

Chapter 18

Cancer of the Vulva

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INTRODUCTION

Cancers of the vulva are relatively rare; they account for slightly less than 5% of all cancers of the female genital organs. Approximately 3,740 women are diagnosed with cancer of the vulva in the United States each year (1).

MATERIALS AND METHODS

Between 1988 and 2001, there were 13,949 cases of cancer of the vulva diagnosed in SEER. The following were excluded from the analysis: patients for whom vulvar cancer was not the first primary, cases identified through autopsy or death certificate only, persons of unknown race, cases without active follow-up or alive with no survival time, patients less than 20 years old, cases without microscopic confirmation, sarcomas, and carcinoids. There were 6,280 in situ cancers of the vulva excluded from analyses. After these exclusions, 4,098 cases remained for analysis (Table 18.1).

RESULTS

Age and Race

Of the 4,098 adult patients, 18.3% were diagnosed in adults under age 50 (Table 18.2). Over 52% of all cases

were diagnosed in women age 70 and over. Almost 89% of the patients were white. The percentage of adult black women diagnosed before the age of 50 is twice that of white women (35% vs. 17%). A slight difference is observed in the percent of black women diagnosed between the ages of 50 and 59 compared to white women (16% vs. 12%). Thirty-two percent of the black women were diagnosed at age 70 or older, compared to 55% of white women.

For all women, survival declined with age (Table 18.3). In 5-year relative survival rates, only minor differences between white and black women (Table 18.3) were observed. The largest survival differential was seen for ages 70+, where white women had a 5-year relative survival rate of 67%, compared to 60% for black females.

Geographic Location

Five-year relative survival rates in the 12 SEER areas included in this chapter ranged from 84.7% in Hawaii to 71.5% in Los Angeles (Table 18.4).

Histology

Distribution by histology is presented in Table 18.5. Tumors classified as squamous comprise 82.7% of all invasive cancers of the vulva with 50.6% Not Otherwise Specified (NOS), 14.2% keratinizing, 9.8% basal cell, and

Table 18.1: Cancer of the Vulva: Number of Cases and Exclusions by Reason, 12 SEER Areas, 1988-2001

Number Selected/Remaining	Number Excluded	Reason for Exclusion/selection
13,949	0	Select 1988-2001 diagnosis (Los Angeles for 1992-2001 only)
11,226	2,723	Select first primary only
11,199	27	Exclude death certificate only or at autopsy
10,671	528	Exclude unknown race
10,619	52	Exclude alive with no survival time
10,486	133	Exclude children (Ages 0-19)
4,206	6,280	Exclude in situ cancers
4,169	37	Exclude no or unknown microscopic confirmation
4,098	71	Exclude sarcomas and carcinoids

Table 18.2: Cancer of the Vulva: Age (20+) and Race Distributions, 12 SEER Areas, 1988-2001

Age Group (Years)	Total		White		Black		Other	
	Cases	Percent	Cases	Percent	Cases	Percent	Cases	Percent
Total (20+)	4,098	100.0	3,637	100.0	296	100.0	165	100.0
20-29	51	1.2	40	1.1	7	2.4	4	2.4
30-39	215	5.2	177	4.9	29	9.8	9	5.5
40-49	486	11.9	400	11.0	69	23.3	17	10.3
50-59	508	12.4	443	12.2	46	15.5	19	11.5
60-69	675	16.5	590	16.2	49	16.6	36	21.8
70-79	1,104	26.9	1,002	27.6	53	17.9	49	29.7
80+	1,059	25.8	985	27.1	43	14.5	31	18.8

8.1% other specified epidermoid types. Tumors classified as adenocarcinoma comprised 8.5% of the total, while melanoma comprised 5.9%. These three histology groups are the ones analyzed here.

Staging

The Federation Internationale de Gynecologie et d'Obstetrique (FIGO) and the American Joint Committee on Cancer (AJCC) have designated staging for cancers of the vulva excluding melanoma. Lack of specificity in the SEER data prevents a detailed breakdown of Stages IA-B and IVA-B for all years. SEER modified AJCC staging, 5th edition, was used for analyses in this chapter. The 5th edition AJCC staging (2) states:

Stage I vulvar cancer is defined as lesions 2 cm or less confined to the vulva or perineum. There are no lymph node metastases

Stage II vulvar cancer is defined as tumor either confined to the vulva and/or perineum or more than 2 cm in the greatest dimension with no nodal metastases.

