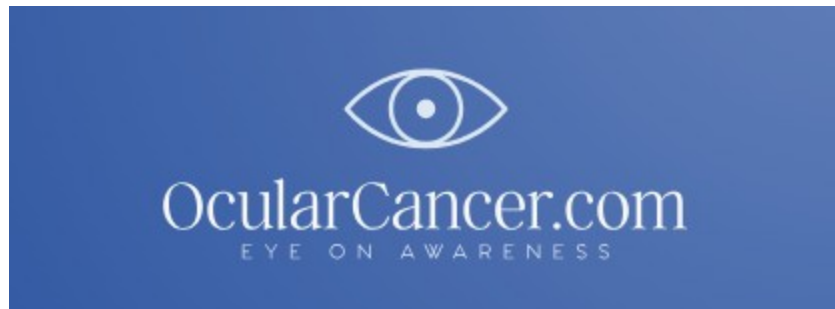




Rhabdomyosarcoma (RMS) Fact Sheet

Cancer Type: Rare Soft Tissue Sarcoma

Common Ocular Site: Orbit (space around the eye)

Most Affected Group: Children under 10

Website Reference: [OcularCancer.com](https://www.OcularCancer.com)

What is Rhabdomyosarcoma?

Rhabdomyosarcoma (RMS) is a **rare and aggressive cancer** that forms in **skeletal muscle cells** that have not yet fully developed. It is the **most common soft tissue sarcoma in children** and can occur anywhere in the body. When it develops in the **orbit**, it is known as **orbital rhabdomyosarcoma**, and is considered a **pediatric ocular cancer emergency** due to its rapid growth.

Key Facts:

- **Most common soft tissue cancer in children**
- **Accounts for ~4–8% of all childhood cancers**
- **Peak onset age:** 1 to 5 years old
- **Gender:** Slight male predominance
- **Orbital RMS:** ~10% of all rhabdomyosarcoma cases

- **Laterality:** Usually **unilateral** (one eye)
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Causes & Risk Factors:

- **Mostly sporadic** (no known cause in most cases)
 - **Genetic syndromes associated with higher risk:**
 - Li-Fraumeni syndrome
 - Neurofibromatosis type 1 (NF1)
 - Beckwith-Wiedemann syndrome
 - **Chromosomal abnormalities:** PAX3/7-FOXO1 fusion genes (seen in alveolar subtype)
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Types of Rhabdomyosarcoma:

1. **Embryonal (ERMS)** – Most common type (especially in the orbit)
 - Better prognosis
 - Seen in younger children
 2. **Alveolar (ARMS)** – More aggressive
 - Often in older children/teens
 - Less common in orbital RMS
 3. **Pleomorphic RMS** – Rare, typically in adults
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Orbital RMS: Signs and Symptoms

- **Rapid-onset bulging of one eye** (proptosis)

- Swelling or redness around the eye
 - Drooping eyelid (ptosis)
 - Eye pain or discomfort
 - Impaired eye movement or double vision
 - Vision changes or loss (in advanced cases)
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Diagnosis:

- **Urgent referral** to ocular oncologist or pediatric oncologist
 - **Imaging:**
 - MRI of the orbit and brain
 - CT scan to evaluate bone involvement
 - **Biopsy:** Required for definitive diagnosis and subtype classification
 - **Bone marrow biopsy** and **lumbar puncture** may be needed to rule out metastasis
 - **Staging:** Based on tumor size, location, lymph node involvement, and distant spread
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Treatment Options:

- **Multimodal therapy is standard:**
 - **Chemotherapy:** Primary treatment (e.g., VAC – Vincristine, Actinomycin-D, Cyclophosphamide)
 - **Radiation therapy:** Often used to preserve eye and vision, especially in orbital RMS
 - **Surgery:** Sometimes performed for biopsy or resection, but often avoided in orbital cases to preserve function

Prognosis:

- **Orbital RMS has a favorable prognosis** with prompt treatment
- **5-year survival rate:** ~85–90% for orbital embryonal RMS
- Prognosis depends on:
 - Subtype (embryonal > alveolar)
 - Stage at diagnosis
 - Tumor location and size
 - Response to therapy

Follow-Up & Monitoring:

- Frequent imaging and eye exams post-treatment
- Monitor for recurrence, side effects, and secondary cancers
- Long-term care may include:
 - Visual rehabilitation
 - Cosmetic or reconstructive surgery (if necessary)
 - Supportive care for growth or cognitive development

Support & Resources:

- Pediatric oncology and ocular oncology teams
- Genetic counseling for high-risk families

- Psychosocial and mental health support
 - Childhood cancer survivor groups
 - Vision support programs and special education services
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Key Takeaways:

- **Rhabdomyosarcoma is rare but highly treatable**, especially in the orbit.
 - **Early diagnosis is critical** - rapid eye bulging in a child is a red flag.
 - Treatment requires a **team approach**: oncology, ophthalmology, radiation, and support services.
 - Survivorship care is essential due to **long-term effects of treatment**.
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Learn more and connect with others at:

 [OcularCancer.com](https://www.OcularCancer.com) – Your resource for rare eye cancers, survivor stories, treatment guidance, and community.