Chapter 30
Cancers of Rare Sites

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INTRODUCTION

There are some anatomic sites in which cancer rarely occurs, for example, the eye, the ureter, the pituitary gland, etc. Most of these rare sites have been excluded from other chapters in this monograph due to the small number of cases involved. This chapter provides a limited analysis of survival for these rare sites and for cancers which could not be assigned to a specific anatomic location at the time of diagnosis due to the advanced stage of the tumor. Cases were obtained from the Surveillance, Epidemiology, and End Results (SEER) Program of the National Cancer Institute (NCI).

MATERIALS AND METHODS

The NCI contracts with medically-oriented, nonprofit institutions located in specific geographic areas to obtain data on all cancers diagnosed in residents of the SEER geographic areas. SEER collects data on all invasive and in situ cancers except basal cell and squamous cell carcinomas of the skin (of non-genital anatomic sites) and in situ carcinomas of the uterine cervix. SEER actively follows all previously diagnosed patients on an annual basis to obtain vital status allowing the calculation of observed and relative survival rates.

This analysis is based on data from 12 SEER geographic areas which collectively cover about 14% of the total US population. The areas are the States of Connecticut, Iowa, New Mexico, Utah, and Hawaii; the metropolitan areas of Detroit, Michigan; Atlanta, Georgia; San Francisco, San Jose, and Los Angeles, California; Seattle, Washington; and 10 counties in rural Georgia. Los Angeles contributed data for diagnosis years 1992 to 2001, all other areas for 1988-2001.

With the exception of male breast cancers, in situ diagnoses have been excluded. Cases diagnosed in children and adolescents aged 0-19 have also been excluded. Some patients have more than one diagnosis of cancer, but only the first diagnosis of cancer has been included. Death certificate only cases, autopsy only cases, and alive cases with no survival time have been excluded. All sarcomas arising in these rare sites have been excluded since they are included in the chapter on sarcomas included elsewhere in this monograph (1). Finally, with the exception of cancers where the primary site could not be determined, cases with no microscopic confirmation have been included.

Survival analysis is based on relative survival rates calculated by the life-table (actuarial) method. Relative survival, defined as observed survival in the cohort divided by expected survival in the cohort, adjusts for the expected mortality that the cohort would experience from other causes of death. Expected survival is based on decennial life tables for the United States in 1990.

In many of the chapters in this monograph, data on stage are presented using the Stage I-IV definitions of the American Joint Committee on Cancer (AJCC) 3rd or 5th Editions. However, for many of the rare tumor sites there are no appropriate AJCC staging definitions. Therefore, the staging definitions utilized in this chapter are those of the 1977 Summary Staging Guide (2). For simplicity, all categories of regional disease have been added together into a single group.

RESULTS

Table 30.1 shows the anatomic sites included in this analysis along with the numbers of cases diagnosed during 1988-2001 classified by sex, by race, and by stage of diagnosis. The sites have been arranged by body systems and show the specific rare cancer sites under the appropriate body system. Relative survival rates at 1, 3, 5, and 10 years are shown in Table 30.2 for both sexes and for males and females separately where appropriate. Table 30.3 presents rates for whites and for blacks where the number of cases permitted meaningful analyses. Finally, Table 30.4 presents relative survival rates by SEER Summary Stage.

Table 30.1: Cancers of rare sites: Number and Distribution of Cases by Primary Site, Sex, Race and SEER Summary Stage 1977 (2), Ages 20+, 12 SEER Areas, 1988-2001

Primary Site	Total	Sex		Race		Stage (%)			
		Male	Female	White	Black	Loc	Reg	Dis	Uns
Respiratory and Intrathoracic Organs									
Nose, Nasal Cavity and Middle Ear	2,299	1,329	970	1,842	223	25.8	51.3	14.1	8.7
Nasal Cavity	1,009	571	438	868	59	46.2	30.7	12.0	11.1
Middle Ear	82	38	44	67	8	17.1	50.0	20.7	12.2
Trachea, Mediastinum and Other Respiratory	573	427	146	474	50	20.8	33.0	24.6	21.6
Trachea	211	112	99	173	20	24.2	42.2	16.1	17.5
Squamous Cell	105	65	40	89	12	21.0	41.9	20.0	17.1
Mediastinum	315	282	33	258	27	21.6	30.5	30.8	17.1
Mediastinum - Germ	250	236	14	205	20	24.4	28.4	28.8	18.4
Other Respiratory	47	33	14	43	<5	0.0	8.5	21.3	70.2
Pleura*	49	28	21	40	<5	16.3	14.3	24.5	44.9
Skin^	2,793	1,517	1,276	2,539	105	73.4	7.4	5.9	13.4
Merkel Cell	1,076	617	459	1,021	9	66.5	11.1	8.2	14.2
Skin Appendage Adenocarcinoma	383	199	184	356	13	77.0	7.6	0.8	14.6
Sweat Gland Adenocarcinoma	208	125	83	185	13	77.4	6.3	2.9	13.5
Sebaceous Adenocarcinoma	458	229	229	413	13	79.9	1.7	6.8	11.6
Peritoneum and Retroperitoneum*	1,461	209	1,252	1,300	80	7.3	15.1	70.9	6.6
Papillary Serous Cystadenocarcinoma	636	<5	635	579	24	1.1	11.3	85.7	1.9
Male Breast (including in situ)	1,905	1,905	0	1,578	232	43.6	37.9	5.2	2.5
In situ	205	205	0	177	20	0.0	0.0	0.0	0.0
Invasive	1,700	1,700	0	1,401	212	48.8	42.5	5.9	2.8
Female Genital									
Ligaments and Adnexa	49	0	49	37	8	44.9	10.2	40.8	4.1
Overlapping	60	0	60	49	7	35.0	21.7	31.7	11.7
Other and Not Otherwise Specified	192	0	192	160	20	0.5	12.0	50.0	37.5
Male Genital									
Penis	1,132	1,132	0	976	94	61.2	29.9	4.3	4.6
Penis - Squamous Cell Carcinoma	996	996	0	858	87	60.5	31.1	4.3	4.0
Scrotum	233	233	0	149	22	69.5	19.3	3.0	8.2
Scrotum - Squamous Cell Carcinoma	87	87	0	65	18	67.8	21.8	4.6	5.7
Scrotum - Paget Disease	74	74	0	34	0	70.3	21.6	1.4	6.8
Other and Not Otherwise Specified	27	27	0	24	0	48.1	18.5	14.8	18.5

* Excludes mesotheliomas and sarcomas
^ Excludes Basal, Squamous, Kaposi sarcoma & Melanoma

Respiratory and Intrathoracic Organs

Cancer of the nose and nasal cavities had the best survival among this group of rare sites. Five-year relative survival rate was 54% overall and was similar for males and females but was better for whites vs. blacks. Cancers of the nasal cavity tended to be diagnosed at an earlier stage which doubtless contributed to a better overall 5-year relative survival rate of 70%. Cancers of the middle ear were much rarer (82 cases during the time period) and relative survival rate was much poorer, 34% at five years (Table 30.2). Some additional tables for nose and nasal cavities are found in the chapter on head and neck cancer (3).

