Volume 2 • Issue 1 • 2020

Research Article

Journal of Ophthalmology and Vision Research

Age-related Features of the Prevalence and Survival of Patients with Malignant Tumors of the Eye and its Adnexa (C69).

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Received: June 08, 2020; **Published:** June 17, 2020

Summary

Malignant tumors of the eye and its adnexa (C69) are rare malignancies. To date state statistics in Russia (Form No. 7) have data only for estimating the incidence of the population. These data, at our proposal, were included in Form No. 7 only since 2011. In 2018 for the first time we were able to have data on the incidence of the population by age separately for men and women but only in Russia as a whole. The estimation of rates of a 1- and 5-year patient survival for this pathology is carried out only by us in the North-West Federal Region of Russia, where, in February 2019, we have formed the first Population-based Cancer Registry (PCR) in Russia at the Federal Region's level with a total database of more than 1 million cases. Previously similar work was conducted on the materials of the Population-based Cancer Registry of St. Petersburg.

For the first time this work presents the features of data on changes in the structure of oncological pathology of the eye and its adnexa (C69) and estimations of a 5-year survival by age. Unfortunately the Office of the Federal State Statistics Service classifies the pathology of the eye (C69) as a group of "other malignant tumors" and does not publish mortality data. At the same time, using the PCR database, it must be borne in mind that index accuracy (the ratio of a number of deaths to a number of sick for the first time in life) according to malignant tumors of the eye (C69) is about 0.50% and is constantly decreasing.

Key words: *Eye; Malignant tumors; Incidence; Structural changes; Index accuracy; Survival of patients; Sex; Age; Russia; St. Petersburg; North-West Federal Region of Russia.*

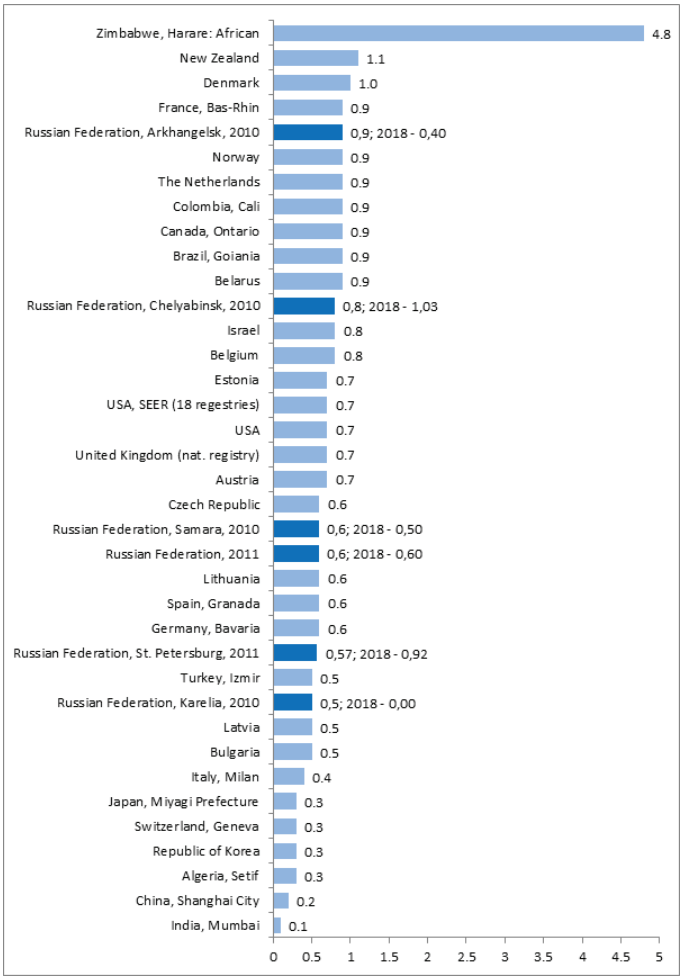
Malignant tumors of the eye and its adnexa (C69) make up 0.18% in Russia (1153 cases out of 624709 in 2018). Being a rare site of malignancies, data on this pathology have only become public in the medical community since 2011 [5]. Prior to this period the frequency of this group of tumors could be judged only by the databases of the Population-based Cancer Registries of separate territories, where even now single cases of diseases are recorded for the calendar year. Even in 2018 only 1 case of malignant tumor of the eye (C69) was registered in such administrative territories as the Kostroma, Murmansk, Magadan regions, the Republics of Ingushetia and Karachay-Cherkess. Not a single case of this disease has been registered in the Chukotka Autonomous Area [5]. In connection with the above, to study the patterns of morbidity dynamics and to assess the survival of patients with malignant tumors of rare