Stage III vulvar cancer is defined as tumor of any size arising on the vulva and/or perineum with either adjacent spread to the lower urethra, the vagina, or the anus, or unilateral regional lymph node metastases.

Stage IV vulvar cancer is defined as tumor invading any of the following: upper urethra, bladder mucosa, rectal mucosa, pelvic bone and/or bilateral regional nodal metastases or any distant metastasis including pelvic lymph nodes.

Vulvar melanoma used the SEER modified AJCC 5th edition staging for melanoma.

Squamous

Survival by Age and Stage

Of the 3,390 patients with squamous cell carcinoma, enough information to establish stage at diagnosis was available for 3,171 (94%). The percent diagnosed in stage I declines with age, from 56% in ages 20-69 to 38% in ages 70+. Stages II and III show increases with age. Percent of stage IV cases is almost equal across age groups (Table 18.6).

Table 18.3: Cancer of the Vulva: Number and Distribution of Cases, Median Survival Time (Months) and 5-year Survival Rates (%) by Race and Age (20+), 12 SEER Areas, 1988-2001

Race/Age (Years)	Cases	Median Survival Time (Months)	5-Year Survival Rate (%)		
			Observed	Expected	Relative
All Races, 20+	4,098	104.2	62.3	81.6	76.4
White, 20+	3,637	99.7	61.5	81.0	76.0
Black, 20+	296	107.7	65.0	86.0	75.3
All Races, 20-69	1,935	> 120	81.6	96.0	85.0
White, 20-69	1,650	> 120	81.8	96.1	85.1
Black, 20-69	200	> 120	77.5	94.6	81.6
All Races, 70+	2,163	51.6	46.0	68.8	66.8
White, 70+	1,987	50.7	45.5	68.4	66.5
Black, 70+	96	42.8	41.1	68.0	59.9

Table 18.4: Cancer of the Vulva: Number and Distribution of Cases and 1-, 2-, 3-, 5-, 8-, & 10-Year Relative Survival Rates (%) by SEER Geographic Area, Ages 20+, 12 SEER Areas, 1988-2001

SEER Geographic Area	Cases	Percent	Relative Survival Rate (%)					
			1-Year	2-Year	3-Year	5-Year	8-Year	10-Year
Total	4,098	100.0	89.5	82.9	79.8	76.4	73.0	69.7
Atlanta and Rural Georgia	286	7.0	91.6	84.2	82.5	78.3	72.1	68.1
Atlanta (Metropolitan) - 1988+	265	6.5	91.1	83.7	81.8	77.9	70.6	66.0
Rural Georgia - 1988+	21	0.5	~	~	~	~	~	~
California								
Los Angeles - 1992+	596	14.5	84.9	78.6	75.5	71.5	65.4	61.5
Greater Bay Area	624	15.2	91.7	85.9	80.8	78.4	73.7	69.9
San Francisco-Oakland SMSA - 1988+	408	10.0	90.9	85.0	80.2	76.7	72.2	66.0
San Jose-Monterey - 1988+	216	5.3	93.1	87.6	82.1	80.6	76.2	76.2
Connecticut - 1988+	616	15.0	87.3	78.5	74.8	73.3	70.3	70.3
Detroit (Metropolitan) - 1988+	592	14.4	88.5	80.3	76.1	72.3	69.2	63.6
Hawaii - 1988+	95	2.3	93.6	93.0	88.0	84.7	80.9	65.9
Iowa - 1988+	519	12.7	92.3	85.3	84.3	80.5	77.2	72.2
New Mexico - 1988+	162	4.0	91.5	86.3	84.4	74.1	69.6	69.6
Seattle (Puget Sound) - 1988+	461	11.2	90.2	85.9	83.6	78.6	77.8	75.1
Utah - 1988+	147	3.6	91.3	84.1	81.3	81.3	80.5	72.1

~ Statistic not displayed due to less than 25 cases.

