

malformations, although the vast majority of children with retinoblastomas apparently have normal chromosomes and intelligence.

Clinical Manifestations

Retinoblastoma has few grossly obvious signs. Typically the parents are the ones who first observe a whitish “glow” in the pupil, known as the cat's eye reflex, or leukocoria (Fig. 25-6). The reflex represents visualization of the tumor as the light momentarily falls on the mass. When a tumor arises in the macular region (which is the area directly at the back of the retina when the eye is focused straight ahead), a white reflex may be visible when the tumor is small. It is best observed when a bright light is shining toward the child as the child looks forward. Sometimes parents accidentally discover it when taking a photograph of their child using a flash attachment.



FIG 25-6 Cat's eye reflex. Whitish appearance of lens is produced as light falls on tumor mass in left eye.

When the tumor arises in the periphery of the retina, it must grow to a considerable size before light can strike it sufficiently to produce the cat's eye reflex. In this situation it is visible only when the child looks in certain directions (sideways) or if the observer stands at an oblique angle to the child's face as the child looks straight ahead. The fleeting nature of the reflex often results in a