Relative survival rates for cancers of the trachea and pleura were poor (35 % and 28%, respectively, at five years) but were better than that for lung cancer (16%) (4). Not surprisingly, most cases were diagnosed with regional or distant disease. For cancers of the mediastinum, the five-year relative survival rate was 48% and was higher among males (50%) than among females (30%). For all respiratory sites, survival was markedly lower for patients diagnosed

Table 30.1 (continued): Cancers of Rare Sites: Number and Distribution of Cases by Primary Site, Sex, Race and SEER Summary Stage 1977 (2), Ages 20+, 12 SEER Areas, 1988-2001

Primary Site	Total	Sex		Race		Stage (%)			
		Male	Female	White	Black	Loc	Reg	Dis	Uns
Urinary System									
Ureter	1,333	808	525	1,158	42	52.6	28.5	10.4	8.6
Ureter - Papillary Transitional Cell	1,251	770	481	1,086	37	54.1	28.5	9.4	7.9
Other Urinary	850	539	311	626	174	27.4	24.8	14.0	33.8
Other Urinary - Papillary Transitional Cell	445	339	106	355	63	28.8	18.9	12.1	40.2
Eye and Orbit	1,904	1,069	835	1,824	33	77.6	7.2	3.0	12.2
Eye and Orbit - Squamous Cell Carcinoma	305	240	65	278	7	74.1	9.5	3.3	13.1
Eye and Orbit - Melanoma	1,504	779	725	1,469	17	80.3	5.3	2.3	12.1
Other Endocrine	1,727	921	806	1,301	184	29.6	36.5	24.8	9.1
Thymus	826	465	361	558	101	20.0	50.6	20.2	9.2
Thymus - Thymoma	678	367	311	441	89	18.9	52.1	19.3	9.7
Adrenal Gland	598	283	315	513	47	37.3	16.9	37.3	8.5
Adrenal Gland - Adrenal Cortical	387	170	217	340	21	43.4	20.2	31.5	4.9
Adrenal Gland - Pheochromocytoma	66	34	32	46	15	40.9	12.1	25.8	21.2
Parathyroid Gland	115	56	59	89	16	51.3	40.0	4.3	4.3
Pituitary Gland	52	25	27	37	8	26.9	40.4	9.6	23.1
Pineal Gland	95	68	27	71	8	48.4	27.4	14.7	9.5
Mesothelioma	3,562	2,795	767	3,239	182	10.9	18.5	58.5	12.1
Mesothelioma - Pleura and Lung	3,148	2,548	600	2,860	160	11.0	19.0	58.4	11.5
Mesothelioma - Peritoneum and Retroperitoneum	354	212	142	328	15	6.5	14.1	65.3	14.1
Reticuloendothelial System Tumors	1,297	779	779	1,174	51	0.0	0.0	99.7	0.3
Waldenstrom's Macroglobulinemia	1,161	709	452	1,050	46	0.0	0.0	100.0	0.0
Myeloma	18,446	9,879	8,567	14,282	3,024	4.5	0.0	95.5	0.0
Solitary Myeloma	526	328	198	433	54	87.3	0.0	12.7	0.0
Multiple Myeloma	17,217	9,103	8,114	13,281	2,870	0.0	0.0	100.0	0.0
Unknown or Ill-defined Primary Site	39,140	18,587	20,553	32,097	4,310	0.0	0.0	0.1	99.9
Microscopically confirmed	30,382	14,917	15,465	24,852	3,334	0.0	0.0	0.1	99.9
Carcinomas	10,700	6,090	4,610	8,891	1,096	0.0	0.0	0.0	100.0
Adenocarcinomas	16,654	7,286	9,368	13,494	1,875	0.0	0.0	0.0	100.0
Other	3,028	1,541	1,487	2,467	363	0.0	0.0	1.3	98.7
Non-microscopically confirmed	8,758	3,670	5,088	7,245	976	0.0	0.0	0.0	100.0

* Excludes mesotheliomas and sarcomas
^ Excludes Basal, Squamous, Kaposi sarcoma & Melanoma
Loc, Localized; Reg, Regional; Dis, Distant; Uns, Unstaged

with distant disease, especially for patients with tracheal cancer (3% at 5 years) (Table 30.4).

Skin

Basal and squamous cell skin cancers are not reported to the SEER program except those of the genital sites. Melanomas of the skin and Kaposi sarcoma of the skin and of visceral organs are discussed elsewhere in this monograph. Still there are other types of cancers which occur, albeit rarely, on the skin. The four major types are Merkel cell, skin appendage adenocarcinomas, sweat gland adenocarcinomas, and sebaceous adenocarcinomas. Among this group of cancers, patients diagnosed with Merkel cell survived more poorly than those with other histologic types; there were more patients with Merkel cell diagnosed at regional or distant stages when compared to the other three groups. The overall 5-year relative rates for the various subtypes were Merkel cell, 63% ; skin appendage 98%; sweat gland, 95%; and sebaceous, 95% (Table 30.2). In general, females

Table 30.2: Cancers of Rare Sites: 1-, 3- ,5- and 10-Year (Yr) Relative Survival Rates (%) by Primary Site and Sex, Ages 20+, 12 SEER Areas, 1988-2001