Table 18.5: Cancer of the Vulva: Number and Distribution of Cases and 5-Year Relative Survival Rates (%) by Histology, Ages 20+, 12 SEER Areas, 1988-2001

Histology	ICD-O Code	Cases	Percent	5-Year Relative Survival Rate (%)
Total	8000-9970	4,098	100.0	76.4
Squamous	8050-8130	3,390	82.7	75.9
Squamous, NOS	8070	2,073	50.6	73.7
Squamous, Keratinizing	8071	582	14.2	60.2
Basal Cell	8090-8110	403	9.8	99.4
Other Epidermoid	8050-8069,8072-8089,8111-8130	332	8.1	84.8
Adenocarcinoma	8140-8147,8160-8162,8180-8221,8250-8506,8520-8550,8560,8570-8573,8940-8941	347	8.5	91.9
Paget, Extramammary	8542	238	5.8	99.0
All Other Adenocarcinoma	8140-8147,8160-8162,8180-8221,8250-8506,8520-8541,8543-8550,8560,8570-8573,8940-8941	109	2.7	68.2
Other Specified Carcinomas	8030-8045,8150-8155,8170-8171,8230-8248,8510-8512,8561-8562,8580-8671	<5	~	~
Carcinoma, NOS	8010-8022	105	2.6	77.1
Other Specified Types	8720-8790,8931-8932,8950-8979,8982,9000-9030,9060-9110,9350-9364,9380-9512,9530-9539	244	6.0	58.9
Melanoma	8720-8790	240	5.9	58.9
All Other Specified Types	8931-8932,8950-8979,8982,9000-9030,9060-9110,9350-9364,9380-9512,9530-9539	<5	~	~
Unspecified	8000-8004	11	0.3	~

~ Statistic not displayed due to less than 25 cases.

A survival differential across age exists for Stages II-IV and unknown stage with women aged 70 and older exhibiting lower survival compared to women aged 20-69. No difference by age is observed in stage I (Table 18.7, Figure 18.1).

Survival by Stage

Table 18.8 and Figure 18.2 show the survival by stage and time since diagnosis for squamous cell carcinoma. In stages III-IV, the steepest declines in survival are observed within the first and second years since diagnosis. Survival continues to decline throughout the 10 years observed in these stages.

Survival by Histology and Stage

Basal cell carcinoma shows a positive survival advantage compared to the other squamous histologic subtypes in stage I and II. Keratinizing has the least favorable survival for these stages. Little difference exists between the other two squamous subtypes in stages I-IV (Table 18.9).

Figure 18.1: Squamous Cell Carcinoma of the Vulva: 5-Year Relative Survival Rate (%) by Age (20+) and AJCC Stage (SEER modified, 5th Edition), 12 SEER Areas, 1988-2001

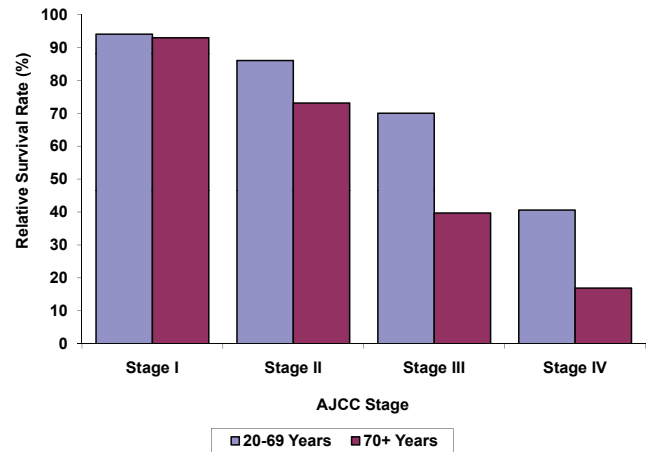


Table 18.6: Squamous Cell Carcinoma of the Vulva: Number and Distribution of Cases by AJCC Stage (SEER modified, 5th Edition) and Age (20+), 12 SEER Areas, 1988-2001

AJCC Stage	Age (Years)					
	Total 20+		20-69		70+	
	Cases	Percent	Cases	Percent	Cases	Percent
Total	3,390	100.0	1,566	100.0	1,824	100.0
Stage I	1,567	46.2	882	56.3	685	37.6
Stage II	650	19.2	253	16.2	397	21.8
Stage III	744	21.9	275	17.6	469	25.7
Stage IV	210	6.2	90	5.7	120	6.6
Unknown/Unstaged	219	6.5	66	4.2	153	8.4

