

EYE, BRAIN AND NERVOUS SYSTEM

JAN ERICSSON, MAGNUS STENBECK

EYE

Malignant tumors of the eye are uncommon. The vast majority of these tumors are either retinoblastomas (occurring almost invariably in young children) or malignant melanomas (usually appearing in adults). The total number of newly diagnosed cancers of the eye in Sweden in 1991 was 100 (49 occurring in males and 51 in females), approximately 90% of these being malignant melanomas (1). The age-standardized incidence was 1.1 per 100 000 males and 1.0 per 100 000 females (1). Since 1971, the age-standardized incidence has decreased by on average 0.7% for males while there has been no significant change in the incidence for females. The incidence figures are approximately the same in the five Nordic countries (2).

The risk factors for development of cancers of the eye are poorly understood. Malignant melanomas of the eye do not seem to share risk factors with their counterparts in the skin. In retinoblastoma, a dominant autosomal hereditary trait has been found in almost half the cases, and is associated with chromosomal changes (3).

Cancer of the eye is diagnosed clinically by ophthalmological inspection and investigation. The diagnosis is not always confirmed by histopathological examination since—with increasing frequency during later years—non-invasive methods of treatment (radiotherapeutic and other) have been applied.

Material

For the period 1961–1989, a total of 1 441 men and 1 311 women were reported to the Swedish Cancer Registry with a diagnosis of malignant tumor of the eye. 44 cases were excluded from the analysis (see Methods and Material) leaving 1 424 men and 1 284 women in the cohort. The mean age at diagnosis was 56 years in men and 55 in women. Over the whole period 42% of men and women were 65 years or older at diagnosis.

Results

In general, crude (CS) and relative (RS) survival rates after diagnosis of eye cancer are good. Due to the small number of cases in the different cohorts, wide variations in CS and RS were noted. Relative 1, 2 and 5-year figures (RS) were 93%, 81% and 67%, respectively for males diagnosed in 1985–87, and 94%, 87% and 66%, for females (Table 1). No great changes were observed across the period. The observed fluctuations are well within the error margin.

Discussion

Treatment of retinoblastomas during the last 10–20 years in Sweden is known to have been extremely successful, only an occasional case having a fatal outcome. However, since less than 10% of all cancers of the eye are retinoblastomas (occurring in children), the impact of the evidently greatly improved survival for patients with this type of eye tumor on total survival in eye cancer is small (Kock E, personal communication). Malignant melanomas account for approximately 90% of the malignant tumors of the eye. The figures on survival obtained in the present study can, therefore, be regarded as largely mirroring the prognosis for patients with this malignancy.

The results do not show any great improvement in survival over the observed period. During the period, the mode of treatment of malignant melanomas of the eye has changed towards non-invasive methods. As a rule, symptoms of altered vision and/or pain lead the patient with a malignant tumor of the eye to examination by an ophthalmologist. Hence, it appears unlikely that tumors of the eye have been detected at earlier stages during later years than in the 1960's. A possible conclusion based on the results is that there is no evidence that the change towards more conservative treatment has led to poorer (or better) survival for patients with malignant melanomas of the eye.

There are few previous population based studies on survival of patients with cancer of the eye. The closest one

Table 1
 Eye
 Percentage crude (CS) and relative (RS) survival

Sex	Age group	Years since diagnosis	Period of diagnosis													
			1961-63		1965-67		1970-72		1975-77		1980-82		1985-87		1987-89	
			CS	RS	CS	RS	CS	RS	CS	RS	CS	RS	CS	RS	CS	RS
Males	0-89	1	89.0	91.3	90.1	93.0	89.4	92.4	94.3	97.2	91.4	94.6	89.5	92.5	92.3	95.4
		2	76.8	80.8	79.6	84.7	80.8	86.4	82.3	87.4	80.7	86.4	76.0	81.2	83.1	88.8
		5	52.9	60.6	54.2	64.0	58.9	70.4	56.3	66.2	56.4	67.4	56.5	67.2	61.5	71.0
		10	43.8	59.5	38.0	54.7	38.4	56.4	39.2	55.8	37.5	54.7
		15	36.6	61.0	28.2	50.9	32.5	59.3	28.8	51.0
		20	28.1	59.1	21.1	49.1	25.6	59.0
	0-54	1	90.5	90.8	95.2	95.7	94.1	94.5	96.0	96.4	92.5	92.8	97.8	98.1	97.8	98.1
		2	85.7	86.3	90.5	91.3	88.2	88.8	88.0	88.7	92.5	93.0	93.5	94.0	93.4	93.8
		5	69.8	71.3	71.4	73.1	80.4	81.9	70.0	71.4	77.5	78.6	87.0	88.1	73.5	74.4
		10	60.2	63.5	66.7	70.5	70.6	73.9	56.0	58.7	65.4	67.7
		15	53.7	59.6	54.8	61.1	66.7	72.6	54.0	58.9
		20	45.6	54.7	47.6	57.4	60.8	70.0
	55-74	1	88.0	90.3	90.2	92.5	88.3	90.8	94.5	97.3	92.8	95.5	86.7	88.9	93.0	95.5
		2	74.7	78.8	76.8	81.0	81.8	86.7	79.1	83.9	79.5	84.4	73.3	77.3	83.1	87.9
		5	46.7	54.4	53.7	62.4	55.8	66.1	52.8	62.4	55.4	65.5	44.3	51.4	60.0	68.0
		10	37.3	55.2	29.3	42.8	27.3	41.8	34.1	51.6	30.6	46.3
		15	29.3	62.2	20.7	42.1	19.5	43.1	19.1	41.2
		20	20.0	70.6	12.2	38.3	9.9	36.3
	75-89	1	88.2	97.9	77.8	88.0	82.6	93.2	88.2	99.4	82.3	94.9	81.5	90.6	80.8	89.8
		2	52.9	65.8	66.7	86.2	60.9	78.0	82.3	104.9	58.8	78.5	51.9	64.6	65.4	81.0
		5	17.6	32.7	16.7	34.7	21.7	44.4	35.3	69.3	11.8	26.3	31.1	58.0	44.0	73.0
		10	11.8	55.5	11.1	66.6	4.3	25.3	17.6	86.6	5.9	39.5
		15	5.9	116.1	0.0	0.0	0.0	0.0	5.9	103.8
		20	0.0	0.0	0.0	0.0	0.0	0.0
Females	0-89	1	88.7	90.6	92.1	94.0	92.1	94.1	88.8	90.7	97.5	99.0	91.8	93.7	93.6	96.1
		2	79.0	82.4	87.0	90.8	87.0	90.9	83.6	87.1	93.3	96.3	83.6	87.2	83.3	87.9
		5	63.2	70.5	69.8	78.0	61.1	68.7	61.2	68.1	74.8	81.7	59.0	66.2	63.5	73.1
		10	48.1	61.1	54.7	69.7	49.6	63.9	50.0	63.0	54.3	66.5
		15	36.8	54.2	44.6	66.6	38.1	57.1	42.4	61.9
		20	24.5	42.7	36.7	65.8	29.2	51.9
	0-54	1	93.0	93.2	96.2	96.4	92.6	92.9	96.0	96.3	100.0	100.2	93.3	93.5	95.5	95.7
		2	87.7	88.1	92.3	92.7	92.6	93.1	96.0	96.5	100.0	100.4	91.1	91.5	93.2	93.5
		5	75.4	76.4	86.5	87.5	75.9	76.8	82.0	83.0	93.8	94.7	76.3	77.0	90.5	91.3
		10	64.9	67.0	82.7	85.0	68.5	70.3	72.0	73.9	76.7	78.4
		15	59.6	63.1	75.0	78.9	57.4	60.2	68.0	71.3
		20	48.7	53.8	71.2	77.5	51.1	55.3
	55-74	1	90.3	92.1	94.3	95.9	98.5	100.1	91.2	92.6	98.1	99.4	95.2	96.5	92.9	94.1
		2	82.3	85.6	87.1	90.2	90.8	94.0	80.9	83.4	94.3	96.9	82.3	84.7	78.6	80.7
		5	62.9	70.6	64.3	71.2	60.0	66.4	55.9	61.1	73.6	79.5	63.5	69.1	54.5	58.9
		10	40.3	54.1	45.7	59.3	47.7	61.9	44.1	55.6	46.8	57.0
		15	24.2	42.3	32.9	54.4	33.9	55.6	33.6	53.1
		20	8.1	20.4	20.0	47.4	20.0	46.5
	75-89	1	64.3	71.7	70.6	77.6	70.0	76.5	56.3	62.6	88.9	94.7	81.5	87.2	92.9	100.2
		2	28.6	35.7	70.6	86.3	60.0	72.3	56.3	69.8	72.2	82.3	74.1	85.4	81.0	94.6
		5	14.3	26.3	41.2	72.1	25.0	42.6	18.8	34.2	27.8	40.4	18.9	28.4	50.2	77.9
		10	14.3	61.6	5.9	22.1	5.0	18.8	6.3	26.5	16.7	44.6
		15	0.0	0.0	0.0	0.0	0.0	0.0	0.0	0.0
		20	0.0	0.0	0.0	0.0	0.0	0.0

