



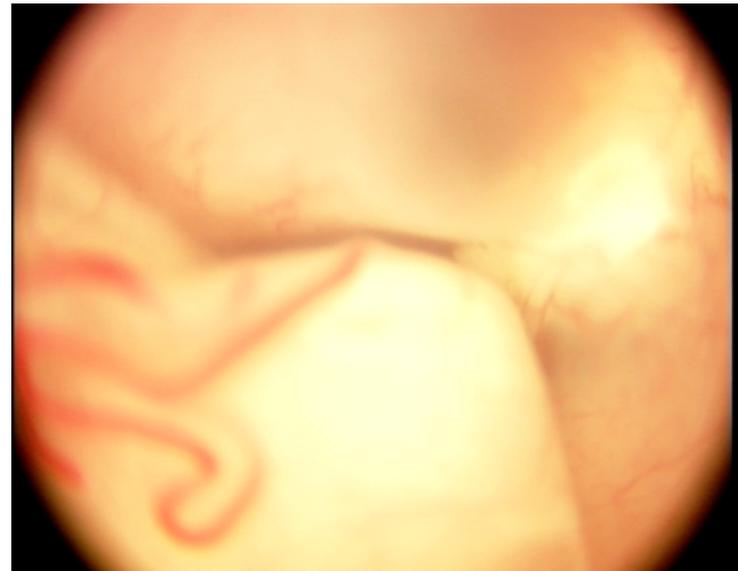
World Eye Cancer Hope
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The Present and the future of retinoblastoma care in Russia

Tatiana Ushakova

Department of head and neck tumors, SRI of pediatric oncology and
hematology of N.N.Blokhin CRC, Moscow, Russian Federation

The disease is so rare that the primary care physicians may meet no more than one case of RB in their entire practice.

