



## Intraocular Lymphoma Fact Sheet

**Cancer Type:** Rare Eye Cancer (Subtype of Primary Central Nervous System Lymphoma)

**Primary Site:** Retina, vitreous, and/or optic nerve within the eye

**Subtype:** Usually Non-Hodgkin's Lymphoma (Diffuse Large B-cell Lymphoma)

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

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### What is Intraocular Lymphoma?

Intraocular lymphoma (IOL) is a **rare and aggressive eye cancer** that develops in the **lymphoid tissue within the eye**, often affecting the **retina and vitreous**. It typically arises as a **primary intraocular lymphoma (PIOL)**, but may also be **secondary**, spreading from systemic or central nervous system lymphoma.

It is considered a subset of **primary central nervous system lymphoma (PCNSL)** when confined to the eye and/or brain.

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### Key Facts:

- **Prevalence:** Extremely rare; exact incidence unknown, but ~1 case per million people annually
- **Most Common Type:** Diffuse Large B-Cell Lymphoma (DLBCL)
- **Age of Onset:** Typically affects individuals aged **50 - 70**
- **Laterality:** Can be **unilateral or bilateral** (often becomes bilateral)

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### **Causes & Risk Factors:**

- No known definitive cause
- Increased risk in **immunocompromised individuals** (e.g., HIV/AIDS, post-transplant patients)
- Association with **Epstein-Barr Virus (EBV)** in some cases
- Most patients are otherwise immunocompetent

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### **Signs and Symptoms:**

- **Painless, progressive vision loss** (most common)
- Floaters or blurred vision
- Redness and inflammation
- Chronic uveitis that does not respond to steroids
- May mimic autoimmune or infectious eye diseases
- Vitreous haze or retinal infiltrates seen on exam

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### **Diagnosis:**

- Often misdiagnosed as **chronic uveitis**
- Requires high suspicion and multiple tests:
  - **Dilated eye exam** with slit lamp and fundus exam
  - **Optical coherence tomography (OCT)**
  - **Fluorescein angiography**

- **Vitreous biopsy** (gold standard)
  - **Cytology and immunohistochemistry**
  - **Flow cytometry & PCR** for gene rearrangements (clonality testing)
  - **MRI brain and lumbar puncture** to assess CNS involvement
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## ⚙️ **Types of Intraocular Lymphoma:**

### 1. **Primary Intraocular Lymphoma (PIOL):**

- Originates in the eye
- Often part of or progresses to **Primary CNS Lymphoma**

### 2. **Secondary Intraocular Lymphoma:**

- Spread from systemic lymphoma
  - Often involves **uvea (choroid)**
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## 💉 **Treatment Options:**

- **Intravitreal chemotherapy:**
  - Methotrexate or rituximab injections directly into the eye
- **Systemic chemotherapy:**
  - High-dose methotrexate-based regimens
  - Rituximab for B-cell lymphoma
- **Radiation therapy:**
  - External beam radiation (especially if bilateral or recurrent)

- **Autologous stem cell transplant:**
    - In recurrent or CNS-involved disease
  - **CNS-directed treatment** is often needed even if lymphoma appears confined to the eye
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### **Prognosis:**

- **Prognosis varies** depending on spread to CNS and response to treatment
  - **Median survival:** ~3 - 5 years in many studies with CNS involvement
  - **Better outcomes** with early detection and CNS management
  - Risk of **relapse** is high; requires ongoing surveillance
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### **Follow-Up & Monitoring:**

- Regular ophthalmic exams (OCT, fundus photography)
  - Neurological monitoring and imaging
  - Ongoing coordination with hematologist/oncologist
  - Long-term surveillance for recurrence or CNS spread
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### **Support & Resources:**

- Lymphoma support organizations
- Rare cancer advocacy groups
- Ocular oncologists and uveitis specialists
- Mental health support for coping with vision loss or cognitive symptoms


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### **Key Takeaways:**

- Intraocular lymphoma is **rare, serious, and often misdiagnosed**.
- It mimics inflammation - any **persistent or steroid-resistant uveitis** should raise suspicion.
- **Diagnosis is challenging** and requires specialized testing and multidisciplinary care.
- Early and aggressive treatment can preserve vision and extend life.
- **Close monitoring is essential** due to high recurrence and CNS involvement risk.

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 **For more information, support, and survivor stories, visit:**

 [OcularCancer.com](https://www.OcularCancer.com) - A home for rare eye cancer awareness, education, and community.