Primary Site	Relative Survival Rate (%)											
	Total				Male				Female			
	1-Yr	3-Yr	5-Yr	10-Yr	1-Yr	3-Yr	5-Yr	10-Yr	1-Yr	3-Yr	5-Yr	10-Yr
Respiratory and Intrathoracic Organs												
Nose, Nasal Cavity and Middle Ear	79.8	61.9	54.0	46.4	80.8	62.4	54.4	47.2	78.3	61.1	53.6	44.9
Nasal Cavity	89.9	77.6	70.1	61.7	89.9	77.6	69.4	62.8	89.9	77.5	70.8	59.7
Middle Ear	68.7	38.5	33.8	27.7	65.7	36.7	36.7	29.6	71.3	39.9	30.4	27.0
Trachea, Mediastinum and Other Respiratory	60.6	45.0	42.3	37.0	61.2	46.1	43.9	38.9	59.0	41.8	37.1	30.6
Trachea	57.7	40.2	34.6	25.5	55.5	36.7	30.5	20.5	60.2	43.8	38.7	28.0
Squamous Cell	47.4	27.6	20.4	13.2	46.6	28.4	21.2	16.2	48.6	26.4	19.1	7.5
Mediastinum	65.5	49.9	48.2	44.0	66.7	51.8	50.3	45.6	55.2	32.9	30.1	25.1
Mediastinum - Germ	69.5	56.8	55.6	51.5	69.4	57.7	56.3	52.0	~	~	~	~
Other Respiratory	39.8	31.0	31.0	15.2	31.6	24.1	24.1	8.3	~	~	~	~
Pleura *	40.4	29.9	28.2	21.2	30.0	19.6	19.6	0.0	~	~	~	~
Skin ^	94.2	85.9	84.0	81.2	93.3	83.5	81.1	79.3	95.2	88.6	87.1	81.9
Merkel Cell	87.4	68.6	62.8	57.5	87.2	65.1	58.6	55.2	87.6	73.2	67.8	60.0
Skin Appendage Adenocarcinoma	99.8	99.1	97.5	97.2	99.8	99.8	97.8	97.8	99.0	96.4	95.7	90.5
Sweat Gland Adenocarcinoma	97.8	94.9	94.5	83.6	95.6	91.8	91.6	78.2	100.0	99.1	97.3	88.9
Sebaceous Adenocarcinoma	98.2	94.6	94.6	87.3	95.2	90.5	90.5	82.8	100.0	98.7	98.7	91.2
Peritoneum and Retroperitoneum *	78.1	49.0	34.3	28.5	78.8	60.6	54.5	51.4	77.9	46.9	30.4	22.8
Papillary Serous Cystadenocarcinoma	84.8	49.5	26.9	17.2	~	~	~	~	84.8	49.6	26.9	17.3
Male Breast (including in situ)	97.3	93.0	85.6	74.2	97.3	93.0	85.6	74.2	N/A	N/A	N/A	N/A
In situ	99.1	99.1	99.1	96.9	99.1	99.1	99.1	96.9	N/A	N/A	N/A	N/A
Invasive	97.0	92.0	83.6	71.3	97.0	92.0	83.6	71.3	N/A	N/A	N/A	N/A
Female Genital												
Ligaments and Adnexa	87.1	72.2	67.1	53.2	N/A	N/A	N/A	N/A	87.1	72.2	67.1	53.2
Overlapping	70.7	55.2	48.8	33.0	N/A	N/A	N/A	N/A	70.7	55.2	48.8	33.0
Other and Not Otherwise Specified	50.4	28.8	25.6	19.1	N/A	N/A	N/A	N/A	50.4	28.8	25.6	19.1
Male Genital												
Penis	88.8	79.1	73.5	64.0	88.8	79.1	73.5	64.0	N/A	N/A	N/A	N/A
Penis - Squamous Cell Carcinoma	87.4	76.7	71.7	62.6	87.4	76.7	71.7	62.6	N/A	N/A	N/A	N/A
Scrotum	94.7	82.0	77.5	62.5	94.7	82.0	77.5	62.5	N/A	N/A	N/A	N/A
Scrotum - Squamous Cell Carcinoma	85.4	63.4	58.6	38.5	85.4	63.4	58.6	38.5	N/A	N/A	N/A	N/A
Scrotum - Paget Disease	100.0	93.0	89.0	82.3	100.0	93.0	89.0	82.3	N/A	N/A	N/A	N/A
Other and Not Otherwise Specified	73.0	62.1	51.8	34.1	73.0	62.1	51.8	34.1	N/A	N/A	N/A	N/A

* Excludes mesotheliomas and sarcomas
^ Excludes Basal, Squamous, Kaposi sarcoma & Melanoma
~ Statistic not displayed due to less than 25 cases.

had better survival rates at both 5 and 10 years. Overall, blacks had higher relative survival rates than whites at 1, 3, and 5 years, but whites had higher rates after 10 years. With the exception of patients with Merkel cell, 5-year relative survival was 97% or higher for all other types diagnosed while localized to the skin.

Peritoneum and Retroperitoneum

Over 70% of cancers occurring in the retroperitoneum were already distant at the time of diagnosis accounting for overall poor relative survival rate, 34% at five-years. Over forty percent of these tumors were papillary serous cystadenocarcinomas which are probably extra-ovarian tumors. Survival from these cancers was even poorer, 27% at five-years. For peritoneum and retroperitoneum

Table 30.2 (continued): Cancers of Rare Sites: 1-, 3- ,5- and 10-Year (Yr) Relative Survival Rates (%) by Primary Site and Sex, Ages 20+, 12 SEER Areas, 1988-2001

