delayed diagnosis because health care professionals fail to appreciate the ominous significance of the parents' findings.

The next most common sign is strabismus resulting from poor fixation of the visually impaired eye, particularly if the tumor develops in the macula, the area of sharpest visual acuity. Blindness is usually a late sign, but it frequently is not obvious unless the parent consciously observes for behaviors indicating loss of sight, such as bumping into objects, slowed motor development, or turning of the head to see objects lateral to the affected eye. Other signs and symptoms include heterochromia (different color of the iris), glaucoma, and pain.

Diagnostic Evaluation

A detailed family history and recording of eye symptoms are essential. Children suspected of having this disorder are referred to an ophthalmologist; the diagnosis is usually based on indirect ophthalmoscopy, ultrasound, CT, and MRI scans.

Metastatic disease at the time of retinoblastoma diagnosis is rare (Hurwitz, Shields, Shields, et al, 2016); therefore, staging procedures such as bone marrow aspiration, bone scan, and LP are not routinely performed.

Staging and Prognosis

Staging of retinoblastomas is done under indirect ophthalmoscopy before surgery to accurately determine the tumor size (measured in disc diameters [DDs]) and location (according to an imaginary line called the *equator* drawn on the midplane of the eye) (Hurwitz, Shields, Shields, et al, 2016).

Various classification systems have been used to stage retinoblastoma. The Reese-Ellsworth system classifies patients according to five groups and predicts survival when patients are treated with radiotherapy. A revised classification system, International Classification of Retinoblastoma, was developed in 2003 and is based on the extent and location of the intraocular tumor; it better predicts globe salvage using contemporary treatments. The overall 10-year survival rate is nearly 90% for unilateral and bilateral tumors (Hurwitz, Shields, Shields, et al, 2016). Retinoblastoma is one of the tumors that may spontaneously