appears to be the Danish report covering the years 1943-1987 (3). In this study, relative one-year survival for men increased from 88% to 93%, and five year survival from 60% to 68%, and for women from 90% to 96% and 61% to 74%, respectively. The figures covering cases diagnosed during the latest years in that study compare well with figures in the present report.

INTRACRANIAL TUMORS

Tumors located within the skull cavity are fairly common. In Sweden they are among the 12-14 most commonly occurring neoplasias to be reported to the Cancer Registry. The true incidence has been said to be difficult to estimate due to incompleteness and inconsequence in the

a) Males

b) Females

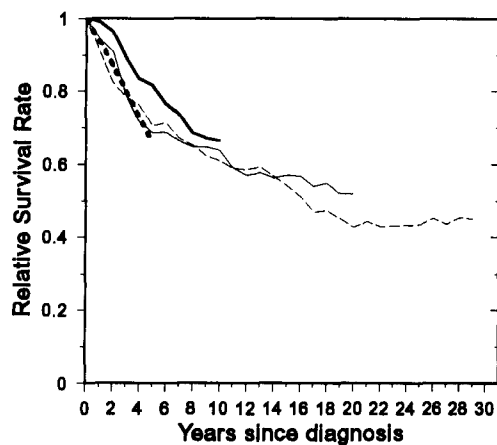
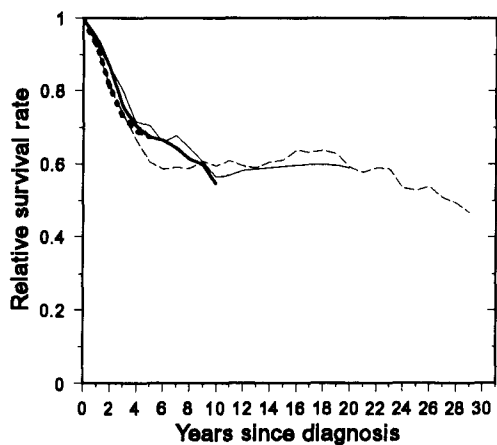


Fig. 1. Eye. Relative survival of a) males and b) females diagnosed in 1961-63 (-----), 1970-72 (—), 1980-82 (—) and 1985-87 (●●●), 0-89 years of age. 3-year moving averages.

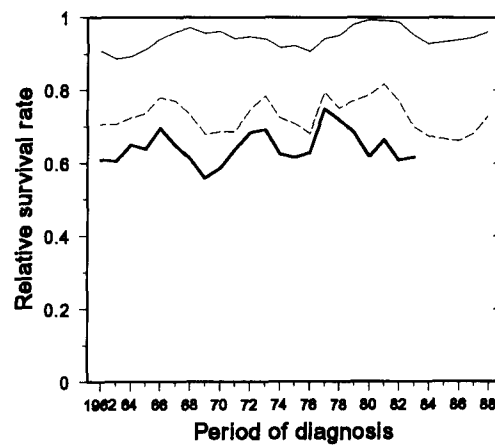
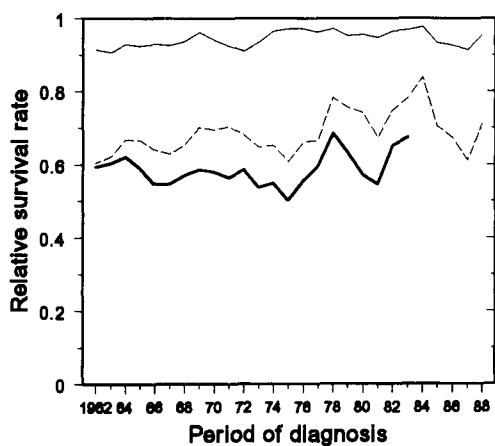


Fig. 2. Eye. 1-year (—), 5-year (-----) and 10-year (—) relative survival rates of a) males and b) females, 0-89 years of age. 3-year moving averages.

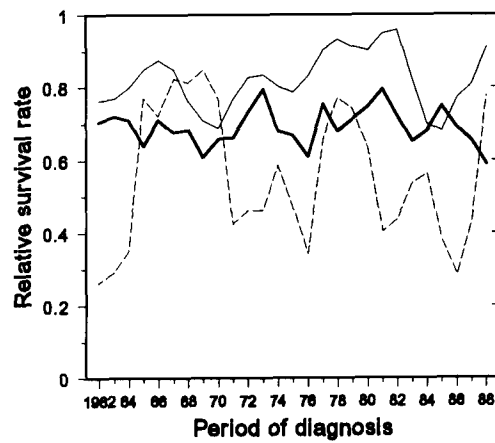
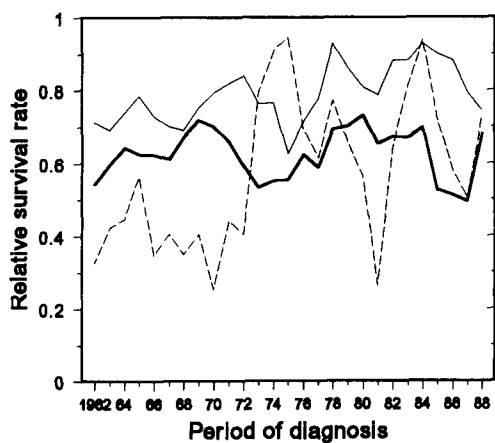


Fig. 3. Eye. Age-group specific 5-year relative survival of a) males and b) females, 0-54 (—), 55-74 (—) and 75-89 (-----) years of age. 3-year moving averages.

