



**Results of treatment rhabdomyosarcoma orbit in children.**  
 Buletov D. (1), Ushakova T. (1), Gorovtsova O. (1), Ivanova N. (1), Yarovoy I. (3), Saakyan C. (2)  
 Polyakov V. (1)  
 (1) – SRI of pediatric oncology and hematology of N.N. Blokhin Cancer Research Center, Moscow, Russia  
 (2) – Moscow Helmholtz Research Institute of Eye Diseases, Moscow, Russia  
 (3) - Department of Ocular Oncology, S.Fyodorov Eye Microsurgery Complex, Moscow Russian Federation

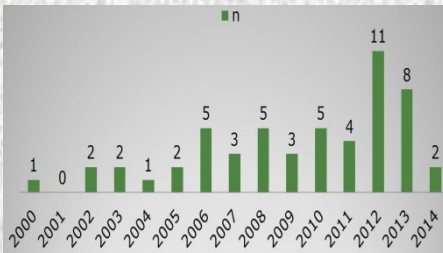
SIOP6  
0628



**Malignant tumors of the orbit:**

- Cancer of the lacrimal gland
- Primary cancer orbit
- Rhabdomyosarcoma
- Liposarcoma
- Leiomyosarcoma
- PNET
- Chondrosarcoma
- Esthesioneuroblastoma
- Osteosarcoma
- Ewing's sarcoma
- Metastatic lesions of the orbit

Most patients were from **Moscow and the Moscow region**, but rhabdomyosarcoma met in every region of the Russian Federation



**Instrumental methods of examination of patients with tumors of the orbit:**

- History of disease
- Consulting of an ophthalmologist, oncologist (if necessary, specialists in related disciplines)
- US orbit and soft tissues of the neck
- CT and/or MRI of the brain and orbits
- Chest X-ray
- US of the abdominal cavity, retroperitoneal space
- Scanning of soft tissue and skeleton

**Our protocol\***

**Carboplatin**  
 360 mg/m<sup>2</sup> 1 day  
 12 mg/kg 1 day  
**Cyclophosphamide** 500 mg/m<sup>2</sup>  
 1,8 days

15 mg/kg 1,8 days  
**Vincristine**  
 1 mg/m<sup>2</sup> 1,8 days  
 0.05 mg/kg 1,8 days  
**Doxorubicine**  
 20 mg/m<sup>2</sup> 2,4 days

1 mg/kg 2.4 days  
 We use doses mg/m<sup>2</sup> for children more then 10 kg body mass.  
 For children less, then 10 kg we use doses for their body mass.

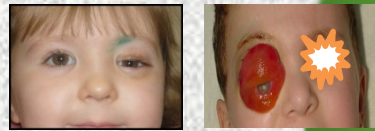
**Differential diagnosis**



Lymphoma  
 Orbital MTS of Neuroblastoma

**Frequency of rhabdomyosarcoma in various parts of the body in children (our data)**

Localisation	%
Head and Neck	35(8% - orbit)
Extremity	25
Genito-urinary system	20
Body	10
Others	10



**Background/Objectives**

**Among all soft tissue sarcomas the most common is rhabdomyosarcoma (4%). The most frequent localization - head and neck, the orbit is 7%.**

**Design/Methods**

**70 children with tumors of the orbit appealed to the Russian Cancer Research Center N.N. Blokhin (RCRC) from January 2000 to December 2014. Diagnosis of RMS is exposed for the first time in 49 cases (92.5%), local recurrence 4 (7.5%) cases. The average age of patients 79.8 ± 47.9 months. Local recurrence had 4 patients. The fate of 3 patients after treatment is unknown, 1 patient with multiple malformations died at a hospice after chemotherapy. 1 of 45 patients came out from the study due to the refusal of parents of treatment. Surgery was performed on 44 patients: Only a biopsy -18, organ-saving surgery - 15, exenteration 11.**

**Results**

**Relapse-free survival in primary embrional RMS group, depending on the program chemotherapy with an average follow-up of 106.8 ± 12.4 months. Using VA -50.9 ± 1.33% (n=18). Local protocol (Carb/VCR/Cpm/Doxo) 81.3 ± 9.8% (n=18). Overall survival in the group of primary patients (n = 44) was 97.7 ± 2.3%, with an average follow-up of 1535 ± 3.4 months. Of the four patients with recurrent rhabdomyosarcoma 2 recovered, 2 died from generalization.**

**Conclusion**

**The preferred method of organ-saving treatment at the present stage is chemoradiotherapy with a rational approach to the selection of the mode of chemotherapy and radiotherapy. The main factors influencing the disease-free survival: are the ability to remove the primary tumor and adequate choice of chemotherapy program. Organ-operation in orbit is preferably carried out initially, if this is feasible, otherwise - just at the height of the chemotherapy effect and are designed to ensure: a radical removal of the tumor within the healthy tissue, maximum safety of orbital structures.**

Correspondence

Department of head and neck tumors, science research institute of Pediatric Oncology and Hematology of N.N. Blokhin Russian Cancer Research Center, 24, Kashirskoe Shosse, Moscow, Russian Federation. 115478

Dmitry Buletov [dmitry@buletov.ru](mailto:dmitry@buletov.ru)  
 Tatiana Ushakova [ushtat07@mail.ru](mailto:ushtat07@mail.ru)