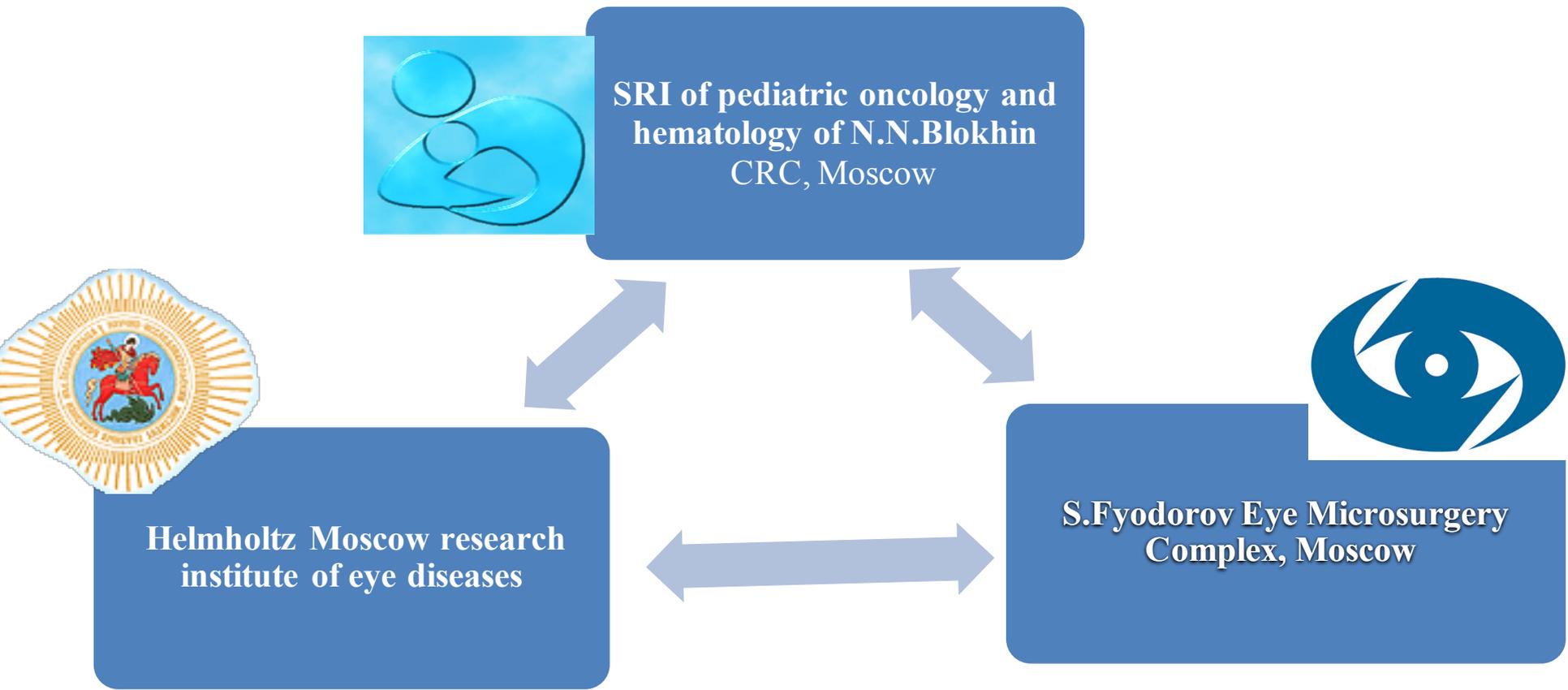


- There are about 100 patients with RB per year in Russia.

In the beginning I would like to say that :

- organ-saving chemoradiotherapy** in Russia for large intraocular RB has been applied since 1976 in the SRI of pediatric oncology and hematology of **N.N.Blokhin** CRC, Moscow (B. Belkina 1994-1997)
- **organ-saving local therapy** in Russia , such as brachytherapy (1976), cryotherapy and laser photocoagulation (1989) in eyes with retinoblastoma, were performed in the Helmholtz Moscow research institute of eye diseases (A. Brovkina , S. Saakyan)
- **Since 2007 Rb local treatment** was introduced in the S.Fyodorov Eye Microsurgery Complex, Moscow (A. Yarovoy 2007)
- **first high-dose chemotherapy** with auto- transplantation of peripheral blood stem cells was performed in the patients with extraocular RB in 2002 in the SRI of pediatric oncology and hematology of **N.N.Blokhin** CRC, Moscow (I.Dolgopolov, 2002)
- **the risk-adapted therapy of unilateral RB after primary enucleation** based on histological criteria was established in 2002 in the SRI of pediatric oncology and hematology of **N.N.Blokhin** CRC, Moscow (T. Ushakova 2012)
- first SIAC was performed -N.N. Burdenko Neuro-Surgery Institute, Moscow in 2010
- **SIAC** was performed in the **N.N.Blokhin** CRC, Moscow in 2011 (I. Trofimov, 2011).
Currently we have 4 SIAC per week.
- **The first intraocular Melphalan injection** was introduced in the SRI of pediatric oncology and hematology of **N.N.Blokhin** CRC, Moscow, in 2011(T. Ushakova, O. Gorovtsova 2011)