Table 2
Brain, excluding neurinoma and meningioma
Percentage crude (CS) and relative (RS) survival

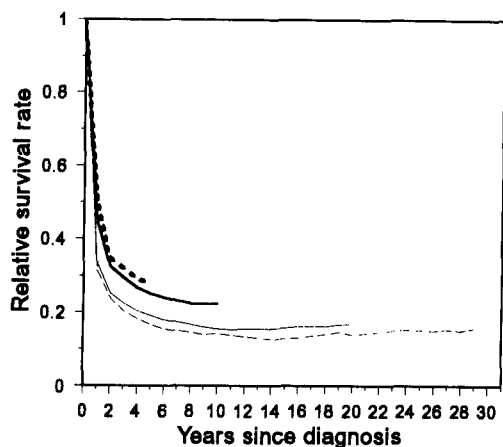
Sex	Age group	Years since diagnosis	Period of diagnosis													
			1961-63		1965-67		1970-72		1975-77		1980-82		1985-87		1987-89	
			CS	RS	CS	RS	CS	RS	CS	RS	CS	RS	CS	RS	CS	RS
Males	0-89	1	30.9	31.4	35.0	35.5	32.8	33.4	35.2	36.0	44.2	45.1	49.6	50.7	47.0	48.0
		2	23.2	23.9	24.4	25.3	24.4	25.4	23.8	24.9	31.3	32.6	33.3	34.8	33.6	34.9
		5	15.4	16.8	17.9	19.6	17.4	19.2	17.2	19.4	22.5	25.1	24.8	27.8	24.9	27.1
		10	11.6	14.2	13.6	16.8	12.3	15.5	12.6	16.4	17.7	22.5
		15	8.9	12.8	12.3	17.6	10.8	15.9	10.4	15.9
		20	8.2	13.9	10.5	18.0	9.7	16.9
	0-54	1	47.2	47.3	50.9	51.0	51.6	51.8	59.5	59.7	66.4	66.6	74.3	74.4	74.7	74.8
		2	36.4	36.7	38.4	38.7	41.4	41.7	45.6	45.9	54.3	54.6	56.3	56.6	58.8	59.0
		5	24.0	24.4	28.7	29.2	30.2	30.7	33.7	34.3	41.4	42.0	45.7	46.3	45.8	46.3
		10	18.8	19.8	22.4	23.4	23.3	24.3	25.5	26.7	34.5	35.9
		15	14.7	16.1	21.2	23.2	21.0	22.8	22.1	23.9
		20	14.4	17.1	19.0	22.1	19.3	22.0
	55-74	1	12.7	13.1	19.0	19.4	16.0	16.4	17.2	17.7	28.5	29.3	32.5	33.3	25.0	25.6
		2	8.1	8.5	9.6	10.1	9.0	9.5	6.5	6.9	13.9	14.6	15.5	16.3	12.2	12.8
		5	5.9	6.8	6.7	7.7	5.7	6.6	4.1	4.8	8.1	9.4	7.8	9.0	6.9	7.7
		10	3.4	4.9	4.3	6.2	2.2	3.2	2.2	3.2	4.5	6.4
		15	2.5	5.0	2.7	5.4	1.3	2.6	1.0	1.9
		20	0.9	3.0	1.3	4.3	0.7	2.1
	75-89	1	3.5	3.9	2.7	3.0	7.4	8.2	5.5	6.1	14.5	15.8	12.4	13.6	5.1	5.6
		2	3.5	4.3	2.7	3.5	3.7	4.6	2.7	3.4	7.9	9.5	5.8	7.0	2.0	2.5
		5	0.0	0.0	0.0	0.0	1.9	3.5	1.4	2.6	4.0	6.6	3.3	5.6	0.0	0.0
		10	0.0	0.0	0.0	0.0	0.0	0.0	1.4	6.1	2.6	9.7
		15	0.0	0.0	0.0	0.0	0.0	0.0	0.0	0.0
		20	0.0	0.0	0.0	0.0	0.0	0.0
Females	0-89	1	32.1	32.4	33.6	34.0	37.6	38.2	37.8	38.3	51.7	52.3	47.9	48.6	52.5	53.2
		2	23.5	24.0	24.9	25.6	29.2	30.1	26.3	27.0	37.4	38.3	36.6	37.6	41.8	42.9
		5	15.6	16.6	19.5	21.1	20.8	22.4	19.7	21.2	29.8	31.9	28.3	30.6	33.1	35.5
		10	12.0	13.8	16.1	19.1	15.3	18.2	15.8	18.8	23.1	27.0
		15	10.3	13.0	14.7	19.5	12.3	16.4	13.1	17.6
		20	9.3	13.3	13.4	20.2	11.2	17.4
	0-54	1	46.5	46.5	49.5	49.5	55.8	55.9	62.8	62.9	75.6	75.7	75.2	75.3	79.4	79.5
		2	36.1	36.3	39.2	39.4	44.3	44.4	46.5	46.6	59.5	59.6	65.6	65.8	70.3	70.4
		5	24.2	24.5	32.3	32.6	31.9	32.2	35.3	35.7	48.7	49.1	55.4	55.8	59.4	59.7
		10	18.7	19.2	27.5	28.2	24.5	25.1	28.8	29.4	39.9	40.6
		15	17.1	18.0	25.6	26.7	21.9	22.9	25.1	26.1
		20	15.8	17.3	24.1	26.0	20.8	22.3
	55-74	1	16.0	16.2	20.2	20.5	24.2	24.5	19.6	19.9	32.9	33.3	29.6	30.0	30.4	30.8
		2	9.6	9.9	11.5	11.9	17.4	17.9	10.6	10.9	19.6	20.2	16.3	16.8	15.9	16.3
		5	6.0	6.5	7.3	8.0	12.1	13.2	7.7	8.4	15.5	16.8	9.5	10.3	9.9	10.7
		10	4.8	6.0	5.2	6.6	7.9	9.8	6.4	7.9	10.1	12.3
		15	2.8	4.3	4.2	6.5	4.8	7.4	4.2	6.3
		20	2.0	4.1	2.8	6.0	3.6	7.7
	75-89	1	13.0	14.3	3.2	3.5	10.0	10.9	9.5	10.3	24.3	25.7	24.1	25.6	17.6	18.8
		2	4.3	5.3	3.2	3.8	10.0	11.9	9.5	11.2	14.3	16.1	14.3	16.2	13.0	14.8
		5	4.3	7.3	1.6	2.6	6.7	10.9	6.3	10.0	5.7	8.1	7.5	10.8	7.9	11.5
		10	0.0	0.0	0.0	0.0	5.0	16.9	1.6	5.0	2.9	7.2
		15	0.0	0.0	0.0	0.0	0.0	0.0	1.6	14.4
		20	0.0	0.0	0.0	0.0	0.0	0.0

reporting systems (4). Furthermore, a large number of different types of tumors occur in the skull cavity, some histologically benign and some malignant, and the rules for reporting and classification vary between cancer registries. In Sweden, all true neoplasias located in the skull cavity are to be reported and registered; however, tumors emanating from the tissues of the pituitary gland are referred to the endocrine glands. Craniopharyngeomas are counted as

located intracranially. The rules for coding intracranial tumors in the Swedish Cancer Registry have remained unaltered since the registry was started. The number of cases lacking histological confirmation is, and has been, small, being some 10% of all incident cases (1).