sites, special importance belongs to the formation of territorial cancer registries at the level of Federal Regions, the first of which is the PCR of the North-West Federal Region of Russia. The estimation of a 1- and 5-year survival rates for this pathology is carried out only by us in the West Federal Region of Russia, where in February 2019 we formed the first Population-based Cancer Registry in Russia at the level of the Federal Region with a total database of more than 1 million cases. Previously similar work was conducted on the materials of the Population-based Cancer Registry of St. Petersburg [6,7,13,14].

First of all let us consider the features of prevalence rate of malignant tumors of the eye (C69) based on the latest IARC monograph "Cancer on 5 Continents", Volume XI, which for the first time included 4 administrative territories of Russia working under our programs in accordance with international requirements: the Arkhangelsk, Samara and Chelyabinsk Regions and the Republic of Karelia (Figure 1,2) [5,12]. The highest incidence of malignant tumors of the eye (C69) is registered among Africans in Zimbabwe (before 1980 - Rhodesia) in both men and women, which can be associated with water containing a large number of dangerous toxins and pathogens. The most typical incidence rate among people of both sexes is from 0.6 to 0.9 0/0000, the minimum incidence of this pathology has been identified in India, Japan and China. The incidence of malignant tumors of the (C69) on average in Russia is at the bottom of the graph. In 2018 in the Republic of Karelia not a single case of this disease was detected (C69), which once again confirms the importance of creating population-based cancer registries across Federal Regions [5,12].

The proportion of malignant tumors of the eye (C69) among all malignancies in different countries is from 0.2% to 0.4% in men and women. The highest incidence rate is registered among patients in the first year of life and at the age of 50 years and older [12].

Table 2 presents the ranking distribution of malignant tumors of the eye (C69) separately for men and women. Among the male population in 8 administrative territories the standardized incidence rate was higher than 1.0 0/0000. The maximum rate of 1.93 0/0000 was registered in the Kurgan Region; in 11 territories not a single case of the disease was registered in 2018. Among the female population the highest incidence rate was observed in the Nenets Autonomous Area - 3.22 0/0000, and more than 1.0 0/0000 was in 12 territories. In 7 territories not a single case of the disease was registered.