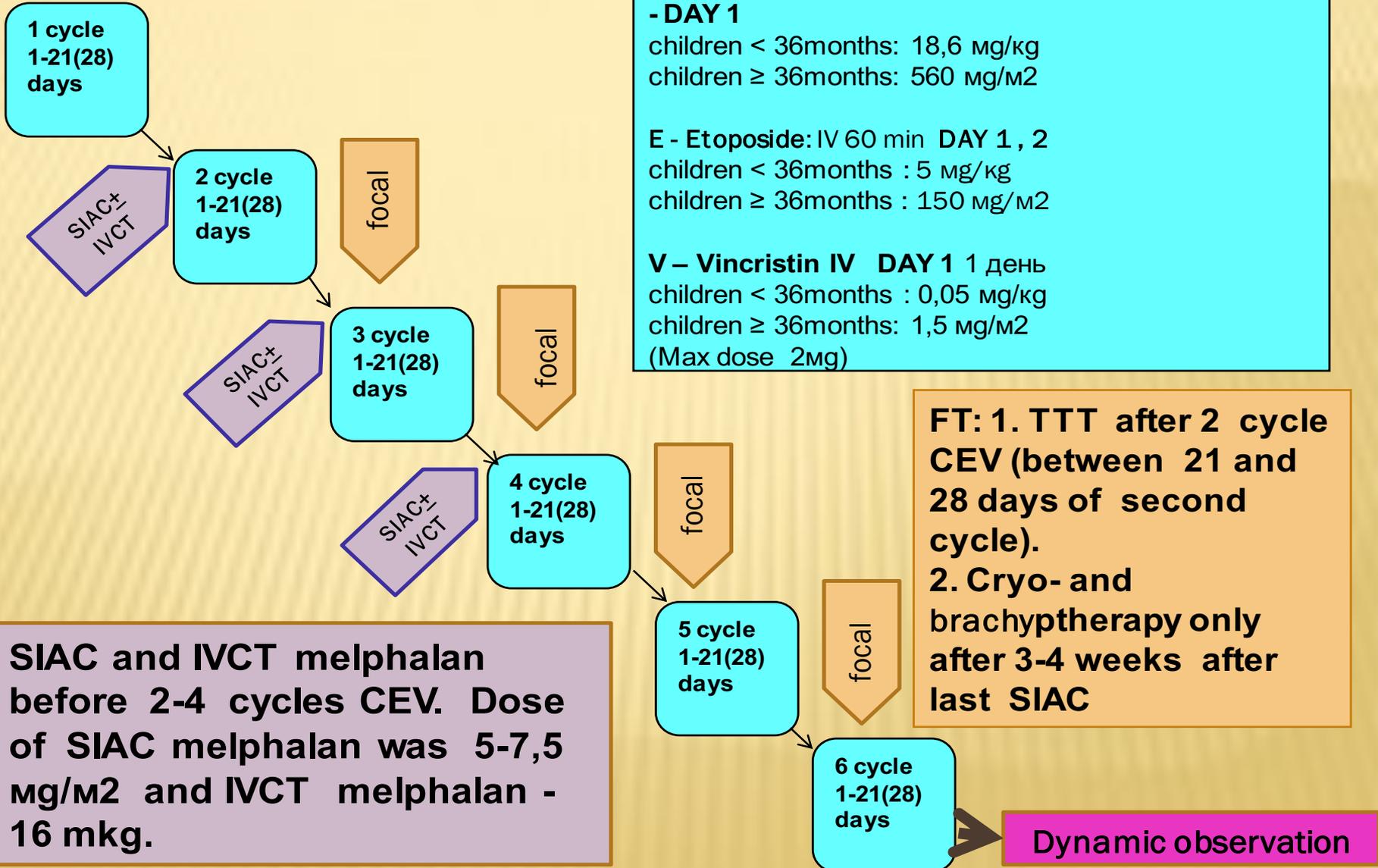
The Protocol RB -2012 of the organ-saving treatment of Rb children was approved and adopted in 2013. This was the first link of multicenter study between three main institutions of Federal significance, where children with retinoblastoma were treated.



The main idea was to use methods which were carried out by every institution for the treatment of Rb patients with different stages.

Institutions	Focal therapy	Systemic CT	SIAC	IViC	RT	Surgery
	no	Yes (100%)	Yes (100%)	Yes (80%)	Yes (100%)	Yes (30%)
	Yes (50%)	no	no	Yes (10%)	no	Yes (30%)
	Yes (50%)	no	no	Yes (10%)	no	Yes (30%)

Protocol RB 2012 (gr. C and D)



CEV:

C - Carboplatin IV 60 min. in 125 ml/m² dextrose 5% - DAY 1

children < 36months: 18,6 mg/kg

children ≥ 36months: 560 mg/m²

E - Etoposide: IV 60 min DAY 1, 2

children < 36months : 5 mg/kg

children ≥ 36months : 150 mg/m²

V – Vincristin IV DAY 1 1 день

children < 36months : 0,05 mg/kg

children ≥ 36months: 1,5 mg/m²

(Max dose 2mg)

FT: 1. TTT after 2 cycle CEV (between 21 and 28 days of second cycle).

