Retinoblastoma

National Cancer Institute

Source

National Cancer Institute. Retinoblastoma. NCI Thesaurus. Code C7541.

A malignant tumor that originates in the nuclear layer of the retina. As the most common primary tumor of the eye in children, retinoblastoma is still relatively uncommon, accounting for only 1% of all malignant tumors in pediatric patients. Approximately 95% of cases are diagnosed before age 5. These tumors may be multifocal, bilateral, congenital, inherited, or acquired. Seventy-five percent of retinoblastomas are unilateral; 60% occur sporadically. A predisposition to retinoblastoma has been associated with 13q14 cytogenetic abnormalities. Patients with the inherited form also appear to be at increased risk for secondary nonocular malignancies such as osteosarcoma, malignant fibrous histiocytoma, and fibrosarcoma.