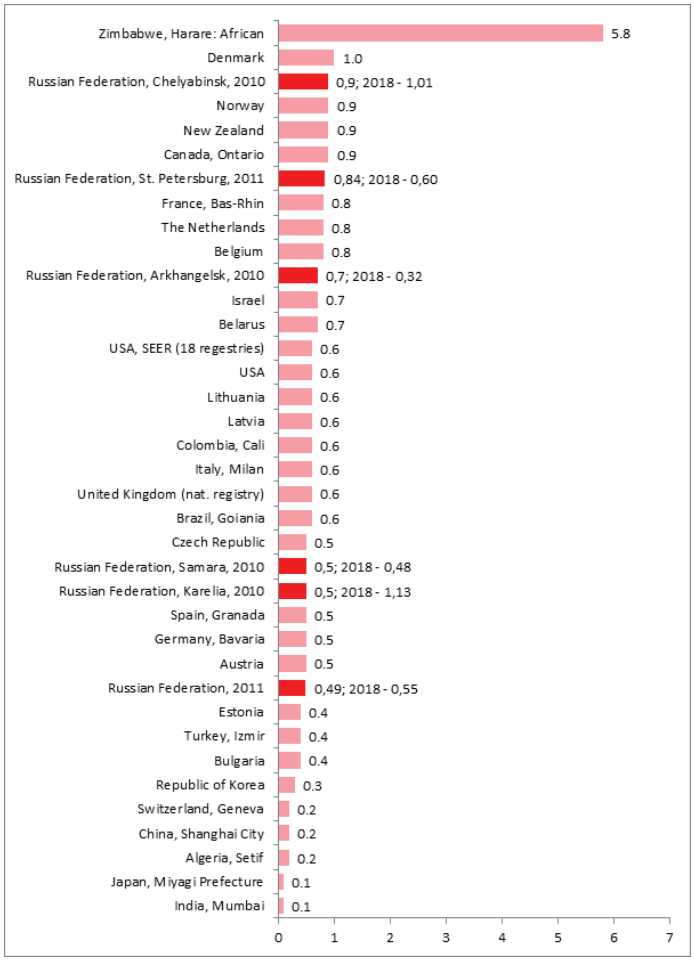
*- directories of the P.A. Gerzen Moscow Cancer Research Institute [1,5].

Figure 1: *Malignant tumors in some countries of the world. The eye and its adnexa. C69. Men. 2008 - 2012. IARC "Cancer on 5 Continents" XI volume.*

2011			
Territory	Absolute number	«Crude» rate	Standardized rate (world standard)
Russia	925	0,65	0,54
North-West Federal Region of Russia	87	0,64	0,50
St. Petersburg	47	0,95	0,72
2018			
Territory	Absolute number	«Crude» rate	Standardized rate (world standard)
Russia	1153	0,79	0,57

North-West Federal Region of Russia	110	0,79	0,59
St. Petersburg	55	1,02	0,71
growth, % (2011-2018 rr.)			
Territory	Absolute number	«Crude» rate	Standardized rate (world standard)
Russia	24,6	21,5	5,6
North-West Federal Region of Russia	26,4	23,4	18,0
St. Petersburg	17,0	7,4	-1,4

Table 1: *The dynamics of the incidence rate of malignant tumors of the eye (C69) in Russia, the West Federal Region of Russia and St. Petersburg from 2011 to 2018 (both sexes) [1,5].*



*- directories of the P.A. Gerzen Moscow Cancer Research Institute [1,5].

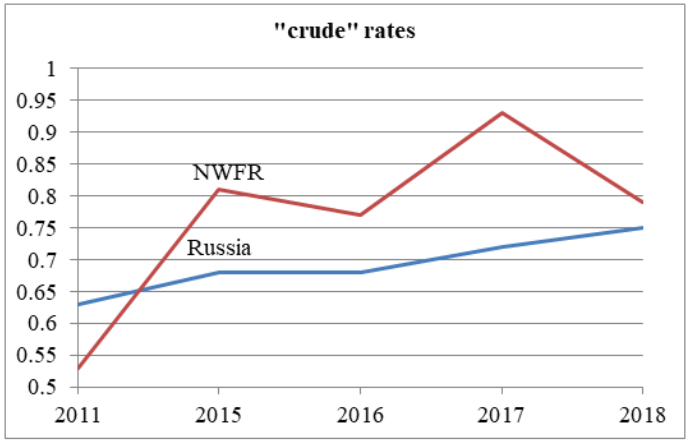
Figure 2: *Malignant tumors in some countries of the world. The eye and its adnexa. C69. Women. 2008 - 2012. IARC "Cancer on 5 Continents" X1 volume.*

