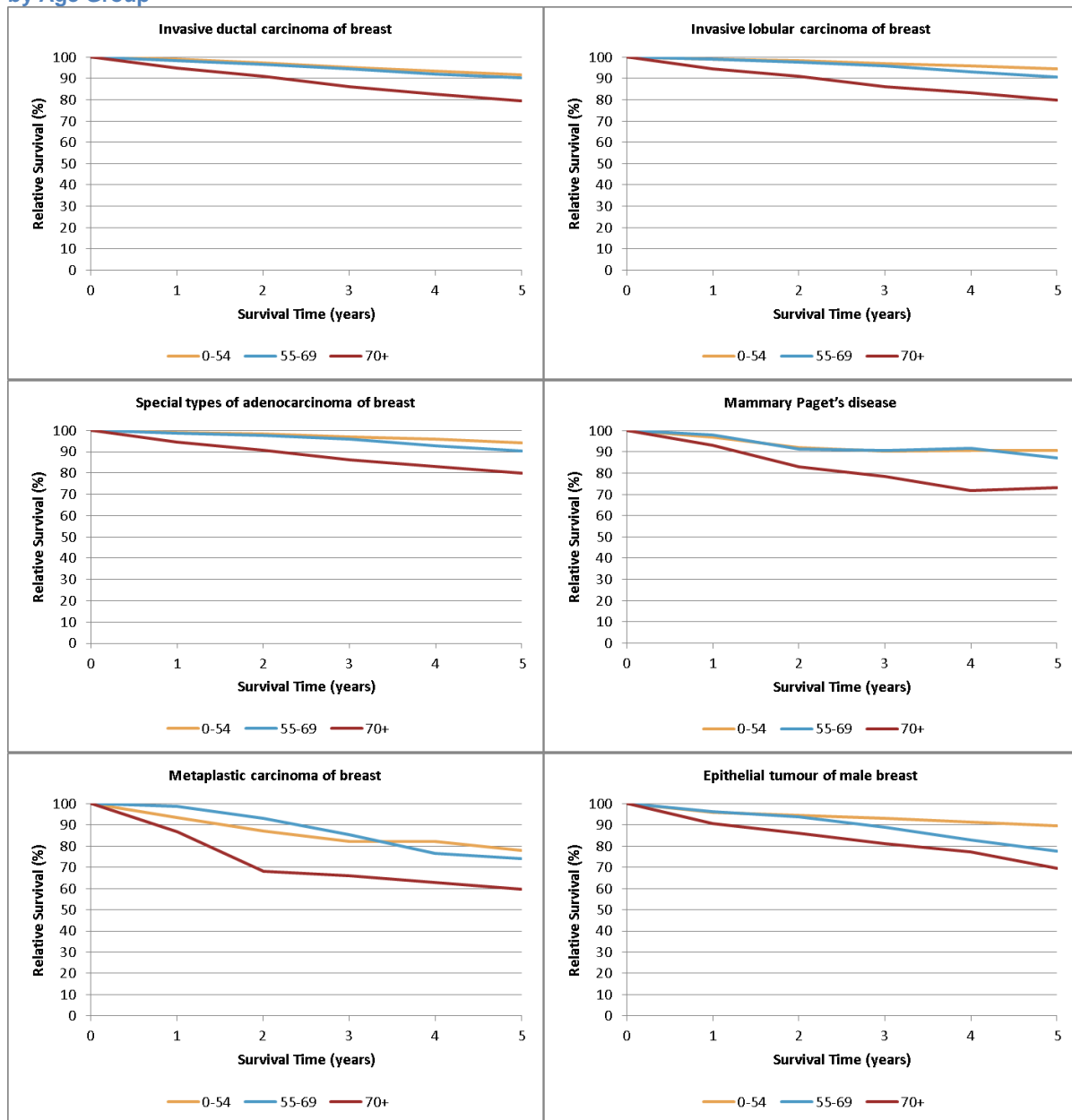
1.4.3 Survival by Age Group

Figure 4. Epithelial Tumours of Breast - Relative Survival by Age Group



- Prognoses of the 0-54 years and 55-69 years age groups are comparable.
- The oldest patients (70 years and older) have a worse prognosis, with a difference in relative survival at 5 years of about 10% compared with younger patients.

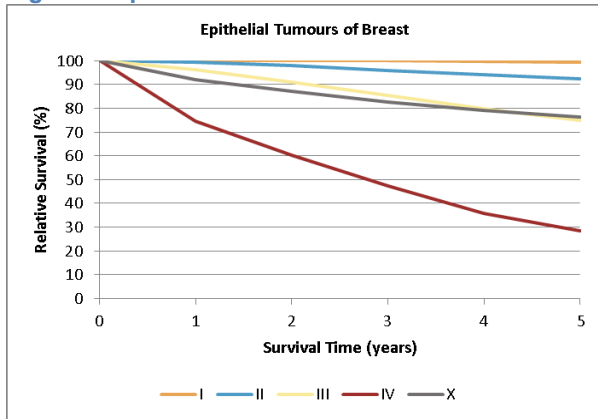
Figure 5. Invasive Ductal Carcinoma, Invasive Lobular Carcinoma, Special Types of Adenocarcinoma, Mammary Paget's Disease, Metaplastic Carcinoma, Epithelial Tumours of Male Breast - Relative Survival by Age Group