Table 18.7: Squamous Cell Carcinoma of the Vulva: Number of Cases and 5-Year Relative Survival Rates (%) by AJCC Stage (SEER modified, 5th Edition) and Age (20+), 12 SEER Areas, 1988-2001

AJCC Stage	Age (Years)					
	Total 20+		20-69		70+	
	Cases	5-Year Relative Survival Rate (%)	Cases	5-Year Relative Survival Rate (%)	Cases	5-Year Relative Survival Rate (%)
Total	3,390	75.9	1,566	84.8	1,824	66.4
Stage I	1,567	93.3	882	94.0	685	92.9
Stage II	650	78.7	253	86.0	397	73.1
Stage III	744	52.7	275	70.0	469	39.7
Stage IV	210	28.7	90	40.6	120	16.9
Unknown/Unstaged	219	57.0	66	74.5	153	44.5

Survival by Stage and Grade

Five-year relative survival rates are shown by stage and grade for the 3,390 patients with squamous cell carcinoma in Table 18.10 & Figure 18.3. Within stage, 5-year relative survival declines as grade increases from well differentiated to poorly differentiated/undifferentiated/anaplastic. In stages II and IV, little difference is seen between well- and moderately-differentiated tumors. In Stage III, little difference is seen between moderately differentiated and poorly differentiated/undifferentiated/anaplastic tumors.

Survival by Tumor Size and Nodal Status for Stage III

In stage III tumors, both lymph node status and tumor size are prognostic, with both larger tumors (those greater than 2 cm) and positive regional lymph nodes predicting poorer survival (Table 18.11 and Figure 18.4).

Conditional Survival

Five-year relative survival rates, conditioned on years since diagnosis, are presented in Table 18.12 and Figure 18.5 for squamous cell carcinoma of the vulva. For stage III, the probability of surviving the next 5 years increases

Figure 18.2: Squamous Cell Carcinoma of the Vulva: Relative Survival Rates (%) by AJCC Stage (SEER modified, 5th Edition), Ages 20+, 12 SEER Areas, 1988-2001

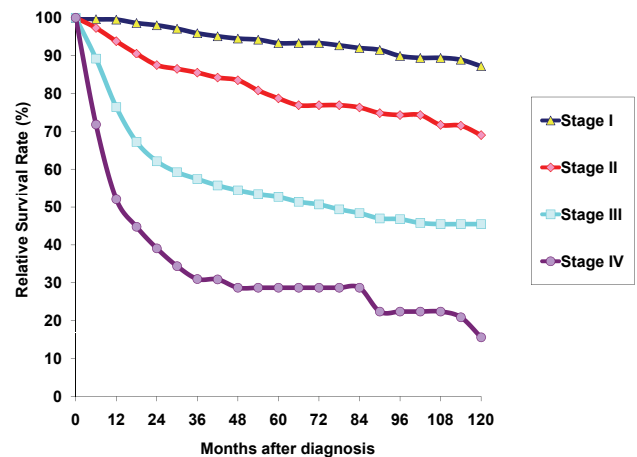


Table 18.8: Squamous Cell Carcinoma of the Vulva: Number and Distribution of Cases and 1-, 2-, 3-, 5-, 8-, & 10-Year Relative Survival Rates (%) by AJCC Stage (SEER modified, 5th Edition), Ages 20+, 12 SEER Areas, 1988-2001

AJCC Stage	Cases	Percent	Relative Survival Rate (%)					
			1-Year	2-Year	3-Year	5-Year	8-Year	10-Year
Total	3,390	100.0	89.1	82.6	79.6	75.9	72.1	69.4
Stage I	1,567	46.2	99.5	98.0	95.9	93.3	89.9	87.2
Stage II	650	19.2	93.8	87.5	85.5	78.7	74.3	69.0
Stage III	744	21.9	76.4	62.1	57.4	52.7	46.8	45.5
Stage IV	210	6.2	52.1	39.1	31.0	28.7	22.4	15.6
Unstaged	219	6.5	77.5	65.9	61.8	57.0	51.4	51.1

