

INTRAOCULAR TUMOURS

Children

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Retinoblastoma, the most common ocular malignancy of childhood, is a malignant tumour of the retina that can be bilateral in around one-third of cases. Half of cases are hereditary (autosomal dominant) and are due to mutation of the gene on chromosome 13; children with a family history should be carefully monitored from birth. Remaining cases occur sporadically. Inherited retinoblastoma is more likely to be bilateral. Retinoblastoma is often not spotted until the tumour fills the globe and presents as a white reflex in the pupil or as a squint (Figure 49.13). The differential diagnosis includes retinopathy of prematurity, persistent fetal vasculature (PFV) and intraocular infections. If the tumour is large, enucleation may be required, but radiotherapy, cryotherapy, chemotherapy or laser treatment can cure small lesions. Liaison with a paediatric oncologist is essential. Summary box 49.2 Intraocular tumours /uni25CF /uni25CF - RB1

Figure 49.12 Capillary haemangioma in a child. An orbital venogram demonstrates displacement of the second part of the superior ophthalmic vein (arrow) (courtesy of Dr Glyn Lloyd). Figure 49.13 Retinoblastoma giving rise to a white pupillary reflex. This child was first seen with a convergent squint and discharged without a fundus examination. He was next seen many years later with a 'white reflex' and died soon after diagnosis (courtesy of MA Bedford, FRCS). Any child with a white pupil (leukokoria) should be referred to an ophthalmologist to exclude retinoblastoma, although congenital cataracts may also cause this sign. A blind painful eye may hide a melanoma or other ocular tumour

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