	Relative Survival Rate (%)											
	Total				Male				Female			
Primary Site	1-Yr	3-Yr	5-Yr	10-Yr	1-Yr	3-Yr	5-Yr	10-Yr	1-Yr	3-Yr	5-Yr	10-Yr
Urinary System												
Ureter	82.3	63.4	55.9	49.6	82.4	63.4	55.6	50.2	82.1	63.3	56.0	46.7
Ureter - Papillary Transitional Cell	83.5	64.6	57.8	51.4	83.4	64.7	57.7	52.1	83.7	64.6	57.5	48.1
Other Urinary	80.5	67.0	60.5	54.2	82.7	71.4	67.6	60.8	76.8	59.5	48.5	42.2
Other Urinary - Papillary Transitional Cell	81.4	68.4	63.8	60.0	83.6	71.9	68.2	62.1	74.2	57.3	49.5	46.3
Eye and Orbit	97.4	85.2	75.4	63.1	97.4	85.9	77.1	65.8	97.3	84.3	73.0	59.8
Eye and Orbit - Squamous Cell Carcinoma	96.5	90.4	86.8	77.3	98.0	91.6	87.7	82.6	90.9	85.2	83.6	54.9
Eye and Orbit - Melanoma	97.8	84.5	73.6	61.1	97.5	84.9	74.8	62.5	98.1	84.1	72.1	59.4
Other Endocrine	78.2	66.2	58.9	47.2	79.2	66.3	59.6	47.5	77.0	66.0	58.1	46.8
Thymus	87.4	74.5	66.3	51.3	88.9	75.0	68.5	54.5	85.5	73.9	63.4	46.8
Thymus - Thymoma	89.0	77.4	70.0	55.7	90.8	76.8	71.4	60.7	86.9	78.3	68.2	49.5
Adrenal Gland	59.4	45.5	38.7	29.6	55.7	40.9	34.1	22.7	62.6	49.6	42.5	35.4
Adrenal Gland - Adrenal Cortical	66.0	48.5	41.2	31.3	62.9	43.7	37.9	27.0	68.4	52.1	43.6	34.6
Adrenal Gland - Pheochromocytoma	84.5	73.0	64.8	44.0	83.9	73.4	67.3	29.7	85.1	72.6	62.4	58.9
Parathyroid Gland	94.1	94.1	93.1	81.6	93.8	93.8	89.6	71.7	94.0	93.1	93.1	88.4
Pituitary Gland	83.7	74.9	63.8	41.7	85.5	70.9	52.6	25.7	81.9	78.9	74.4	53.4
Pineal Gland	87.7	76.2	71.7	63.9	90.1	80.0	75.2	69.8	81.7	66.2	62.4	47.8
Mesothelioma	39.0	12.8	8.2	5.6	37.0	9.8	5.5	3.0	46.1	23.1	17.3	12.9
Mesothelioma - Pleura and Lung	38.2	10.5	6.4	4.3	36.9	8.5	4.8	2.4	43.7	18.7	12.8	9.9
Mesothelioma - Peritoneum and Retroperitoneum	41.8	25.9	18.4	9.5	34.1	19.0	11.7	5.6	53.1	35.6	28.2	14.7
Reticuloendothelial System Tumors	88.0	78.5	66.6	44.0	88.9	77.3	65.6	45.6	86.6	80.3	67.9	41.9
Waldenstrom's Macroglobulinemia	90.2	81.0	69.3	45.6	91.6	80.0	67.9	47.4	88.1	82.6	71.3	43.1
Myeloma	74.7	48.1	31.7	14.2	75.1	49.4	33.4	15.7	74.1	46.7	29.8	12.6
Solitary Myeloma	90.3	73.7	64.9	44.1	90.4	78.8	69.1	52.8	90.0	65.2	57.5	29.1
Multiple Myeloma	73.7	46.4	29.4	11.7	74.0	47.2	30.6	12.3	73.4	45.6	28.3	11.1
Unknown or Ill-defined Primary Site	24.2	13.5	11.3	9.8	25.2	14.8	12.8	11.7	23.2	12.4	10.0	8.1
Microscopically confirmed	27.4	15.0	12.4	10.6	28.4	16.5	14.1	12.7	26.5	13.6	10.7	8.8
Carcinomas	35.9	23.4	20.9	18.3	39.6	26.6	24.0	21.7	30.9	19.3	16.8	14.0
Adenocarcinomas	19.4	7.5	5.2	4.1	16.3	5.8	4.0	3.3	21.8	8.7	6.0	4.7
Other	41.2	26.6	21.8	18.5	40.9	27.1	22.6	19.9	41.5	26.1	21.0	16.7
Non-microscopically confirmed	12.5	7.7	6.6	5.4	12.0	7.0	5.9	5.4	12.9	8.1	7.1	5.4

* Excludes mesotheliomas and sarcomas
^ Excludes Basal, Squamous, Kaposi sarcoma & Melanoma
~ Statistic not displayed due to less than 25 cases.

combined, males had a survival advantage at 1, 3, 5, and 10 years, and the relative survival rate for whites was considerably higher than that of blacks at five years, 35% vs. 24% (Table 30.3).

Male Breast

About one percent of all breast cancers occur among males. As among females, some cases are diagnosed at the in situ stage. The majority of male breast cancers are ductal carcinomas. Relative survival rates for cases diagnosed at the in situ stage are quite good, overall, 99% at five-years. Among the invasive tumors, almost half were diagnosed while still localized. Five-year relative survival for all males was 84% (compared to 89% in females) (5) with white males having a survival advantage of 86% vs. 67% among black males. A similar advantage existed at 10 years also, 73% among whites vs. 52% among blacks. Five-year relative survival for males with localized breast cancer was 97%.

Female genital

Survival from cancers of the vagina, vulva, uterus, ovary, fallopian tube, and placenta are included in other chapters in this monograph. Among the remaining cases occurring within specific female genital sites – round and broad ligaments, parametrium, and adnexa – there were too few cases for a separate analysis, so these sites have been grouped together. The five-year relative survival rate for this group of 67% was intermediate to that for ovary (44%) (6) vs. cervix (72%) (7) and corpus uteri (85%) (8), this despite the fact that over 40% of the patients were diagnosed at an advanced stage. For those women whose cancer overlapped two or more sites within the female genital tract, relative survival rate was much poorer, 49% at five years, even though only one third of these women were diagnosed with distant disease. The poorest relative survival rate was experienced by those women whose cancer could not be specifically assigned to any specific genital site at the time of diagnosis, only

Table 30.3: Cancers of Rare Sites: 1,3,5 and 10-Year Relative Survival Rates (%) by Site and Race, 12 SEER Areas, 1988-2001