Table 18.9: Squamous Cell Carcinoma of the Vulva: Number of Cases and 5-Year Relative Survival Rates (RSR) by Histology and AJCC Stage (SEER modified, 5th Edition), Ages 20+, 12 SEER Areas, 1988-2001

Histology	AJCC Stage											
	Total		I		II		III		IV		Unknown/Unstaged	
	Cases	5-Year RSR (%)	Cases	5-Year RSR (%)	Cases	5-Year RSR (%)	Cases	5-Year RSR (%)	Cases	5-Year RSR (%)	Cases	5-Year RSR (%)
Total	3,390	75.9	1,567	93.3	650	78.7	744	52.7	210	28.7	219	(%)
Squamous, NOS*	2,073	73.7	900	91.5	376	77.8	510	54.7	142	27.6	145	49.1
Squamous, Keratinizing	582	60.2	186	82.7	145	64.4	187	43.7	45	23.9	19	~
Basal Cell	403	99.4	292	100.0	61	97.7	5	~	<5	~	43	85.2
Other Epidermoid	332	84.8	189	94.7	68	88.3	42	57.5	21	~	12	~

~ Statistic not displayed due to less than 25 cases.
 * NOS: Not Otherwise Specified

Table 18.10: Squamous Cell Carcinoma of the Vulva: Number of Cases and 5-Year Relative Survival Rates (%) by AJCC Stage (SEER modified, 5th Edition) and Grade, Ages 20+, 12 SEER Areas, 1988-2001

AJCC Stage	Grade									
	Total		Well Differentiated		Moderately Differentiated		Poorly/Undifferentiated		Unknown	
	Cases	5-Year Relative Survival Rate (%)	Cases	5-Year Relative Survival Rate (%)	Cases	5-Year Relative Survival Rate (%)	Cases	5-Year Relative Survival Rate (%)	Cases	5-Year Relative Survival Rate (%)
Total	3,390	75.9	746	83.6	1,045	66.0	494	49.2	1,105	90.9
Stage I	1,567	93.3	403	95.5	337	86.9	107	77.4	720	96.4
Stage II	650	78.7	167	79.8	242	75.8	87	51.1	154	94.5
Stage III	744	52.7	110	64.6	332	46.6	202	43.4	100	76.2
Stage IV	210	28.7	29	33.7	85	32.0	65	18.5	31	31.3
Unknown/Unstaged	219	57.0	37	41.1	49	50.2	33	43.6	100	68.9

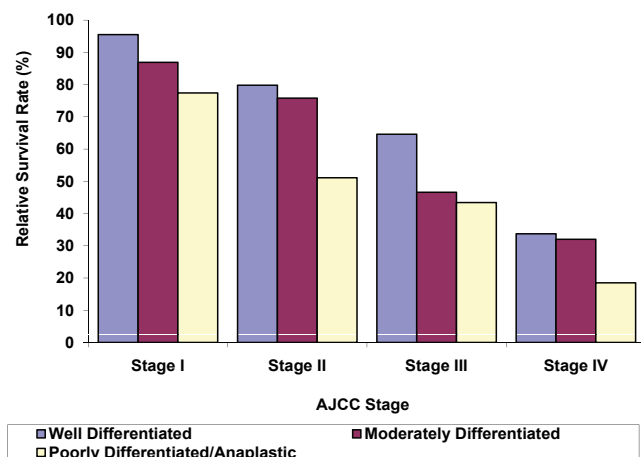
as time since diagnosis increases. For stage IV cases, the 5-year relative survival rate from time of diagnosis is 29%, but for those individuals who survive one year post-diagnosis, 5-year survival increases to 54%. This increases to 71% for those individuals who survive 4 years. However, 5-year survival decreases to 46% for those who have already survived five years after diagnosis. Stage I, which has a 93% 5-year survival rate from diagnosis, exhibits little gain in 5-year survival several years after diagnosis.

Adenocarcinoma

Survival by Stage

Survival by stage at diagnosis is shown for the 347 cases of adenocarcinoma. The most common stage at diagnosis was stage I, with nearly half of the cases. Over 70% were diagnosed in stages I-II (Table 18.13). Very few adult women were diagnosed with stage IV (5%).

