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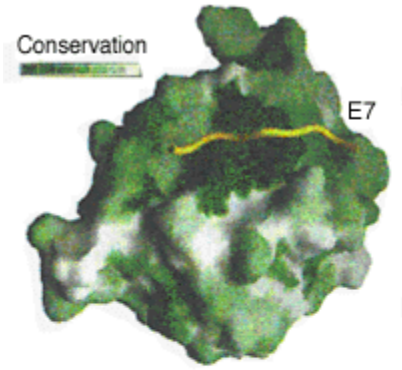
Retinoblastoma



A complex of retinoblastoma protein (RB) with E7 - a viral oncoprotein that frequently binds to RB and blocks its function in cervical cancer. The degree of green color shows the conservation of amino acids in RB and related proteins. [Reproduced from Lee, J-O., Russo, A.A. and Pavletich, N.P. (1998) Nature 391, 859-865, with permission.]

Retinoblastoma occurs in early childhood and affects about 1 child in 20,000. The tumor develops from the immature retina - the part of the eye responsible for detecting light and color. There are both hereditary and non-hereditary forms of retinoblastoma. IN the hereditary form, multiple tumors are found in both eyes, while in the non-hereditary form only one eye is effected and by only one tumor.

In the hereditary form, a gene called Rb is lost from chromosome 13. Since the absence of Rb seemed to be linked to retinoblastoma, it has been suggested that the role of Rb in normal cells is to suppress tumor formation. Rb is found in all cells of the body, where under normal conditions it acts as a brake on the cell division cycle by preventing certain regulatory proteins from triggering DNA replication. If Rb is missing, a cell can replicate itself over and over in an uncontrolled manner, resulting in tumor formation.

Untreated, retinoblastoma is almost uniformly fatal, but with early diagnosis and modern methods of treatment the survival rate is over 90%. Since the Rb gene is found in all cell types, studying the molecular mechanism of tumor suppression by Rb will give insight into the progression of many types of cancer, not just retinoblastoma.

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