Site	Relative Survival Rate (%)							
	White				Black			
	1-Year	3-Year	5-Year	10-Year	1-Year	3-Year	5-Year	10-Year
Respiratory and Intrathoracic Organs								
Nose, Nasal Cavity and Middle Ear	80.4	63.5	56.4	48.1	70.4	47.1	36.3	30.5
Nasal Cavity	90.0	77.9	71.0	61.4	81.3	64.1	59.1	58.4
Middle Ear	68.8	41.1	39.2	31.5	~	~	~	~
Trachea, Mediastinum and Other Respiratory	61.4	45.8	43.5	39.0	51.1	30.2	21.1	17.4
Trachea	55.8	38.9	35.1	27.8	~	~	~	~
Squamous Cell	47.8	29.0	23.5	15.0	~	~	~	~
Mediastinum	68.3	52.1	50.0	46.0	52.6	31.6	31.6	22.8
Mediastinum - Germ	71.1	58.2	56.7	53.3	~	~	~	~
Other Respiratory	41.1	31.4	31.3	9.7	~	~	~	~
Pleura *	39.0	25.0	22.2	12.8	~	~	~	~
Skin ^	94.1	85.4	83.3	81.3	97.1	87.7	87.7	70.9
Merkel Cell	87.6	68.4	62.5	57.2	~	~	~	~
Skin Appendage Adenocarcinoma	99.7	98.8	96.6	96.5	~	~	~	~
Sweat Gland Adenocarcinoma	97.8	95.3	94.8	85.4	~	~	~	~
Sebaceous Adenocarcinoma	97.8	94.1	94.1	91.2	~	~	~	~
Peritoneum and Retroperitoneum *	78.1	49.3	35.0	28.9	80.6	45.5	23.5	9.6
Papillary Serous Cystadenocarcinoma	83.7	48.9	27.0	16.7	~	~	~	~
Male Breast (including in situ)	98.1	94.5	87.7	75.1	92.6	81.8	70.4	58.1
In situ	99.2	99.2	99.2	93.2	~	~	~	~
Invasive	97.9	93.7	85.9	72.6	92.1	80.2	67.2	52.4
Female Genital								
Ligaments and Adnexa	93.4	72.9	70.5	54.3	~	~	~	~
Overlapping	71.6	57.4	51.9	36.3	~	~	~	~
Other and Not Otherwise Specified	50.7	30.7	27.4	21.4	47.2	25.7	25.7	16.9
Male Genital								
Penis	90.0	79.9	73.9	64.6	77.3	64.3	64.3	52.4
Penis - Squamous Cell Carcinoma	88.6	77.6	71.9	62.9	77.3	64.3	64.3	52.4
Scrotum	97.5	88.3	82.5	67.0	~	~	~	~
Scrotum - Squamous Cell Carcinoma	90.4	72.8	67.0	47.5	~	~	~	~
Scrotum - Paget Disease	100.0	100.0	94.4	86.8	~	~	~	~
Other and Not Otherwise Specified	~	~	~	~	~	~	~	~

* Excludes mesotheliomas and sarcomas
^ Excludes Basal, Squamous, Kaposi sarcoma & Melanoma
~ Statistic not displayed due to less than 25 cases.

26% at five years. There were too few cases occurring among blacks to allow a comparison of white vs. black females.

Male genital

Survival from cancers of the prostate and testis are analyzed elsewhere in this monograph. Cancer of the penis is indeed rare within the United States, and the majority of the tumors occurs on the skin of the penis and is of squamous cell origin. Relative survival rates were quite high and were better for white males than for black males. Similarly, the majority of other cancers occurring in the remainder of the male genital system was of the skin of the scrotum and was squamous cell in origin. However, men with squamous cell carcinoma of the scrotum versus the penis survived more poorly at 5 years with relative rates of 59% vs. 72%, respectively. Men with localized squamous cell carcinoma of the scrotum had much poorer 5-year

Table 30.3 (continued): Cancers of Rare Sites: 1,3,5 and 10-Year Relative Survival Rates (%) by Site and Race, 12 SEER Areas, 1988-2001

Site	Relative Survival Rate (%)							
	White				Black			
	1-Year	3-Year	5-Year	10-Year	1-Year	3-Year	5-Year	10-Year
Urinary System								
Ureter	82.8	64.6	57.2	50.7	72.5	53.9	40.3	39.0
Ureter - Papillary Transitional Cell	84.3	66.2	59.1	52.6	68.3	51.7	44.2	42.5
Other Urinary	81.3	68.9	63.9	58.8	77.2	58.5	48.1	40.3
Other Urinary - Papillary Transitional Cell	82.3	69.3	64.9	61.9	76.8	60.2	55.6	47.6
Eye and Orbit	97.5	85.1	75.7	63.7	89.7	75.8	41.7	10.1
Eye and Orbit - Squamous Cell Carcinoma	96.0	89.9	86.4	76.7	~	~	~	~
Eye and Orbit - Melanoma	97.9	84.5	74.2	61.7	~	~	~	~
Other Endocrine	76.5	64.3	57.5	46.4	79.5	70.4	61.5	44.8
Thymus	86.8	73.6	65.9	49.4	85.5	76.2	66.7	51.1
Thymus - Thymoma	87.9	77.0	70.1	54.2	90.2	79.4	71.1	54.6
Adrenal Gland	58.5	45.3	39.1	31.1	64.9	47.6	37.3	13.1
Adrenal Gland - Adrenal Cortical	65.9	50.0	43.5	33.5	~	~	~	~
Adrenal Gland - Pheochromocytoma	81.6	66.9	60.3	44.9	~	~	~	~
Parathyroid Gland	94.2	93.7	92.8	86.5	~	~	~	~
Pituitary Gland	89.9	78.0	66.3	40.4	~	~	~	~
Pineal Gland	87.7	73.3	67.5	60.2	~	~	~	~
Mesothelioma	39.4	12.8	8.1	5.3	37.9	12.0	11.7	8.6
Mesothelioma - Pleura and Lung	38.8	10.7	6.3	4.1	37.3	9.4	8.8	6.2
Mesothelioma - Peritoneum and Retroperitoneum	40.7	24.4	17.4	8.4	~	~	~	~
Reticuloendothelial System Tumors	87.8	78.9	66.7	43.9	95.6	76.1	68.2	46.6
Waldenstrom's Macroglobulinemia	90.3	81.5	69.7	45.4	94.9	77.3	71.4	51.6
Myeloma	74.2	48.0	31.2	13.7	76.4	49.5	34.1	15.7
Solitary Myeloma	89.8	74.1	63.7	42.1	92.9	68.7	68.0	40.3
Multiple Myeloma	73.2	46.2	28.8	11.0	75.7	48.5	32.6	14.1
Unknown or Ill-defined Primary Site	24.5	13.9	11.6	10.0	21.6	11.0	8.9	7.8
Microscopically confirmed	28.1	15.7	12.9	11.0	22.2	10.6	8.6	7.7
Carcinomas	37.4	24.9	22.3	19.4	24.9	13.5	11.4	10.1
Adenocarcinomas	19.6	7.5	5.2	4.0	16.9	5.7	4.1	3.5
Other	41.4	26.7	21.6	18.3	40.8	26.2	23.0	19.9
Non-microscopically confirmed	11.3	6.8	5.9	4.6	19.6	12.6	10.1	8.4

* Excludes mesotheliomas and sarcomas
^ Excludes Basal, Squamous, Kaposi sarcoma & Melanoma
~ Statistic not displayed due to less than 25 cases.