Figure 18.3: Squamous Cell Carcinoma of the Vulva: 5-Year Relative Survival Rate (%) by AJCC Stage (SEER modified, 5th Edition) and Grade, Ages 20+, 12 SEER Areas, 1988-2001



Both stages I and II show 5-year relative survival rates of 92% or higher. Five-year relative survival falls to 74.1% at stage III. There are not enough cases to calculate survival at stage IV, (Table 18.13)

Melanoma

Survival by Stage

Melanoma of the vulva has been staged using the melanoma staging scheme (2). Of the 240 cases of melanoma, enough information to analyze stage at diagnosis was available for 223 (93%). The most common stage at diagnosis was stage I with 40% of the cases. Twenty-nine percent of the adult women were diagnosed with stage III and 19% were diagnosed with stage II (Table 18.14).

The Stage I five-year survival rate is 83%. Survival falls to 64% at stage II and 35% at stage III (Table 18.14).

Figure 18.4: Stage III Squamous Cell Carcinoma of the Vulva: 5-Year Relative Survival Rate (%) by Nodal Status and Tumor Size, Ages 20+, 12 SEER Areas, 1988-2001

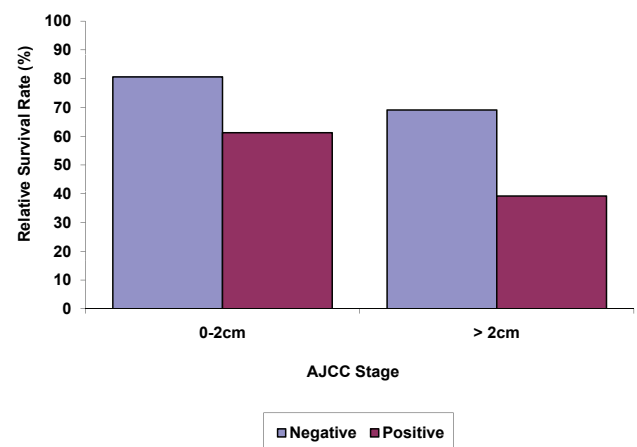


Table 18.11: Stage III Squamous Cell Carcinoma of the Vulva: Number of Cases and 5-Year Relative Survival Rates (%) by Nodal Status and Tumor Size, Ages 20+, 12 SEER Areas, 1988-2001

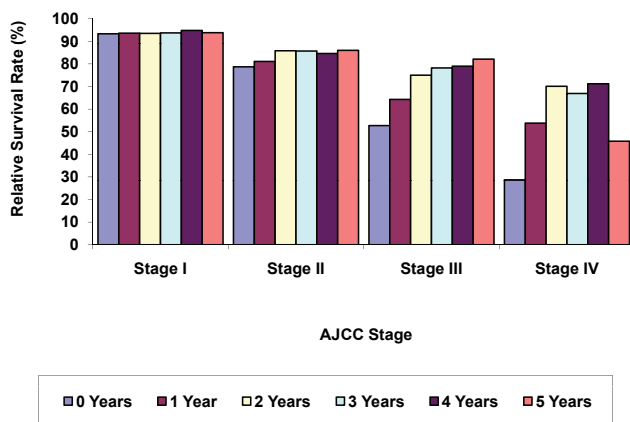
Nodal Status	Tumor Size							
	Total		<= 2cm		> 2cm		Unknown	
	Cases	5-Year Relative Survival Rate (%)	Cases	5-Year Relative Survival Rate (%)	Cases	5-Year Relative Survival Rate (%)	Cases	5-Year Relative Survival Rate (%)
Total	744	52.7	111	67.4	488	48.1	145	54.8
Negative	179	71.9	23	~	114	69.1	42	71.1
Positive	476	43.1	76	61.2	337	39.2	63	39.2
Unknown	89	59.6	12	~	37	52.7	40	57.2

~ Statistic not displayed due to less than 25 cases.

