Cortical visual impairment (CVI) is the most common cause of bilateral visual impairment in children in the developed world. In less-affluent countries, the incidence is increasing because the survival rate of premature babies is improving. As a consequence, the mortality of children with complex medical problems has begun to decline. Retinopathy of prematurity (ROP) is also a major cause of visual handicap: its rate is increasing, and it may become the commonest cause of visual impairment in children. A risk factor for CVI is prematurity, which is also a risk for ROP. Thus, these two disease processes often coexist.

CVI is defined as visual impairment caused by damage to the central nervous system. Visual acuity is reduced as a result of a disease process that does not involve the ocular structures.

HISTORICAL PERSPECTIVE

CVI has come into prominence recently partly because of its increasing incidence and also the greater understanding that is being developed of the pathophysiology. Whiting, Jan, and Wong first coined the term cortical visual impairment in 1985; before then, the problem was referred to as cortical blindness. Cortical blindness is a term more relevant to adults who experience a devastating injury to their occipital cortices. Infants and children who experience such insults tend not to be blind but rather impaired. Their brains are still growing, and as a consequence some aspects of their vision improve over time.
INCIDENCE

Cortical visual impairment is now recognized as the most frequent cause of bilateral visual impairment in the Western world. The Blind Babies Foundation of Northern California has found that, in children under 5 years of age with visual impairment, CVI is the most prominent causative factor.\(^{15}\) The Oxford Registry of Early Childhood Impairments found that nearly 30% of children with bilaterally poor vision had CVI.\(^{16}\) In Liverpool (U.K.), it was found that in children with neurological disorders and visual impairment, CVI was the most common cause of poor vision.\(^{17}\) A study in 1996 in the Nordic countries found that brain damage is causing an increasing number of children to have visual impairment.\(^{18}\)

ETIOLOGY: PATHOPHYSIOLOGY, HISTOPATHOLOGY

The cause of CVI can be diverse: hypoxia-ischemia; congenital brain malformation (schizencephaly, holoprosencephaly, lissencephaly); central nervous system infection (meningitis, encephalitis); blockage of a ventriculo-peritoneal shunt; head injury, particularly that resulting from child abuse; or metabolic derangements.\(^{12}\)

Hypoxia-Ischemia

The most common cause of CVI is an hypoxic-ischemic event. The consequence of an hypoxic-ischemic event can best be assessed by the age at which the insult occurred. At each age level, a different area of the brain is more susceptible to damage in an hypoxic-ischemic event.

In premature babies, the germinal matrix is most at risk of being damaged.\(^{6}\) The germinal matrix is a watershed area of the brain, but only in preterm babies. It is situated in the walls of the lateral ventricles. The optic radiations are supplied with blood from the germinal matrix, as are the long motor tracts. Thus, the preterm baby who suffers CVI is very likely to also have cerebral palsy.\(^{1}\) Periventricular leukomalacia ensues after an hypoxic-ischemic event in preterm babies (Fig. 7-1).

In the term infant, an hypoxic-ischemic insult is more likely to affect the watershed area of the brain, which is now an area