2. Cryo- and brachytherapy only after 3-4 weeks after last SIAC

SIAC and IVCT melphalan before 2-4 cycles CEV. Dose of SIAC melphalan was 5-7,5 mg/m² and IVCT melphalan - 16 mkg.

Dynamic observation

Preliminary results were presented at the SIOP ASIA 2016

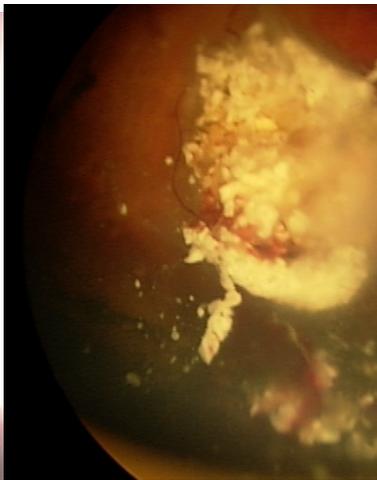
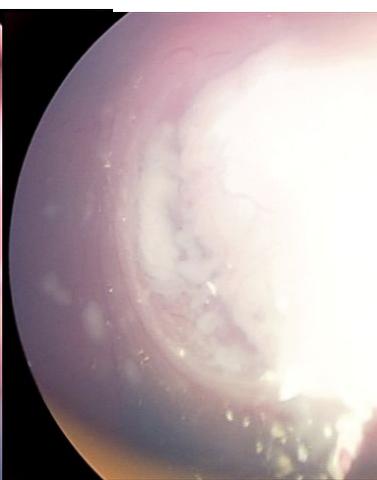
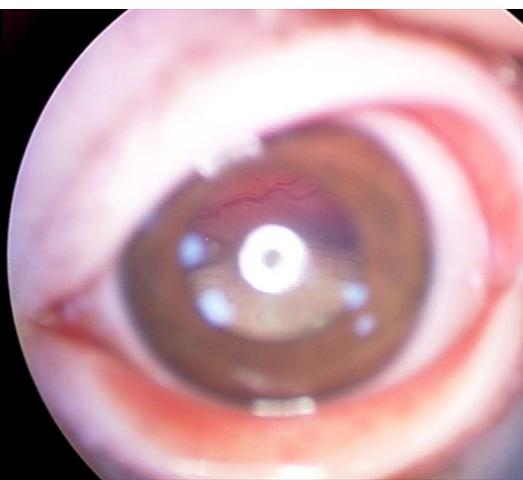
The Rb patients of groups «C» and «D» had disease free survival (DFS) of 56,1+8,9% with a median period of follow-up of 26,9+ 2,5months.

At a median follow up of 20 months (range 3-43 months) 44 of 45 patients with Rb of groups «C» and «D» are alive with no metastatic disease. One of 45 patients died of secondary malignant tumor - acute leukemia. 14 of 15 eyes (93,3%) with group «C» and 31 of 34 eyes (91,2%) with group «D» were preserved.

It should be noted that the second line chemotherapy was used in 2 cases, RT - in 3 cases.

In our opinion, these methods of treatment, should be considered as an act of desperation and failure of treatment.

Focal therapy and local chemotherapy were the main treatment for progression, recurrence and stabilization disease.



The Future of retinoblastoma care in Russia

It is necessary to create

- a single coordinating center with the participation of leading specialists from leading centers: oncologists, ophthalmologists, radiation diagnosticians, chemo - and radiotherapists, molecular biologists, genetics, pediatricians, anesthesiologists, psychologists, specialists in medical rehabilitation, social workers, programmers , charities

- a unified database for Rb patients

- a unified database of the morphological material of Rb patients

- molecular genetic studies will make a significant contribution to enhancing the effectiveness of genetic counseling, will give a more accurate disease prognosis , the prognosis of progeny

- multi-level study module on RB (students, pediatric oncologists, residents in ophthalmology, for the qualification exam of ophthalmologists and oncologists)

- the international network and cooperation with other centers and internships for young scientists



*Thank you for your
attention*