Table 18.12: Squamous Cell Carcinoma of the Vulva: 5-Year Relative Survival Rates (%) , Conditioned on Years Since Diagnosis, by AJCC Stage (SEER modified, 5th Edition), Ages 20+, 12 SEER Areas, 1988-2001

AJCC Stage	5-Year Relative Survival Rate (%)					
	Years Since Diagnosis					
	0	1	2	3	4	5
Total	75.9	83.2	87.4	88.5	89.4	89.3
Stage I	93.3	93.6	93.5	93.7	94.8	93.8
Stage II	78.7	81.1	85.8	85.7	84.6	86.0
Stage III	52.7	64.3	75.0	78.2	79.0	82.1
Stage IV	28.7	53.8	70.1	66.9	71.2	45.8
Unstaged	57.0	68.6	75.1	77.3	78.8	82.9

Figure 18.5: Squamous Cell Carcinoma of the Vulva: 5-Year Relative Survival Rate (%), Conditioned on Years Since Diagnosis, by AJCC Stage (SEER modified, 5th Edition), Ages 20+, 12 SEER Areas, 1988-2001



DISCUSSION

A larger proportion of black women than of white women are diagnosed with vulvar cancer at younger ages. The proportion of adult black women diagnosed before the age of 50 (35%) is nearly double that of whites (17%). Overall, there are no major differences in survival between blacks and whites for cancer of the vulva. For older women, 70 years and over, however, white women have better survival. By stage, there is little difference in survival by age groups (20-69 years compared to 70+ years) for women with stage I (2). In contrast, women over 70 years of age have increasingly worse survival across stages II to IV, compared to women 20-69 years of age with the same stage.

Early stage of disease is associated with the most favorable survival. Among adenocarcinomas, stages I and II have the most favorable survival among this histologic category. Among squamous histologies, representing 83% of all cancers of the vulva, basal cell has a distinct survival advantage in stage I and II over other squamous cell carcinomas. Survival declines with advanced stage of disease.

Table 18.13: Adenocarcinoma of the Vulva: Number and Distribution of Cases and 1-, 2-, 3-, 5-, 8-, & 10-Year Relative Survival Rates (%) by AJCC Stage (SEER modified, 5th Edition), Ages 20+, 12 SEER Areas, 1988-2001

AJCC Stage	Cases	Percent	Relative Survival Rate (%)					
			1-Year	2-Year	3-Year	5-Year	8-Year	10-Year
Total	347	100.0	96.2	95.1	93.1	91.9	89.9	82.5
Stage I	172	49.6	100.0	100.0	100.0	100.0	97.5	88.5
Stage II	75	21.6	98.6	98.6	94.1	92.2	86.7	72.0
Stage III	51	14.7	97.6	86.5	83.3	74.1	74.1	70.9
Stage IV	18	5.2	~	~	~	~	~	~
Unstaged	31	8.9	91.0	91.0	90.9	84.8	84.8	84.8

~ Statistic not displayed due to less than 25 cases.

Table 18.14: Melanoma of the Vulva: Number and Distribution of Cases and 1-, 2-, 3-, 5-, 8-, & 10-Year Relative Survival Rates (%) by AJCC Stage (SEER modified, 5th Edition - Melanoma Staging Used), Ages 20+, 12 SEER Areas, 1988-2001

AJCC Stage	Cases	Percent	Relative Survival Rate (%)					
			1-Year	2-Year	3-Year	5-Year	8-Year	10-Year
Total	240	100.0	89.1	72.3	63.5	58.9	53.0	46.6
I	96	40.0	99.4	91.8	84.5	83.0	76.5	71.3
II	45	18.8	96.2	76.9	76.4	64.3	64.3	57.4
III	69	28.8	81.1	55.7	39.7	35.1	27.0	21.5
IV	13	5.4	~	~	~	~	~	~
Unknown/Unstaged	17	7.1	~	~	~	~	~	~

~ Statistic not displayed due to less than 25 cases.

REFERENCES

1. American Cancer Society, Cancer Facts and Figures 2006, American Cancer Society, #500806, 2006.
2. Fleming ID, Cooper JS, Henson DE, Hutter RVP, Kennedy BJ, Murphy GP, O'Sullivan B, Sobin LH, Yarbro, JW (eds). AJCC Cancer Staging Manual, Fifth edition, American Joint Committee on Cancer. Philadelphia: Lippincott-Raven, 1997.