In Sweden, intracranial tumors constitute about 2.6% of all tumors in males reported to the Cancer Registry; the corresponding figure for females is 2.7%. The majority of

a) Males



b) Females

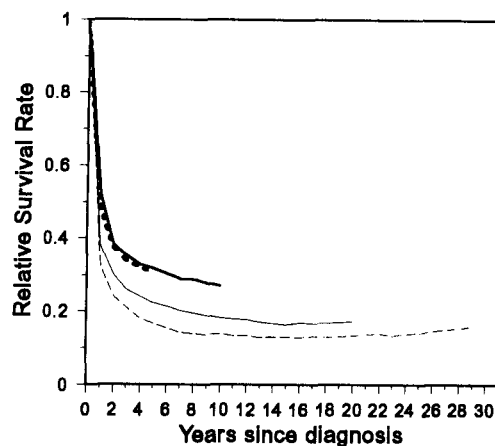


Fig. 4. Brain excised meningioma and neuroma. Relative survival of a) males and b) females diagnosed in 1961-63 (-----), 1970-72 (—), 1980-82 (—) and 1985-87 (••••), 0-89 years of age. 3-year moving averages.

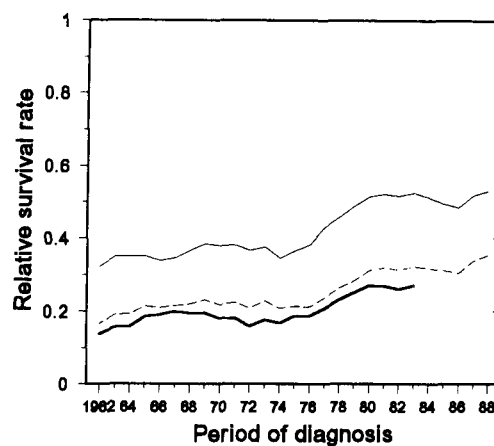
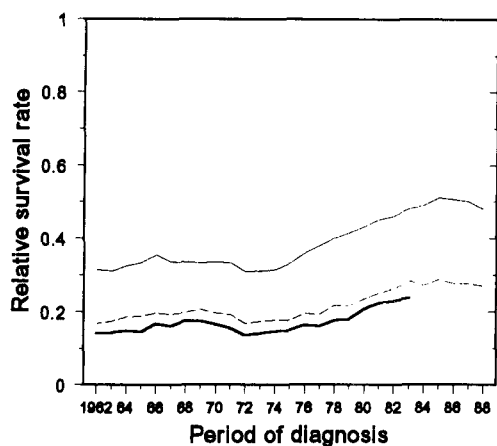


Fig. 5. Brain excised meningioma and neuroma. 1-year (—), 5-year (-----) and 10-year (—) relative survival rates of a) males and b) females, 0-89 years of age. 3-year moving averages.

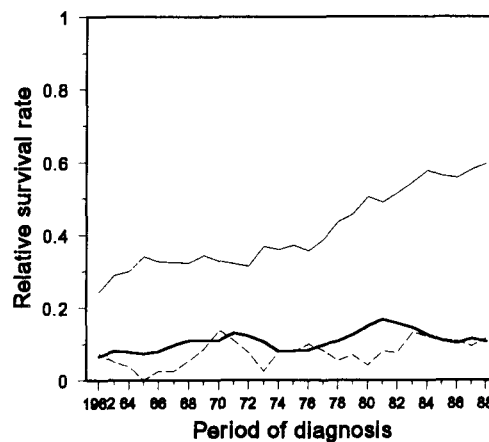
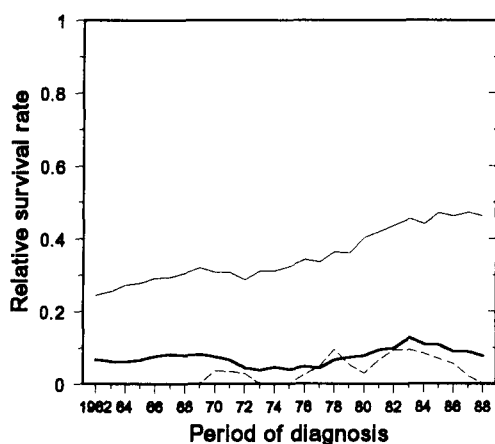


Fig. 6. Brain excised meningioma and neuroma. Age-group specific 5-year relative survival of a) males and b) females, 0-54 (—), 55-74 (—) and 75-89 (-----) years of age. 3-year moving averages.

