FAKTA OM

RETINOBLASTOMA

Retinoblastoma is a tumour in the retina at the back of the eye which affects very young children, generally before the age of two. The disease is very rare, occurring in an average of six to seven children in Sweden a year. Most children affected develop a tumour in only one eye (unilateral retinoblastoma) but a third have tumours in both eyes (bilateral retinoblastoma).

All bilateral tumours are hereditary, but only 10-15 per cent of unilateral tumours. However, it is more common for a child to be the first in the family to suffer from hereditary retinoblastoma than for it to be passed on from a relative. Half of all children who have a parent with hereditary retinoblastoma will inherit the genes in question, and almost all children who carry these genes will develop one or more eye tumours. The risk of developing a tumour is therefore almost 50 per cent for each child with the genes behind hereditary retinoblastoma.

Unilateral retinoblastoma, on the other hand, is a sporadic, randomly occurring form of the disease and so there is only a small risk of the genes being passed on.

SYMPTOMS

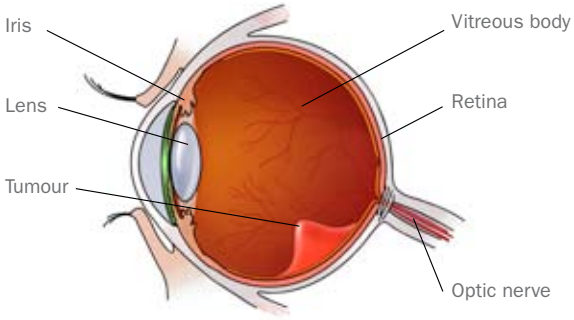
The symptoms of retinoblastoma can be diffuse and difficult to interpret. Most common is an abnormal appearance of the pupil, which is normally black but turns white or yellowish. Some parents may notice that the pupil catches the light from certain angles in dim lighting, while others spot changes in photographs. Squinting may also be a symptom of an eye tumour, but this is a very unusual reason for squinting.

DIAGNOSIS

Children with suspected retinoblastoma are referred by their regional eye clinic to St Erik's Eye Hospital in Stockholm for diagnosis.

TREATMENT

Treatment depends on how far the tumour has spread and whether it is unilateral or bilateral. Surgical removal of the eye with the most widespread tumour growth is common, while any tumours in the other eye can nowadays be treated successfully with systemic chemotherapy



followed by repeated focal laser therapy. In some cases this treatment is complemented with ionising radiation administered using a radioactive applicator which is attached to the surface of the eye for about a day, and, if this is not enough, external radiotherapy of the whole of the back of the eye. New tumours can develop after the treatment, and chemotherapy and focal laser treatment are followed by regular check-ups – monthly to begin with and then gradually less often. These examinations are initially given under anaesthetic, and then in an outpatient clinic without anaesthetic from the age of four or five before normally coming to an end at the age of seven or eight.

PROGNOSIS

A century ago the prognosis for retinoblastoma was quite bleak, but survival has since improved significantly. Today more than 95 per cent of all children in the Western World are disease-free five years from the onset of the disease, and it is now extremely rare for the disease to spread. Vision also remains good with modern treatment, and relatively few children with retinoblastoma suffer from a significant visual handicap. Full vision in one eye is, for example, quite enough to be able to drive a car.

Factual accuracy verified by professor Stefan Seregard from St Erik's Eye Hospital, April 2009