Table 30.4: Cancers of Rare Sites: 5- & 10-Year (Yr) Relative Survival Rates (%) by Primary Site and SEER Summary Stage 1977 (2), 12 SEER Areas, 1988-2001

Primary Site	Summary Stage									
	Total		Local		Regional		Distant		Unstaged	
	Relative Survival Rate (%)		Relative Survival Rate (%)		Relative Survival Rate (%)		Relative Survival Rate (%)		Relative Survival Rate (%)	
	5-Yr	10-Yr	5-Yr	10-Yr	5-Yr	10-Yr	5-Yr	10-Yr	5-Yr	10-Yr
Respiratory and Intrathoracic Organs										
Nose, Nasal Cavity and Middle Ear	54.0	46.4	82.5	77.4	47.3	37.6	25.3	21.7	55.6	45.6
Nasal Cavity	70.1	61.7	86.3	79.4	57.5	47.0	37.6	29.7	73.2	63.6
Middle Ear	33.8	27.7	~	~	37.7	29.2	~	~	~	~
Trachea, Mediastinum and Other Respiratory	42.3	37.0	66.2	60.0	44.2	36.2	17.1	13.6	43.5	35.8
Trachea	34.6	25.5	59.7	43.5	31.2	20.7	3.4	!	32.8	21.6
Squamous Cell	20.4	13.2	~	~	12.4	12.4	~	~	~	~
Mediastinum	48.2	44.0	69.9	67.7	54.6	46.0	23.3	17.5	53.0	50.6
Mediastinum - Germ	55.6	51.5	73.4	70.9	62.0	59.8	29.5	21.5	60.1	57.7
Other Respiratory	31.0	15.2	!	!	~	~	~	~	36.1	18.1
Pleura *	28.2	21.2	~	~	~	~	~	~	~	~
Skin ^	84.0	81.2	91.7	87.6	62.1	58.3	46.4	44.1	70.0	68.8
Merkel Cell	62.8	57.5	74.7	68.4	38.7	32.4	28.4	28.4	45.6	39.2
Skin Appendage Adenocarcinoma	97.5	97.2	99.1	98.6	88.2	79.1	~	~	86.8	84.7
Sweat Gland Adenocarcinoma	94.5	83.6	97.4	87.9	~	~	~	~	80.9	73.6
Sebaceous Adenocarcinoma	94.6	87.3	97.1	84.2	~	~	85.4	85.4	86.0	86.0
Peritoneum and Retroperitoneum *	34.3	28.5	77.7	74.1	52.6	41.5	25.0	18.9	39.5	36.5
Papillary Serous Cystadenocarcinoma	26.9	17.2	~	~	46.3	!	23.8	16.4	~	~
Male Breast (including in situ)	85.6	74.2	96.9	92.1	78.1	58.1	23.0	5.8	64.2	34.8
In situ	99.1	96.9	!	!	!	!	!	!	!	!
Invasive	83.6	71.3	96.9	92.1	78.1	58.1	23.0	5.8	64.2	34.8
Female Genital										
Ligaments and Adnexa	67.1	53.2	~	~	~	~	~	~	~	~
Overlapping	48.8	33.0	~	~	~	~	~	~	~	~
Other and Not Otherwise Specified	25.6	19.1	~	~	~	~	13.9	13.1	43.0	31.8
Male Genital										
Penis	73.5	64.0	84.9	74.5	59.2	52.6	11.4	!	68.2	54.4
Penis - Squamous Cell Carcinoma	71.7	62.6	84.5	75.9	55.8	47.9	11.9	!	61.5	40.5
Scrotum	77.5	62.5	88.5	70.6	56.4	34.3	~	~	~	~
Scrotum - Squamous Cell Carcinoma	58.6	38.5	67.0	48.2	~	~	~	~	~	~
Scrotum - Paget Disease	89.0	82.3	98.3	86.3	~	~	~	~	~	~
Other and Not Otherwise Specified	51.8	34.1	~	~	~	~	~	~	~	~

* Excludes mesotheliomas and sarcomas
^ Excludes Basal, Squamous, Kaposi sarcoma & Melanoma
~ Statistic not displayed due to less than 25 cases.
! Not enough intervals to produce rate

Table 30.4 (continued): Cancers of Rare Sites: 5- & 10-Year (Yr) Relative Survival Rates (%) by Primary Site and SEER Summary Stage 1977 (2), 12 SEER Areas, 1988-2001