Table 3
Meningioma
Percentage crude (CS) and relative (RS) survival

Sex	Age group	Years since diagnosis	Period of diagnosis													
			1961-63		1965-67		1970-72		1975-77		1980-82		1985-87		1987-89	
			CS	RS	CS	RS	CS	RS	CS	RS	CS	RS	CS	RS	CS	RS
Males	0-89	1	47.9	50.1	50.0	52.2	46.8	49.3	56.8	59.5	87.6	89.8	86.2	88.5	87.0	89.3
		2	45.8	50.1	48.0	52.4	44.8	49.8	52.4	57.4	82.2	86.5	81.9	86.5	83.1	87.7
		5	38.7	48.4	40.3	50.7	40.4	52.8	43.6	55.1	71.9	82.5	71.1	82.1	74.2	83.6
		10	31.0	47.9	34.7	56.1	34.8	59.6	35.7	57.1	53.4	72.7
		15	23.9	46.3	27.6	59.1	28.0	62.6	28.4	58.2
		20	19.0	47.0	23.0	67.1	21.9	64.3
	0-54	1	80.8	81.1	92.0	92.4	84.7	85.1	88.7	89.1	96.8	97.1	100.0	100.3	98.4	98.7
		2	78.8	79.5	92.0	92.9	83.3	84.1	82.3	83.0	95.2	95.9	95.2	95.9	95.2	95.8
		5	67.3	69.0	84.0	86.3	80.6	82.7	77.4	79.4	87.1	88.9	93.7	95.4	95.2	96.6
		10	59.6	63.6	82.0	88.2	75.0	80.3	67.5	71.9	80.3	84.4
		15	53.9	61.5	70.0	80.9	70.8	80.7	59.3	66.6
		20	44.2	56.0	64.0	82.3	65.3	81.2
	55-74	1	41.7	42.8	49.5	50.9	50.5	52.0	63.1	64.8	87.2	89.6	86.3	88.6	88.9	91.2
		2	38.3	40.6	45.7	48.5	47.7	50.8	58.6	61.9	81.7	86.4	82.3	87.0	85.5	90.0
		5	31.7	37.4	35.2	41.7	39.5	47.3	45.0	52.7	70.6	82.8	65.4	76.2	71.8	80.6
		10	21.7	32.9	25.7	39.6	30.3	48.1	35.1	51.7	45.2	66.9
		15	10.0	22.0	18.1	40.7	17.4	41.1	25.8	52.7
		20	6.7	24.4	12.4	46.4	7.1	29.1
	75-89	1	3.3	3.9	0.0	0.0	1.4	1.7	8.9	10.2	50.0	55.7	47.8	52.5	51.9	56.8
		2	3.3	4.7	0.0	0.0	0.0	0.0	7.1	9.4	28.6	35.7	43.5	52.8	44.4	53.5
		5	3.3	8.2	0.0	0.0	0.0	0.0	3.6	7.7	14.3	26.5	34.2	59.3	36.4	56.2
		10	0.0	0.0	0.0	0.0	0.0	0.0	1.8	10.5	0.0	0.0
		15	0.0	0.0	0.0	0.0	0.0	0.0	0.0	0.0
		20	0.0	0.0	0.0	0.0	0.0	0.0
Females	0-89	1	46.2	47.8	50.1	52.2	48.8	50.6	61.1	62.9	90.6	91.7	89.8	91.0	89.8	91.0
		2	45.3	48.6	47.8	51.9	46.6	50.3	58.9	62.4	88.0	90.3	88.1	90.6	87.9	90.3
		5	40.4	48.4	42.8	52.8	43.3	52.8	53.0	61.8	80.0	85.9	81.0	87.7	84.1	90.6
		10	34.0	49.7	36.9	56.1	37.5	56.9	44.8	62.3	67.3	79.9
		15	29.1	52.6	30.8	57.9	32.5	61.5	37.9	64.2
		20	25.5	57.8	26.1	61.3	26.9	63.7
	0-54	1	80.4	80.7	88.1	88.3	84.3	84.5	94.9	95.1	96.0	96.2	98.2	98.4	97.7	97.9
		2	78.3	78.7	85.3	85.8	82.1	82.5	93.2	93.7	93.6	94.0	97.6	98.0	97.7	98.0
		5	70.7	71.8	82.6	83.9	79.1	80.2	89.0	90.2	91.2	92.3	95.1	96.1	94.4	95.4
		10	62.0	64.5	75.2	78.3	75.4	77.9	83.9	86.6	87.2	89.7
		15	59.8	64.4	66.9	72.2	70.2	74.6	81.3	86.3
		20	56.4	64.2	61.4	69.8	67.8	75.2
	55-74	1	47.1	48.0	58.3	59.2	58.1	59.0	67.4	68.4	89.2	90.4	90.5	91.6	91.0	92.2
		2	47.1	49.0	56.1	58.0	55.2	57.1	63.7	65.7	86.7	89.0	88.9	91.3	87.9	90.2
		5	42.0	47.0	47.6	52.4	51.0	56.2	54.2	59.3	79.0	85.0	79.4	85.6	83.8	90.3
		10	34.4	45.7	39.0	50.0	41.0	52.7	41.6	52.3	63.4	76.8
		15	26.1	44.9	31.0	49.9	33.3	54.4	29.4	46.1
		20	20.4	50.6	23.5	52.8	21.3	49.7
	75-89	1	5.0	5.6	6.2	7.0	6.9	7.6	10.7	11.7	77.4	81.5	62.1	65.6	56.9	60.2
		2	3.8	4.7	3.9	4.9	5.6	6.9	10.7	12.8	74.2	82.7	56.9	63.9	54.9	61.7
		5	2.5	4.6	2.3	4.5	3.1	5.5	9.7	16.0	41.9	57.6	47.0	65.9	50.4	70.8
		10	1.3	5.3	1.6	6.9	1.3	5.1	5.8	19.9	12.3	28.9
		15	0.0	0.0	0.0	0.0	0.0	0.0	3.9	40.2
		20	0.0	0.0	0.0	0.0	0.0	0.0

these neoplasias are intracerebral, mainly astrocytomas and gliomas of variable histological malignancy grades in adults, and medulloblastomas in children; however, meningiomas and acoustic neurinomas are of relatively frequent occurrence.

The total number of intracranial tumors reported to the Swedish Cancer Registry in 1991 was 1 063 (543 in men and 520 in women). The age-standardized incidence was 12.8

per 100 000 males and 11.0 per 100 000 females. Since 1972, the age-standardized incidence has increased annually by 1.4% for males and 1.1% for females. For the small subgroup of neurinomas there has been a much sharper change with an annual increase of 6.4% for males and 4.1% for females. The incidence of intracranial tumors in Sweden is higher than in the other Nordic countries, but the differences are small (2).

a) Males

b) Females

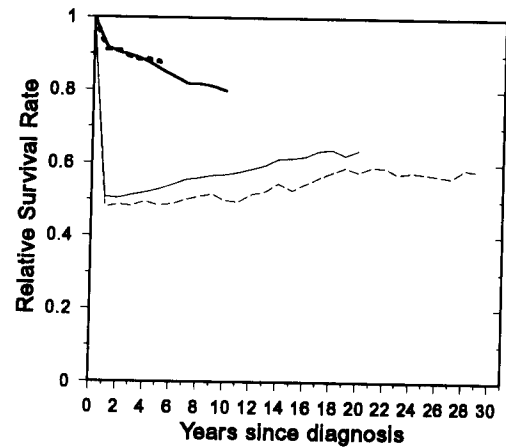
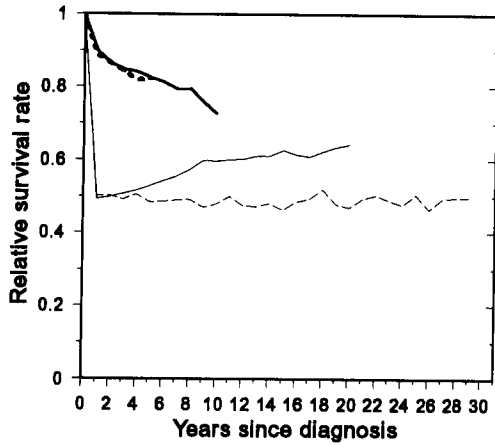


Fig. 7. Meningioma. Relative survival of a) males and b) females diagnosed in 1961-63 (-----), 1970-72 (—), 1980-82 (—) and 1985-87 (••••), 0-89 years of age. 3-year moving averages.