- Except for the epithelial tumors of the male breast, the 5-year relative survival for the 0-54 years age group and 55-69 years age group is comparable in the different histological groups.
- The oldest patients (70 years and older) have the worst prognosis in the different histological groups

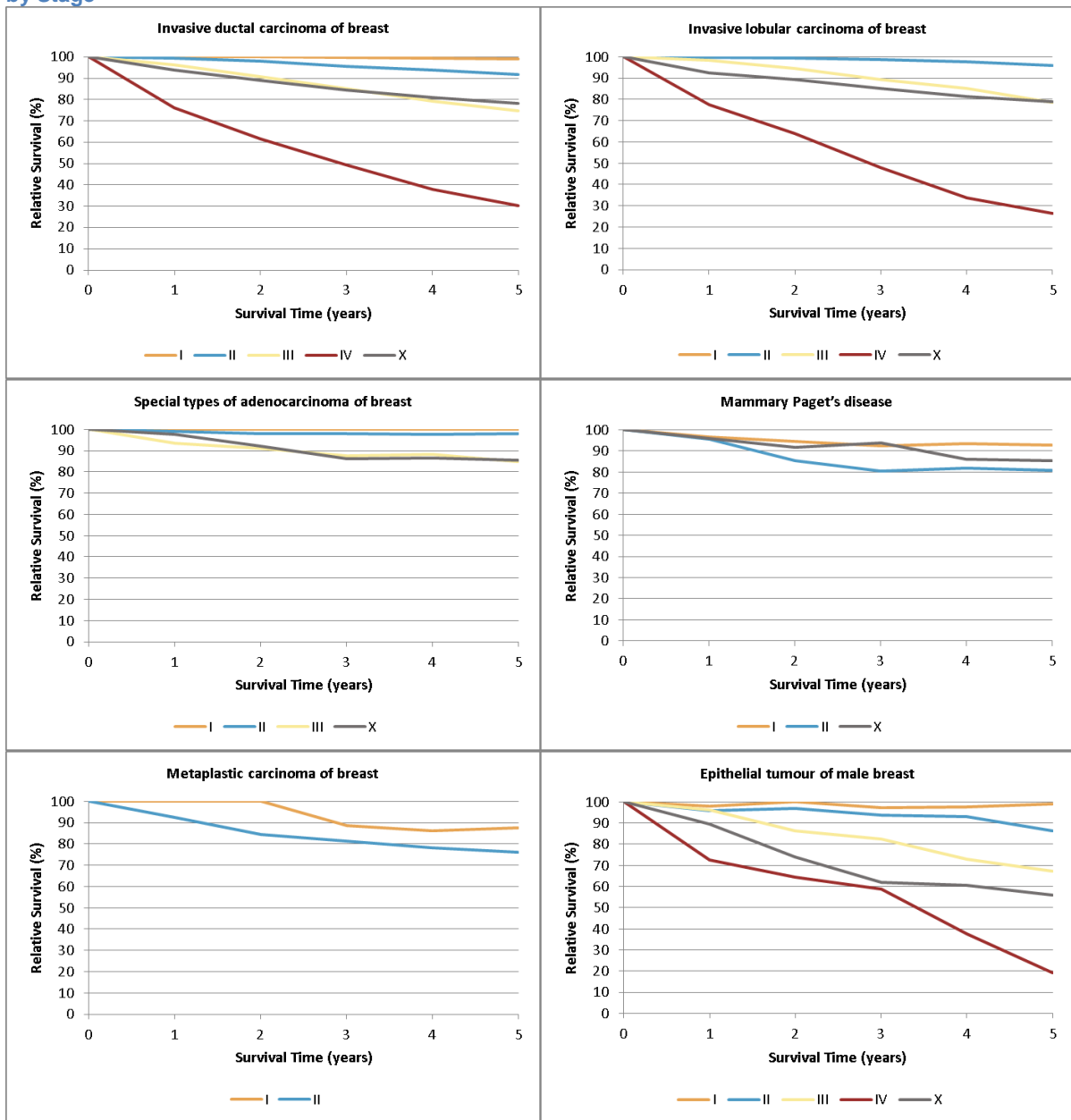
1.4.4 Survival by Stage

Figure 6. Epithelial Tumours of Breast - Relative Survival by Stage



- Survival depends on stage, with the best prognosis in stage I and the worst in stage IV.

Figure 7. Invasive Ductal Carcinoma, Invasive Lobular Carcinoma, Special Types of Adenocarcinoma, Mammary Paget's Disease, Metaplastic Carcinoma, Epithelial Tumours of Male Breast - Relative Survival by Stage



- Survival depends on stage, with the best prognosis in stage I and the worst in stage IV.
- The relative 5 year survival of stage IV tumor varies between 20 and 30%, with the worst prognosis seen in epithelial tumors of the male breast.