Primary Site	Summary Stage									
	Total		Local		Regional		Distant		Unstaged	
	Relative Survival Rate (%)		Relative Survival Rate (%)		Relative Survival Rate (%)		Relative Survival Rate (%)		Relative Survival Rate (%)	
	5-Yr	10-Yr	5-Yr	10-Yr	5-Yr	10-Yr	5-Yr	10-Yr	5-Yr	10-Yr
Urinary System										
Ureter	55.9	49.6	76.6	65.5	35.6	32.6	9.8	9.4	44.7	41.4
Ureter - Papillary Transitional Cell	57.8	51.4	77.3	66.5	36.3	33.6	10.1	9.7	50.4	46.3
Other Urinary	60.5	54.2	80.1	70.0	50.5	45.2	15.9	12.7	69.2	63.4
Other Urinary - Papillary Transitional Cell	63.8	60.0	77.6	70.7	52.8	43.3	4.8	!	73.9	70.2
Eye and Orbit	75.4	63.1	79.6	69.1	56.0	37.3	30.2	24.3	69.2	45.7
Eye and Orbit - Squamous Cell Carcinoma	86.8	77.3	89.2	82.2	64.3	!	~	~	88.5	57.8
Eye and Orbit - Melanoma	73.6	61.1	77.3	66.2	57.2	34.2	14.8	14.8	65.7	42.4
Other Endocrine	58.9	47.2	77.6	69.3	68.6	51.2	22.5	15.2	54.7	42.4
Thymus	66.3	51.3	88.8	82.1	70.5	48.6	35.4	24.3	61.6	51.6
Thymus - Thymoma	70.0	55.7	87.8	86.1	74.4	52.6	41.4	29.8	65.1	56.3
Adrenal Gland	38.7	29.6	65.5	52.6	45.7	39.2	8.6	3.4	31.4	19.4
Adrenal Gland - Adrenal Cortical	41.2	31.3	64.7	51.5	43.9	36.3	7.1	2.0	~	~
Adrenal Gland - Pheochromocytoma	64.8	44.0	83.0	60.5	~	~	~	~	~	~
Parathyroid Gland	93.1	81.6	95.6	85.8	95.5	82.2	~	~	~	~
Pituitary Gland	63.8	41.7	~	~	~	~	~	~	~	~
Pineal Gland	71.7	63.9	81.1	78.5	69.5	49.6	~	~	~	~
Mesothelioma	8.2	5.6	19.3	16.2	10.3	9.0	4.9	2.1	10.1	5.4
Mesothelioma - Pleura and Lung	6.4	4.3	15.2	11.9	9.1	7.8	3.7	1.4	6.6	3.3
Mesothelioma - Peritoneum and Retroperitoneum	18.4	9.5	~	~	24.8	14.3	13.1	5.3	26.6	16.9
Reticuloendothelial System Tumors	66.6	44.0	!	!	!	!	66.6	43.9	~	~
Waldenstrom's Macroglobulinemia	69.3	45.6	!	!	!	!	69.3	45.6	!	!
Myeloma	31.7	14.2	68.4	51.8	!	!	30.0	12.3	!	!
Solitary Myeloma	64.9	44.1	64.9	47.1	!	!	64.0	25.0	!	!
Multiple Myeloma	29.4	11.7	!	!	!	!	29.4	11.7	!	!
Unknown or Ill-defined Primary Site	11.3	9.8	~	~	!	!	46.7	41.6	11.2	9.7
Microscopically confirmed	12.4	10.6	~	~	!	!	47.2	47.2	12.3	10.6
Carcinomas	20.9	18.3	!	!	!	!	!	!	20.9	18.3
Adenocarcinomas	5.2	4.1	!	!	!	!	!	!	5.2	4.1
Other	21.8	18.5	~	~	!	!	47.2	47.2	21.4	18.0
Non-microscopically confirmed	6.6	5.4	!	!	!	!	~	~	6.6	5.4

* Excludes mesotheliomas and sarcomas
^ Excludes Basal, Squamous, Kaposi sarcoma & Melanoma
~ Statistic not displayed due to less than 25 cases.
! Not enough intervals to produce rate

relative survival than men with localized squamous cell carcinoma of the penis (67% vs. 85%, respectively).

Urinary System

Cancers of the ureter, urethra, urachus and other urinary organs were diagnosed most commonly as localized or regional disease and occurred more commonly among males. The majority of these cancers were papillary transitional cell carcinomas. Five-year relative survival rates from cancers of the ureter were similar among males vs. females, but for the other rare urinary sites, males had a distinct survival advantage at 5 years, 68% vs. 49%. The survival rates among whites were consistently higher than those for blacks. Five-year relative survival rates were uniformly poor for patients diagnosed with distant disease.

Eye and Orbit

Since children are excluded from this analysis, by definition, retinoblastomas are excluded as well. Among adults, nearly 80% of the malignancies of the eye were melanomas with most of the remainder being squamous cell carcinomas. Over 70% of all eye tumors were diagnosed while still localized, resulting in five-year relative survival rates of 87% for squamous cell carcinomas and 74% for melanomas. For melanomas of the eye 1, 3, and 5-year rates were quite similar in males and females while for squamous cell carcinomas of the eye, there appears to be a male survival advantage. There were very few cases among blacks, but for these few, survival was much poorer than among whites, especially at 5 and 10 years. Patients with squamous cell carcinoma of the eye had higher relative survival rates than patients with melanoma of the eye.

Endocrine System

Cancers of the thymus gland were primarily thymomas and patients tended to be diagnosed with regional disease. For thymomas, males and females survived similarly at 1, 3, and 5 years and the survival rates were similar among whites and blacks at 1, 3, 5, and 10 years. Sixty-five percent of adrenal gland malignancies were of the adrenal cortical type while another 10 percent were malignant pheochromocytomas. Relative 5-year survival rates were much better for the pheochromocytomas, 65% than for the adrenal cortical carcinomas, 41%. Males with pheochromocytomas had a survival advantage at 5 years compared to females, but at 10 years, the advantage was in favor of females, 59% vs. 30%. There were too few cases of pheochromocytomas among blacks to yield meaningful comparisons by race.

For adrenal cortical carcinomas, females had a survival advantage at 1, 3, 5, and 10 years. Malignant tumors of the parathyroid, pituitary, and pineal glands were indeed rare with too few cases to allow comparisons by race. The majority of these tumors were diagnosed at either the localized or regional stage with relative 5-year survival rates of 93%, 64%, and 72% respectively. For malignancies of the parathyroid and the pituitary, females survived better than males at 5 and 10 years whereas the opposite was true for pineal malignancies. For each stage, patients with adrenal gland cancers had much poorer survival than patients with other endocrine tumors.

Mesothelioma

Mesotheliomas arose more frequently in the pleura for both males (91%) and females (78%) with most of the remaining mesotheliomas arising from the (retro)peritoneum. The majority of mesotheliomas were diagnosed as distant disease. While uniformly dismal, the 5- and 10-year relative survival rates for females were more than triple those for males. Survival rates were slightly higher for blacks compared to whites. Interestingly for all races combined and both sexes combined, 5-year relative survival rates from mesotheliomas which arose in the (retro)peritoneum were at least double those which arose in the pleura. This was true even for patients diagnosed with regional or distant disease (Table 30.2).