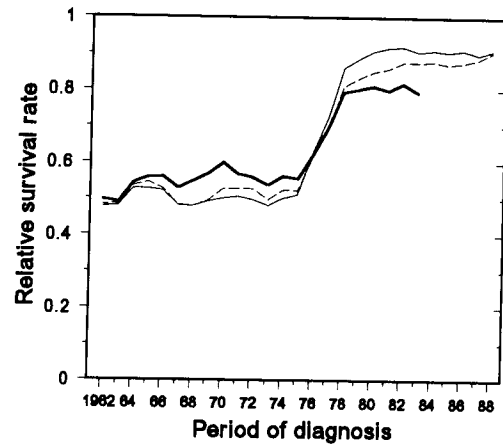
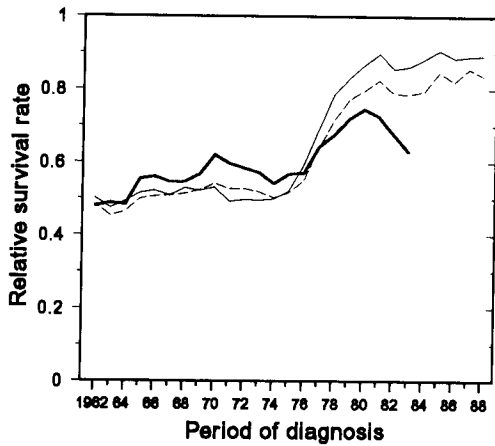


Fig. 8. Meningioma. 1-year (—), 5-year (-----) and 10-year (—) relative survival rates of a) males and b) females, 0-89 years of age. 3-year moving averages.

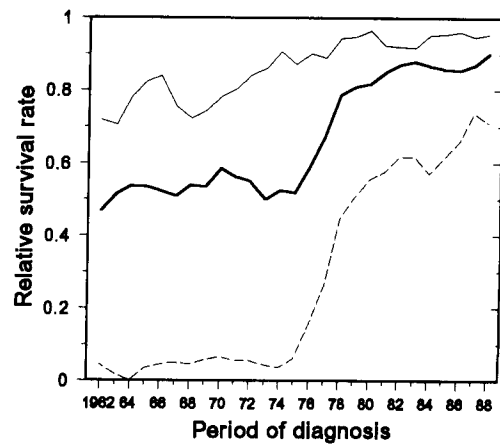
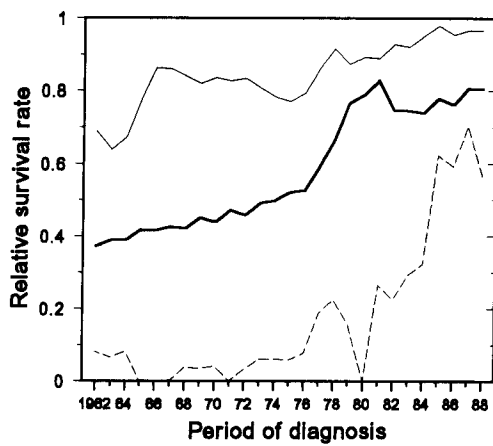


Fig. 9. Meningioma. Age-group specific 5-year relative survival of a) males and b) females, 0-54 (—), 55-74 (—) and 75-89 (-----) years of age. 3-year moving averages.

Table 4
Neurinoma of intracranial nerves
Percentage crude (CS) and relative (RS) survival

Sex	Age group	Years since diagnosis	Period of diagnosis													
			1961-63		1965-67		1970-72		1975-77		1980-82		1985-87		1987-89	
			CS	RS	CS	RS	CS	RS	CS	RS	CS	RS	CS	RS	CS	RS
Males	0-89	1	70.6	71.5	67.4	68.5	67.3	68.6	76.3	77.7	87.5	88.8	96.2	97.0	96.0	97.2
		2	67.7	69.4	67.4	69.8	65.3	67.8	76.3	79.3	85.7	88.4	95.2	97.0	93.9	96.3
		5	67.7	72.8	54.4	59.8	63.3	69.8	73.7	81.7	85.7	93.1	91.0	95.8	85.3	90.1
		10	64.7	77.1	50.0	62.4	61.2	76.0	68.4	86.6	82.1	99.3
		15	52.9	72.3	45.6	66.8	55.1	78.5	65.8	98.0
		20	50.0	80.5	41.3	72.8	50.8	84.3
	0-54	1	86.4	86.7	84.0	84.3	84.0	84.2	88.2	88.5	92.3	92.6	98.5	98.7	98.1	98.3
		2	86.4	87.0	84.0	84.6	84.0	84.5	88.2	88.8	92.3	92.8	98.5	99.0	98.1	98.6
		5	86.4	88.1	76.0	77.7	84.0	85.4	88.2	89.9	92.3	93.8	98.5	99.9	98.1	99.2
		10	81.8	86.3	72.0	76.3	80.0	83.3	88.2	92.2	92.3	96.0
		15	77.3	86.0	68.0	76.2	76.0	82.0	88.2	95.6
		20	72.7	88.1	64.0	78.0	76.0	86.3
	55-74	1	41.7	42.9	47.4	48.6	57.1	58.5	73.7	75.4	85.7	87.5	94.7	96.6	95.3	97.4
		2	33.3	35.5	47.4	49.9	52.4	55.0	73.7	77.3	85.7	89.4	92.1	95.8	93.0	97.1
		5	33.3	39.8	31.6	36.7	47.6	54.7	68.4	78.7	85.7	96.4	80.8	90.4	68.2	75.1
		10	33.3	52.2	26.3	38.8	47.6	67.6	57.9	82.4	78.6	105.0
		15	8.3	19.4	21.1	44.2	38.1	72.4	52.6	102.9
		20	8.3	33.8	15.8	53.8	26.7	75.3
	75-89	1														
		2														
		5														
		10														
		15														
		20														
Females	0-89	1	76.6	77.9	78.7	79.9	67.5	68.4	73.8	74.6	96.8	97.7	98.5	99.1	97.4	98.2
		2	75.3	77.8	73.8	76.1	66.2	68.0	72.1	73.8	95.8	97.5	97.7	99.0	97.4	98.9
		5	71.4	78.0	70.5	76.8	63.6	68.5	67.2	71.4	91.6	96.3	93.8	97.4	97.4	101.8
		10	68.8	83.9	65.6	79.1	58.4	69.2	63.8	73.5	84.9	96.3
		15	61.0	84.7	63.9	87.0	58.4	78.5	57.1	72.8
		20	52.0	84.2	54.1	84.7	55.8	88.2
	0-54	1	93.5	93.7	100.0	100.2	93.8	94.0	78.8	79.0	100.0	100.2	98.4	98.6	98.0	98.1
		2	93.5	94.0	92.6	93.0	93.8	94.2	78.8	79.2	100.0	100.4	98.4	98.7	98.0	98.3
		5	93.5	94.7	88.9	89.9	93.8	94.9	75.7	76.7	97.4	98.4	93.4	94.2	98.0	98.8
		10	90.3	93.0	85.2	87.6	90.6	93.4	75.7	78.1	94.7	97.2
		15	83.9	88.6	85.2	89.8	90.6	95.8	72.5	77.0
		20	83.9	92.1	81.5	89.0	87.5	96.0
	55-74	1	71.0	72.0	75.0	76.0	53.7	54.4	75.0	76.0	94.6	95.8	98.5	99.5	96.8	97.9
		2	71.0	73.1	71.4	73.5	51.2	52.8	70.8	72.8	92.9	95.3	97.0	99.1	96.8	98.9
		5	65.8	71.5	67.9	73.8	46.3	50.6	66.7	72.1	87.5	94.1	94.0	100.0	96.8	103.1
		10	63.2	77.9	60.7	75.0	39.0	48.8	58.3	71.3	77.9	94.0
		15	52.6	78.7	57.1	85.5	39.0	60.0	45.8	67.7
		20	34.2	68.0	39.3	77.8	36.6	76.8
	75-89	1														
		2														
		5														
		10														
		15														
		20														