Men		Women	
Territory	Standardized rate	Territory	Standardized rate
Kurgan Region	1,93	Nenets Autonomous Area	3,22
Vologda Region	1,36	Republic of Khakassia	1,96
Jewish Autonomous Region	1,21	Komi Republic	1,85
Irkutsk Region	1,15	Republic of Mordovia	1,54
Vladimir Region	1,10	Kamchatka Region	1,21
Chelyabinsk Region	1,03	Republic of Kalmykia	1,16
Altai Region	1,02	Kursk Region	1,14
Novosibirsk Region	1,02	Republic of Karelia	1,13
Tver Region	0,95	Sakhalin Region	1,08
Republic of Crimea	0,94	Ivanov Region	1,06
St. Petersburg	0,92	Orel Region	1,02
…		Chelyabinsk Region	1,01
Komi Republic	0,83	…	
…		Moscow	0,68
Russia	0,60	…	
Kaliningrad Region	0,59	St. Petersburg	0,60
…		Krasnodar Region	0,58
Tomsk Region	0,55	…	
Pskov Region	0,54	Russia	0,55
Moscow	0,52	…	
…		Samara Region	0,48
Samara Region	0,50	…	
Novgorod Region	0,49	Leningrad Region	0,43
Krasnodar Region	0,49	Kaliningrad Region	0,38
…		Arkhangelsk Region	0,32
Arkhangelsk Region	0,40	Murmansk Region	0,12

Table 2: *Rank distribution of malignant tumors of the eye (C69) in the administrative territories of Russia. 2018. [5].*

Thus it is possible to conclude that the assessment of the prevalence of malignant tumors of the eye (C69) should be carried out only at the level of the Federal Region, where the probability elements of random statistical effects can influence the dynamics of processes to a much lesser extent. There is a second option as it is accepted by IARC in the formation of the monographs "Cancer on 5 Continents" to summarize the averaged data for a 5-year time groups, which may be the only analysis option for territories with a small population.

Figures 3 and 4 present the dynamics of "crude" and standardized rates of the incidence of malignant tumors of the eye (C69) in Russia and the North-West Federal Region of Russia. The figures clearly show an element of random scatter of a small number of observations, even at the level of the Federal Region, while in Russia as a whole the dynamics of the incidence of rare tumor sites has a smoother movement.



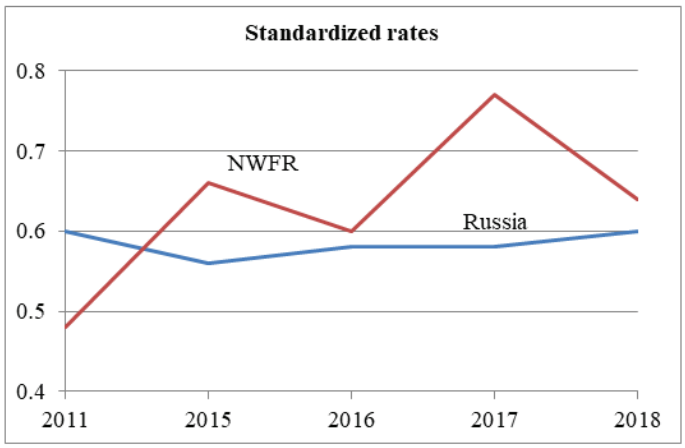
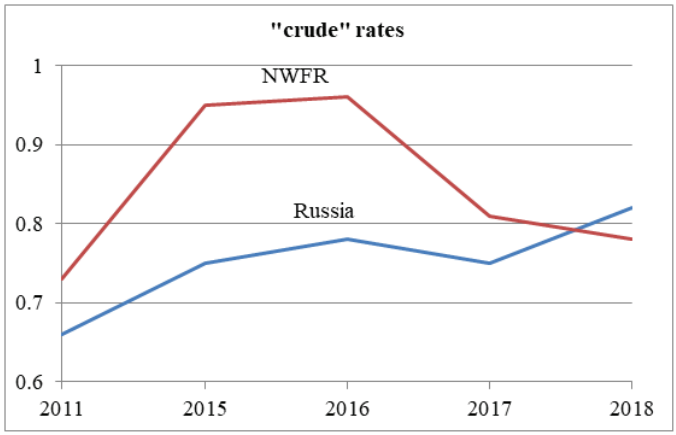


