

Although retinoblastoma is a rare disorder, it is the most common cancer of the eye in children, accounting for about 3% of all childhood malignancies. Retinoblastoma affects males slightly more often than females. The incidence in the United States and Europe is estimated to be 2-5 children per 1,000,000 people in the general population. The age-adjusted annual incidence for children aged 0-4 in the United States is 10-14 children per 1,000,000. This equates to about 1 in 14,000-18,000 live births. Incidence is the number of newly diagnosed people with a disorder identified in a given year. Two-thirds of children are affected before the age of 2 and more than 90% of retinoblastomas become apparent before the age of five years. The diagnosis of retinoblastoma is made based upon a thorough clinical evaluation, detailed patient history, the identification of characteristic symptoms, and a variety of specialized tests. The presenting symptom is usually leukocoria. A complete examination of the interior of the eye (fundoscopic examination under anesthesia – EUA) may be performed to locate the presence of a tumor or tumors. Magnetic resonance imaging (MRIs) may be used to determine the extent of the tumor(s) and determine if the tumor has spread to surrounding structures or tissue. Ultrasonography may be used to rule out other conditions. Computed tomography (CT) scans are generally avoided because of the potential risk of additional radiation-induced tumors if the child has hereditary retinoblastoma.