Reticuloendothelial System

Survival from leukemias and multiple myeloma are discussed elsewhere. Ninety percent of the remaining tumors occurring in the reticuloendothelial system were classified as Waldenstrom's macroglobulinemia, a systemic disease always staged as distant at the time of diagnosis. Survival rates were similar among males and females (Table 30.2) and whites versus blacks (Table 30.3).

Myelomas

Multiple myelomas accounted for 93% of the myelomas. Patients with multiple myeloma survived much more poorly than patients with a solitary myeloma, especially at 5 (29% vs. 65%) and 10 (12% vs. 44%) years. For multiple myeloma survival was similar among males and females and blacks had slightly higher survival rate compared to whites. However, for solitary myeloma, males had a distinct survival advantage at 10 years, 53% vs. 29% for females.

Ill-defined and Unknown Sites

Some tumors are so disseminated at the time of diagnosis that it is impossible to determine the exact anatomic site in which the tumor arose. Sometimes these tumors can be ascribed to a body region such as the abdomen, but still, an exact primary site cannot be determined with certainty. For this group of tumors only, those without microscopic confirmation have been included but are shown separately in Tables 1-3. Twenty-two percent of these tumors did not have microscopic confirmation. By convention, these tumors are always classified with an unknown stage.

Not surprisingly, relative survival rates for this group as a whole were poor – 11% at five years. Patients whose disease was classified as "carcinoma" had a better survival than those with "adenocarcinoma" – 21% vs. 5% at five years. Patients with non-microscopically confirmed cancers experienced a five-year survival rate of only 7% vs. 12% for those with microscopic confirmation. Survival was essentially equal for males and for females, but whites had a slight survival advantage when compared to blacks, especially those with a tumor classified as carcinoma, 22% vs. 11% at five years.

DISCUSSION

It is not clear as to why there are certain anatomic sites in which cancer rarely arises. This is particularly curious in systems such as the endocrine system where with the exception of the thyroid gland few tumors arise, particularly in the pituitary and pineal glands. However, when cancer does arise in one of these rare sites, survival, in general, is similar to survival from other primary sites in the same system. For example survival from cancers of the pleura and from pleural mesotheliomas was very similar to survival from lung cancers.

Survival from cancer of the male breast, in general was similar to survival among females with breast cancer.

Among females, survival from cystadenocarcinomas of the (retro)peritoneum was very similar to survival from cystadenocarcinomas of the ovary. Roffers et al. documented that these tumors are actually extra ovarian tumors (9).

The extremely poor survival among patients with unknown or ill-defined primary site was probably reflective of the fact that the disease was already widely disseminated at the time of diagnosis so that the site of origin could not be identified. While this group was the largest of the rare site groups, overall unknown site accounts for less than 3% of all primary cancers.

Because of the small numbers of cases involved, detailed analysis of survival by races other than white cannot be made. Further, time trends in survival are also difficult due to the small numbers involved. As more survival data are accumulated from a larger group of registries for a longer time period, these analyses should be repeated.

REFERENCES

1. Ries LAG, Ward KC and Young JL Jr., Chapter 11: Sarcomas in Ries LAG, Young JL, Keel GE, Eisner MP, Lin YD, Horner, M-J (editors). SEER Survival Monograph: Cancer Survival Among Adults: U.S. SEER Program, 1988-2001, Patient and Tumor Characteristics. National Cancer Institute, SEER Program. NIH Pub. No. 07-6215, 2007.
2. Shambaugh, EM, Weiss, MA. Axtell, LM (eds), The 1977 Summary Staging Guide for the Cancer Surveillance, Epidemiology and End Results Reporting (SEER) Program, National Cancer Institute, Bethesda, MD, April 1977.
3. Piccirillo JF, Costas I, and Reichman ME, Chapte 2 Cancer of the Head and Neck in Ries LAG, Young JL, Keel GE, Eisner MP, Lin YD, Horner, M-J (editors). SEER Survival Monograph: Cancer Survival Among Adults: U.S. SEER Program, 1988-2001, Patient and Tumor Characteristics. National Cancer Institute, SEER Program. NIH Pub. No. 07-6215, 2007.
4. Ries LAG, and Eisner MP, Chapter 9: Cancer of the Lung in Ries LAG, Young JL, Keel GE, Eisner MP, Lin YD, Horner, M-J (editors). SEER Survival Monograph: Cancer Survival Among Adults: U.S. SEER Program, 1988-2001, Patient and Tumor Characteristics. National Cancer Institute, SEER Program. NIH Pub. No. 07-6215, 2007.
5. Ries, LAG and Eisner MP, Chapter13: Cancer of the Female Breast in Ries LAG, Young JL, Keel GE, Eisner MP, Lin YD, Horner, M-J (editors). SEER Survival Monograph: Cancer Survival Among Adults: U.S. SEER Program, 1988-2001, Patient and Tumor Characteristics. National Cancer Institute, SEER Program. NIH Pub. No. 07-6215, 2007.
6. Kosary CL, Chapter 16: Cancer of the Ovary in Ries LAG, Young JL, Keel GE, Eisner MP, Lin YD, Horner, M-J (editors). SEER Survival Monograph: Cancer Survival Among Adults: U.S. SEER Program, 1988-2001, Patient and Tumor Characteristics. National Cancer Institute, SEER Program. NIH Pub. No. 07-6215, 2007.
7. Kosary CL, Chapter 14: Cancer of the Cervix Uteri in Ries LAG, Young JL, Keel GE, Eisner MP, Lin YD, Horner, M-J (editors). SEER Survival Monograph: Cancer Survival Among Adults: U.S. SEER Program, 1988-2001, Patient and Tumor Characteristics. National Cancer Institute, SEER Program. NIH Pub. No. 07-6215, 2007.
8. Kosary CL, Chapter 15: Cancers of the Corpus Uteri in Ries LAG, Young JL, Keel GE, Eisner MP, Lin YD, Horner, M-J (editors). SEER Survival Monograph: Cancer Survival Among Adults: U.S. SEER Program, 1988-2001, Patient and Tumor Characteristics. National Cancer Institute, SEER Program. NIH Pub. No. 07-6215, 2007.
9. Roffers SD, Wu XC, Johnson CH, and Correa CN. Incidence of Extraovarian Primary Cancers in the United States, 1992-1997. Cancer; Vol. 15;97(10 Suppl):2643-7; May 2003.