For obvious reasons, the etiology of intracranial tumors varies with the type of tumor and tissue of origin (4). Suggested risk factors for intracerebral tumors include genetic predisposition, occupation (e.g. dental personnel exposed to amalgam), and exposure to viruses, chemicals, lead, trauma and x-rays (1, 3). Von Recklinghausen's disease often shows an autosomal dominant pattern and is associated both with tumors of the eighth cranial nerve

(acoustic neurinomas) and, rarely, with meningiomas and ependymomas. Otherwise, theories about the etiology of meningioma and neurinoma are few.

The choice of therapy for intracranial tumors depends on the size, precise location, histological type, and the grade of malignancy of the tumor. In general, surgical intervention has been the method of choice for treatment of extracerebral tumors like meningiomas and neurinomas,

a) Males

b) Females

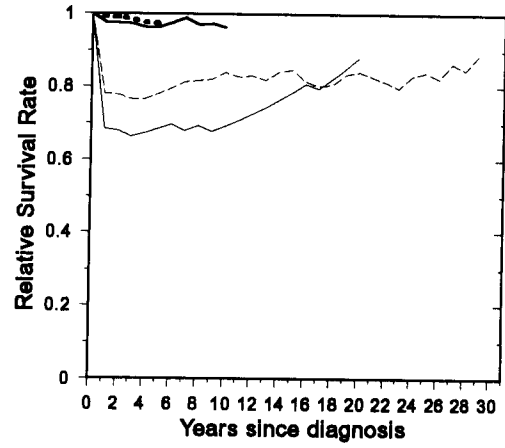
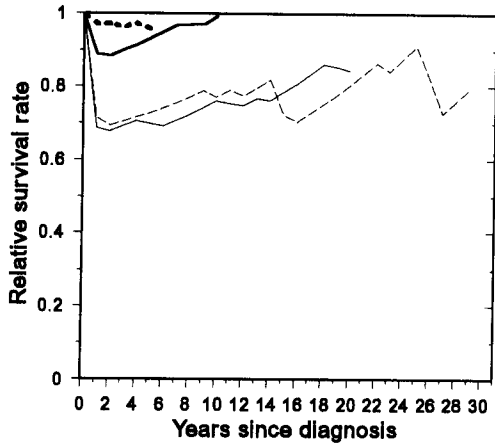


Fig. 10. Intracranial nerves. Relative survival of a) males and b) females diagnosed in 1961–63 (-----), 1970–72 (—), 1980–82 (—) and 1985–87 (••••), 0–89 years of age. 3-year moving averages.

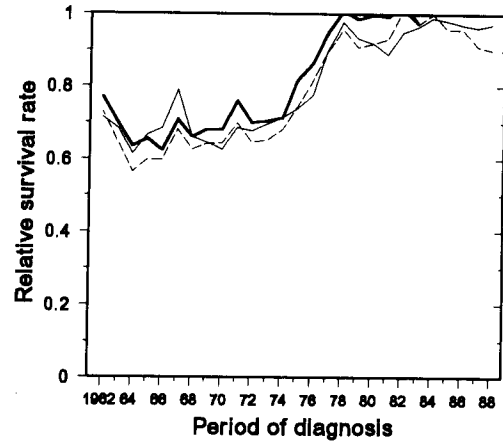
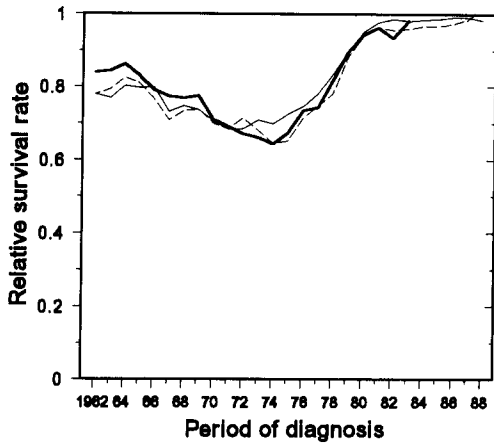


Fig. 11. Intracranial nerves. 1-year (—), 5-year (-----) and 10-year (—) relative survival rates of a) males and b) females, 0–89 years of age. 3-year moving averages.

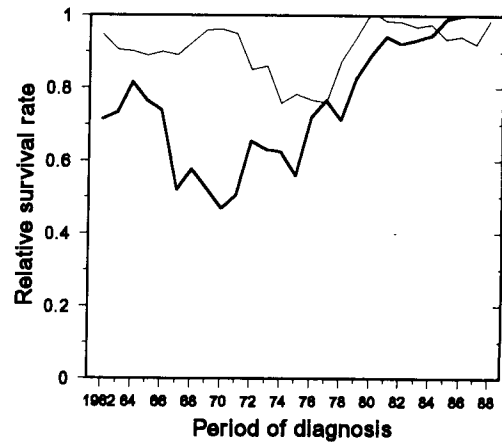
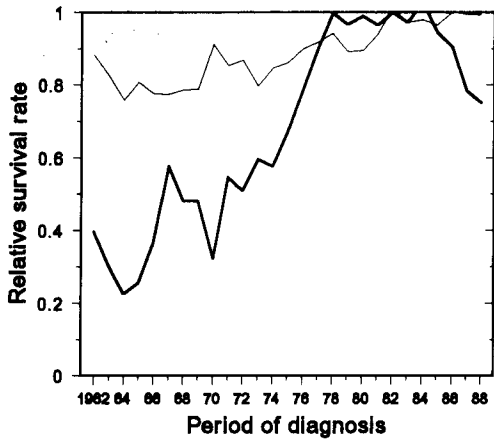


Fig. 12. Intracranial nerves. Age-group specific 5-year relative survival of a) males and b) females, 0–54 (—) and 55–74 (—) years of age. 3-year moving averages.

while combinations of surgery, radiotherapy and chemotherapy have been applied in the treatment of intracerebral lesions.

Materials

For the period 1961–89, 2 557 men and 3 369 women were excluded from the analysis due to the exclusion of extracranial nerves and other criteria described in Methods and Material. A total of 11 858 men and 12 610 women with a diagnosis of intracranial tumor were included in the analysis.

