🧬 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

🔍 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)

🧬 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)

🧁 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

👁️ 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)

🩺 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

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🎯 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

📌 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**.

📚 **Learn more and connect with others at:**
🌐 **OcularCancer.com** – Your resource for rare eye cancers, survivor stories, treatment guidance, and community.