Figure 3: *The dynamics of the incidence of malignant tumors of the eye (C69) in Russia and the North-West Federal Region of Russia. "Crude" and standardized rates (men). [1,5].*



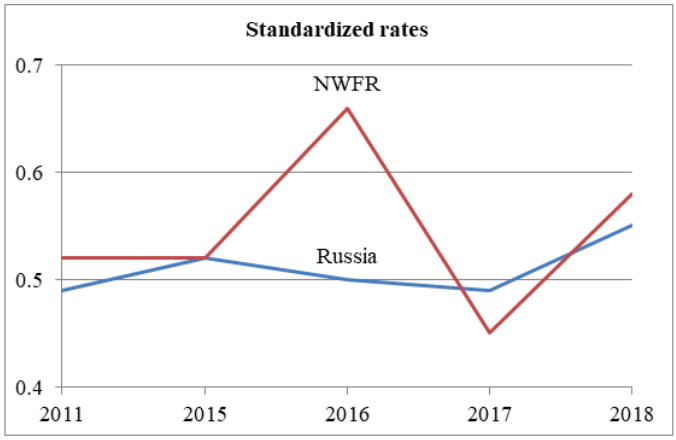


Figure 4: *The dynamics of the incidence of malignant tumors of the eye (C69) in Russia and the North-West Federal Region of Russia. "Crude" and standardized rates (women). [1,5].*

Database of the Population-based Cancer Registry of the North-West Federal Region of Russia allows studying in more detail the patterns of the dynamics of the site and histological structure of malignant tumors of the eye. The total number of cases was 1760. Table 3 presents the dynamics of the detailed localizations structure of the oncopathology of the eye (C69) and its adnexa according to both sexes over two observation periods: 2000-2008 and 2009-2017. The leading role in the site structure of malignant tumors of the eye (C69) belongs to the choroid of the eye (C69.3), the proportion of which in the second observation period was 52.5%. Retinal malignant tumors (C69.2) -10.7% are on the second place, and conjunctival tumors (C69.0)- 8.9% are on the third place. There was also noted a decrease in the proportion of malignant tumors of the unspecified part of the eye (C69.9) from 19.8% to 16.4%, which indicates a slight increase in the quality of primary diagnostics in the region.

Nosology	2000-2008		2009-2017		2000-2017	
	M + F	%	M + F	%	M + F	%
C69.0 malignant tumors of conjunctiva	25	3,3	89	8,9	114	6,5
C69.1 malignant tumors of cornea	10	1,3	7	0,7	17	1,0
C69.2 malignant tumors of retina	65	8,6	107	10,7	172	9,8
C69.3 malignant tumors of choroid	337	44,2	526	52,5	863	49,0
C69.4 malignant tumors of ciliary body	116	15,3	53	5,3	169	9,6
C69.5 malignant tumors of lacrimal gland	5	0,7	11	1,1	16	0,9
C69.6 malignant tumors of eye sockets	30	4,0	31	3,1	61	3,5
C69.8 malignant tumors of the eye and its adnexa, extending beyond one or more of the above sites	21	2,8	13	1,3	34	1,9
C69.9 malignant tumors of undefined site	150	19,8	164	16,4	314	17,8
Total	759		1001		1760	

Table 3: *The dynamics of the detailed structure of the oncopathology of the eye (C69). The North-West Federal Region of Russia. Both sexes. Database of the Population-based Cancer Registry.*

Table 4 presents the dynamics of the histological structure of malignant tumors of the eye (C69). The total number of cases was 1760. The overwhelming majority of malignant tumors account for malignant melanoma NOS* (M8420/3)-42.0%, the second place belongs to spindle cell melanoma NOS (M-8772/3) -10.0%, and the third place belongs to retinoblastoma NOS (M95-10/3)-4.1%. In general it is possible to conclude that the histological structure of malignant tumors of the eye (C69) can be attributed to persistent signs, it has practically not changed over 17 years.