When cases aged 90 years or older at diagnosis had been excluded, the mean age at diagnosis was 52 years in men and 54 years in women. Over the entire period, 30% of men and 35% of women were between 65 and 89 years of age.

Results

Intracerebral tumors

The diagrams (Figs. 4a, b) on relative survival reveal a rather poor prognosis both for males and females; low levels of survival are reached as early as the first year. Stabilization of the relative survival (RS) curve occurs, however, at a clearly higher level for cases diagnosed in 1980–82 than for those diagnosed earlier, and is reached after some 6 years after diagnosis. 1-year crude (CS) and relative survival (RS) have improved by some 16–17 percentage points for men and 20–21 percentage points for women to reach 47–48% for males and 52–53% for females diagnosed in 1987–89 (Table 2). 1-year RS has improved more than 5 and 10-year RS (Figs. 5a, b). The 5-year survival rates (CS and RS) have increased from 1961–63 to 1985–87 from 15–17% to approximately 25–30% (Table 2). 10-year RS rates reached 23% for men and 27% for women in the 1980–82 cohorts from the 14% observed for both sexes in the beginning of the observation period. The survival figures are much better and have improved more for persons who are 54 years or younger at diagnosis than for older persons (Figs. 6a, b).

Meningiomas

As with intracerebral tumors, the diagrams on RS (Figs. 7a, b) show that most fatalities occurred during the first year after diagnosis. Survivors after the first year have good survival. The 1-year CS improved radically from 48% in 1961–63 to 87% in 1985–87 for males and from 46% to 90% for females. The figures on 5 and 10-year CS are just as encouraging; 5-year CS increased by 33 percentage points up to 71% for males and 41 percentage points up to 81% for females in 1985–87, and 10-year CS increased by 22 percentage points for males and 33 percentage points for females from 1961–63 to 1980–82 (Table 3).

5-year RS for males was 82% and for females 88% in 1985–87 and the 10-year RS for males was 73% and for females 80% in 1980–82. It is seen in Figs. 7a and b that the increase is due to the radical improvement in the first year after diagnosis, an improvement that seemed to persist for subsequent years.

1, 5, and 10-year RS increased slowly for both sexes from around 50% to between 55% and 63% from 1961–63 to 1975–77. Between 1975–77 and 1980–82 a radical increase was observed when 1-year RS rose by 30 percentage points to 90–92%, 5-year RS reached 83% for males and 86% for females. The steep increase is obvious in Figs. 8a and b. After the jump in the early 1980's, RS has stayed on a constantly high level.

Survival is less favorable in older age groups, but in contrast to the intracerebral tumors there has been a great improvement in survival both in the 0–54 and 55–74 age groups (Figs. 9a, b). Although the same development also appears to have taken place in the oldest age group, the incidence of meningioma is too low here to make any definite statements about this group.

Neurinomas

The relative survival curves (Figs. 10 and 11) are close to those for meningiomas, also displaying a radical increase in survival in the 1970's with even higher levels of current survival. Towards the end of the observation period, 1 and 2-year RS for men was 97% and 96%, and for women 98% and 99% respectively (Table 4). Neuroma patients survived to the same extent as the control group in the short and long-term. The 5-year RS was 95% for men in 1985–87 and 97% for women, and the 10-year rates were 99% and 96% respectively, for men and women diagnosed in 1980–82.

As with meningiomas, there was a clear age gradient putting older patients at a disadvantage in terms of RS at the beginning of the period, but the differences between age groups were completely eliminated across the observed period. Age comparisons were only possible for the age groups 0–54 and 55–74 (Figs. 12a, b).

Discussion

The results emphasize the importance of giving separate accounts of survival in extracerebral and intracerebral tumors. Neurinomas and meningiomas are frequent and have had a radically different development of survival. If included in the group of intracerebral tumors, their presence in the material would have considerably affected the survival figures.

Being almost 100% histologically benign (and localized) tumors, it is well understandable that meningiomas and neurinomas have an extremely good prognosis. It would appear that the radically improved survival of patients

with these tumors can, to a large extent, be explained by achievements in operation techniques and preoperative care and treatment. Judging from the shapes of the curves in the diagrams (Figs. 8 and 11), the most important improvements were seen in the late 1970's and in the beginning of the 1980's. The same appears to apply to the treatment of patients with intracerebral tumors, although the situation is more complex in these cases in that not only achievements in surgical technique but also in radiotherapy and chemotherapy must have played important roles. The latter notion seems to be supported by the fact that improved survival among patients with intracerebral tumors was most conspicuous in the youngest age-group (Figs. 6a, b). This group contains a rather large number of children with medulloblastomas, for which greatly improved methods of treatment have been applied during recent decades.

Figures on survival after a diagnosis of intracranial tumor are available from the Danish, Finnish and Norwegian cancer registries. The figures are, however, not directly comparable with those in the present study, since they cover other time periods and the tumor types are not the same (3, 5, 6). Hence, it is not possible to evaluate whether the strong increase in survival of meningioma and neuroma patients has also occurred in other Nordic countries. A slightly improved survival was found in Norway between patients diagnosed in 1963–67 and 1968–71 concerning all tumors (intracerebral and extracerebral) (6). Finnish figures on changes in survival between 1953 and

1974 showed steady improvement for all tumor types (5). For gliomas, the highest 5-year survival rate was 30% for males and 34% for females, while corresponding figures for meningiomas were 85% and 84%, respectively. Danish figures also covered later time periods but the study did not separate extracerebral from intracerebral tumors (3). In the latest cohort studied (patients diagnosed in 1983–87), relative 5-year survival in males had increased to 36% (from 25%), and in females from 34% to 48%.

REFERENCES

1. Cancer Incidence in Sweden 1991. Centre for Epidemiology. National Board of Health and Welfare. Fritzes, Stockholm, 1994.
2. Tulinius H, Storm HH, Pukkala A, Andersen A, and Ericsson J. Cancer in the Nordic Countries 1981–1986. Association of Nordic Cancer Registries, Nordic Cancer Union. *APMIS* 1992; 100 (Suppl 31): 1–194.
3. Frisch M and Olsen JH. Eye, brain and nervous system, thyroid, bone and connective tissue. In: Carstensen B, Storm HH and Schou G, eds. *Survival of Danish cancer patients*. *APMIS* 1993; 101 (Suppl 33): 156–82.
4. Schoenberg BS. Nervous system. In: Schottenfeld D and Fraumeni JF, eds. *Cancer epidemiology and prevention*. WB Saunders, Philadelphia, 1982: 968–83.
5. Hakulinen T, Pukkala E, Hakama M, Lehtonen M, Saxen E, and Teppo L. Survival of cancer patients in Finland in 1953–1974. *Ann Clin Res* 1981; 13 (Suppl 31): 1–101.
6. Survival of cancer patients. Cases diagnosed in Norway 1968–1975. The Cancer Registry of Norway. Fr Salvesen AS. Oslo, 1980.