Retinoblastoma (RB) is the most frequent intraocular malignancy of childhood. Tumorigenesis has been firmly associated with mutations of the tumor suppressing RB1 gene, while, recently, involvement of additional genetic modifications has been illustrated. Clinically, RB is most commonly presented with leukocoria and strabismus. Imaging techniques supplement diagnosis and contribute to a more accurate tumor assessment. Several classification systems have been introduced for the purpose of planning therapeutic efforts more precisely. Current available treatment options include surgical removal, radiation, chemotherapy and focal modalities. Despite intravenous chemotherapy being the standard treatment, localized methods of administration havealso shown efficiency the past few years. Enucleation still remains a potential alternative, especially in cases with poor prospect of eye salvage. Focal therapy has been utilized mostly as a complementary treatment to chemotherapy.
Key words: Intraocular Malignancy, Leukocoria, Strabismus.