*NOS – not otherwise specifide

ICD-O-2	Histological type of tumors	2000 - 2005			2006 - 2011			2012 - 2017		2000 - 2017	
		Abs. number	Survival 1-year	Survival 5-year	Abs. number	Survival 1-year	Survival 5-year	Abs. number	Survival 1-year	Abs. number	Survival 1-year
8720/3.	Malignant melanoma, NOS	225	91,1	58,9	270	93,0	65,5	287	92,2	782	92,1
8770/3.	Mixed epithelioid and spindle cell melanoma	13	92,3	46,2	9	88,9	43,2	19	93,9	41	92,2
8771/3.	Epithelioid cell melanoma	18	83,3	44,4	18	88,2	56,7	37	82,6	73	84,2
8772/3.	Spindle cell melanoma, NOS	49	98,0	73,0	68	93,9	63,3	68	93,2	185	94,8
9510/3.	Retinoblastoma, NOS	23	95,7	87,0	36	100,0	93,8	28	87,5	87	95,2
	Other	139	83,8	61,8	209	79,8	61,3	244	90,1	592	84,9
	Total	467	89,6	61,7	610	88,8	64,9	683	90,9	1760	89,8

Table 4: *The dynamics of the histological structure of malignant tumors of the eye (C69) in the North-West Federal Region of Russia. Both sexes. Database of the Population-based Cancer Registry of the North-West Federal Region of Russia. The North-West Federal Region of Russia.*

Survival.

To estimate survival rates there were selected 1184 patients with malignant tumors of the eye (C69) including 546 men and 638 women. A 5-year survival rate of these patients for both sexes for three observation periods (1999-2003, 2004-2008, 2004-2013) increased in the North-West Federal Region of Russia from 62.3% to 66.9%, or by 7.4% This rate represents a particular interest for assessing the survival of individuals of various age groups. They were divided into 2 parts: men up to retirement age (from 0 to 59 years old) and older (60+), women up to retirement age up to 55 years old and 55 years old and older (Figure 5, 6). First of all it is important to draw attention to the fact that a 5-year observed survival of men is significantly lower than that of women - 58.5% and 64.8%, respectively.

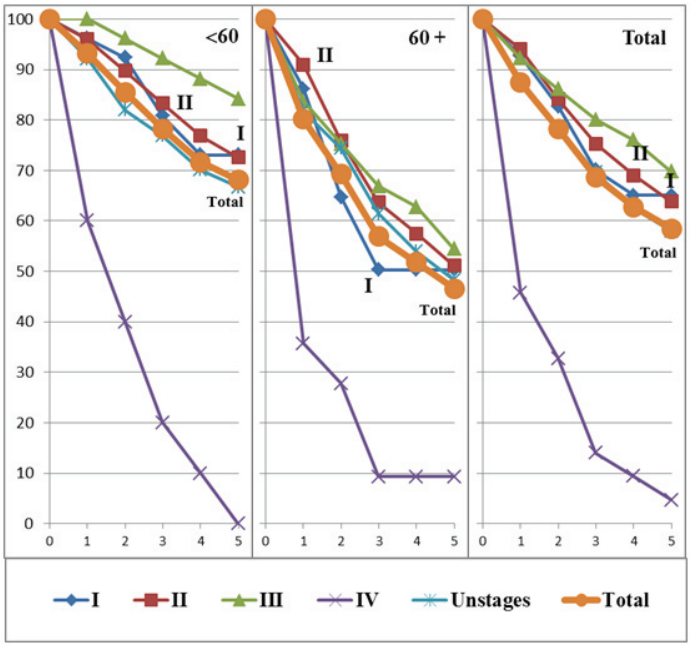


Figure 5: *An observed 5-year survival of men younger 60 years old, and 60 years and older. Malignant tumors of the eye (C69) by the stage of the disease in the North-West Federal Region. Database of the Population-based Cancer Registry of the North-West Federal Region of Russia. The North-West Federal Region of Russia.*

Men of retirement age die within 5 years more often than before retirement age - 46.6% and 68.1%, respectively. Malignant tumors of the eye (C69) are noticeably less frequently detected among the elderly in the early stages (I + II) -34.0% and 43.1%, respectively. We did not reveal such differences in women. A 5-year survival among young women (under 55 years old) is significantly higher than among persons of retirement age (55 years and older) - 81.6% and 53.6%, respectively. These rates are noticeably higher for patients diagnosed at various stages of the disease (Figure 6) [6-11].

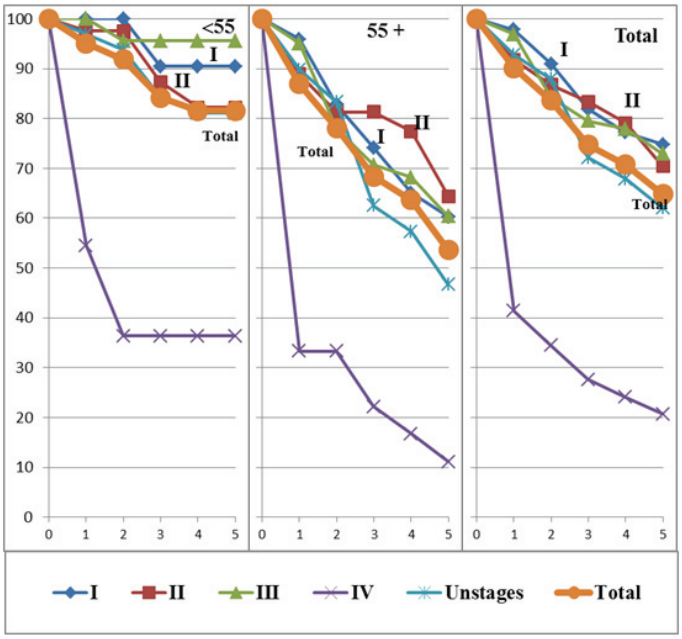


Figure 6: *An observed 5-year survival of women younger 55 years old, and 55 years and older. Malignant tumors of the eye (C69) by the stage of the disease in the North-West Federal Region. Database of the Population-based Cancer Registry of the North-West Federal Region of Russia. The North-West Federal Region of Russia.*

Table 5 demonstrates not only the difference in levels of a 5-year survival but also the paradoxical phenomenon of a higher survival rate of patients with a higher stage, which can be attributed to the diagnostic errors of its establishment when planning treatment of patients.

Thus the use of new opportunities of the consolidated database of population-based cancer registries of the North-West Federal Region of Russia, formed at the federal level made it possible to more clearly determine the dynamic processes of the prevalence of malignant tumors of the eye and survival of patients. It has been established that in the vast majority of economically developed countries the standardized incidence rate of malignant tumors of the eye in both men and women is in the range of 0.6-0.90/0000. The increase in the rate mainly associated with the process of aging of the population and the intensification of work on the prevention of

cancer; the histological structure of this malignancy remains virtually unchanged. Survival of patients is growing rather slowly but an unfavorable trend is clearly identified for the worst survival rates of the population of retirement age.

Age	Stages of disease					
	I	II	III	IV	No stage	Total
Men						
< 60 years	73,1	72,6	84,2	0	66,7	68,1
60 years >	50,3	51,1	54,4	9,3	48,3	46,6
Women						
< 55 years	90,5	82,2	95,6	36,4	81,1	81,6
55 years >	60,3	64,4	60,4	11,1	46,7	53,6

Table 5: *An observed 5-year survival of patients with malignant tumors of the eye (C69) by the stage of the disease and age group in the North-West Federal Region. Database of the Population-based Cancer Registry of the North-West Federal Region of Russia. The North-West Federal Region of Russia. 1999-2018.*

References

1. Злокачественные новообразования в России в (2011) году (заболеваемость и смертность)/Под ред. В.И. Чиссова, В.В. Старинского, Г.В. Петровой.-М.: ФГБУ «МНИОИ им. П.А. Герцена» Минздрава России, 2013.-289 с. (in Russian)
2. Злокачественные новообразования в России в (2015) году (заболеваемость и смертность)/Под ред. А.Д. Каприна, В.В. Старинского, Г.В. Петровой. -М.: МНИОИ им. П.А. Герцена-филиал ФГБУ «НМИРЦ» Минздрава России, 2017.-250 с. (in Russian)
3. Злокачественные новообразования в России в (2016) году (заболеваемость и смертность)/Под ред. А.Д. Каприна, В.В. Старинского, Г.В. Петровой.-М.: МНИОИ им. П.А. Герцена-филиал ФГБУ «НМИЦ радиологии» Минздрава России.-2018.- 250 с. (in Russian)
4. Злокачественные новообразования в России в (2017) году (заболеваемость и смертность)/Под ред. А.Д. Каприна, В.В. Старинского, Г.В. Петровой.-М.: МНИОИ им. П.А. Герцена-филиал ФГБУ «НМИЦ радиологии» Минздрава России.-2018.- 250 с. (in Russian)
5. Злокачественные новообразования в России в (2018) году (заболеваемость и смертность)/ Под ред. А.Д. Каприна, В.В. Старинского, Г.В. Петровой - М.: МНИОИ им. П.А. Герцена-филиал ФГБУ «НМИЦ радиологии» Минздрава России, 2019.- 250 с. (in Russian)
6. Мерабишвили В.М. Выживаемость онкологических больных. Выпуск второй. Часть I. / Под ред. Ю.А. Щербука - СПб.: ООО «Издательско-полиграфическая компания «КОСТА», (2011).- 332 с. (in Russian)
7. Мерабишвили В.М. Выживаемость онкологических больных. Выпуск второй. Часть II/ Под ред. Ю.А. Щербука.- СПб.: ООО «Издательско-полиграфическая компания «КОСТА», (2011).- 408 с. (in Russian)
8. Мерабишвили В.М. Онкологическая статистика (традиционные методы, новые информационные технологии): руководство для врачей. Часть I.- СПб.: ООО «Издательско-полиграфическая компания «КОСТА», (2011).- 221 с. (in Russian)
9. Мерабишвили В.М. Онкологическая статистика (традиционные методы, новые информационные технологии): руководство для врачей. Часть II. - СПб. : ООО «Издательско-полиграфическая компания «КОСТА», (2011). - 248 с. (in Russian)
10. Мерабишвили В.М. Злокачественные новообразования в Северо-Западном федеральном округе России (заболеваемость, смертность, контингенты, выживаемость больных). Экспресс-информация. Выпуск третий/ под ред. А.М. Беляева.- СПб.: Т8. Издательские технологии, (2017).- 282 с. (in Russian)
11. Мерабишвили В.М. Злокачественные новообразования в Северо-Западном федеральном округе России (заболеваемость, смертность, контингенты, выживаемость больных). Экспресс-информация. Выпуск четвертый. Пособие для врачей/Под ред. А.М. Беляева.-СПб.:Т8. Издательские технологии, (2018).-444 с. (in Russian)
12. Cancer incidence in five continents. Vol. XI. IARC Scientific Publication №166. Lyon. (2019). (http://ci5.iarc.fr/CI5-XI/default.aspx 11.01.2020)
13. Sant M., Aareleid T., Berrino F., Bielska Lasota M., Carli P.M., Faivre J., Grosclaude P., Hedelin G., Matsuda T., Moller H., Moller T., Verdecchia A., Capocaccia R., Gatta G., Micheli A., Santaquilani M., Roazzi P., Lisi D., editors. Eurocare-3: survival of cancer diagnosed 1990-1994- results and commentary. Annals of Oncology. (2003) 14(5): v61-118.
14. Sant M., Allemani C., Santaquilani M., Knijn A., Marchesi F., Capocaccia R., editors. Eurocare-4. Survival of Cancer patients diagnosed in 1995-1999. Results and commentary. European Journal of Cancer. (2009) 45(6): 931-91.