Cortical visual impairment (CVI) is the most common cause of bilateral visual impairment in children in the developed world.\(^1\) 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.\(^1^8\) 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.\(^9\)

**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\(^{1^9}\); 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. The Oxford Registry of Early Childhood Impairments found that nearly 30% of children with bilaterally poor vision had CVI. 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. A study in 1996 in the Nordic countries found that brain damage is causing an increasing number of children to have visual impairment.

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.

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. 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. 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
including the striate cortex [see Fig. 7-2]. In term children, an hypoxic-ischemic insult causing permanent CVI is much less common.

**Shunt Blockage**

Ventriculoperitoneal shunt blockage has been described as a cause of CVI.\(^4\) The occipital lobes become infarcted because the posterior cerebral arteries are compressed against the edge of the tentorium.\(^{13}\)

**Twin Pregnancy**

A risk factor for CVI is twin pregnancy.\(^6\) Twin pregnancy increases the risk of premature birth, which is a risk factor for CVI in itself. There is an even higher chance of prematurity if the twins are monozygotic.\(^2\) Twin–twin transfusion syndrome can occur if the twins are monozygous and also share one pla-
If one twin dies, then there may be an acute twin–twin transfusion of blood to the dead fetus. The surviving fetus may become hypovolemic and develop neurological abnormalities as a consequence. Emboli and thromboplastin from the dead fetus may enter the survivor’s circulation and induce disseminated intravascular coagulation.\(^\text{14}\) The developing brain is then exposed to hypoxia-ischemia.

**CLINICAL FEATURES**

Neurobehavioral characteristics, or mannerisms, exhibited by children with CVI are helpful in deciding the position of a lesion in the visual pathways. These behaviors tend to be adaptations to the anatomic defect.\(^\text{7}\)

**Differential Diagnosis**

The differential diagnoses of CVI are varied, and include conditions in which the child appears to have poor vision but either
improves later (delayed visual maturation); has a poor motor response (oculomotor apraxia); or has no apparent interest in their surroundings, as may be exhibited in autism.\textsuperscript{9}

**CLINICAL ASSESSMENT: WORKUP, EXAMINATION TECHNIQUE, LABORATORY, PATHOLOGY**

The child who presents with CVI can usually be diagnosed with a clinical examination. In pure cases the ocular examination is normal. It should be remembered that ocular and cortical abnormalities can coexist. Children with CVI have poor visual function and do not exhibit eye contact. They also do not regard a face. Parents may comment that sometimes the child sees better than other times; variability in visual performance is a characteristic of CVI.

**SYSTEMIC ASSOCIATIONS**

Any child who is diagnosed with CVI will have an associated neurological abnormality.\textsuperscript{11} Most children with CVI have a co-existent ocular problem.\textsuperscript{19} An example of this is the child who is extremely premature and has a risk of developing retinopathy of prematurity. In this child, the anterior (ROP) and posterior (CVI) visual pathways are involved, causing the visual impairment.

**INHERITANCE**

CVI is usually the result of an insult to the developing brain, often an episode of hypoxia-ischemia. Thus inheritance is not considered to be a major factor in the causation. Nevertheless, some clinical features suggest a partial genetic cause. Some children have a remarkable recovery from a neurological insult, and this invulnerability might be genetically determined.

**NATURAL HISTORY**

CVI improves with time, although full, normal vision is rarely achieved.\textsuperscript{8} It is more usual for gradual improvement to occur over months and years. Visual behavior can change from hour
to hour depending on fatigue or distractibility of the child, so it is important for parents to realize that the best vision which they observe is more indicative of the child’s potential.

Visual improvement after an hypoxic-ischemic event seldom regresses. Parents need not be concerned that the vision will decrease unless there is a progressive neurodegenerative disorder, or unless some other neurological event occurs that interferes with vision (e.g., intractable seizures).

TREATMENT: MEDICAL OR SURGICAL

Surgical: Indication, Technique, and Complications

Surgical treatment is seldom helpful in CVI. Exceptions include a shunt blockage that needs to be relieved or a tumor that requires resection. Cerebral edema or hemorrhage secondary to trauma may require relieving.

Medical: Specific Medication and Dose

Medical treatment for CVI is limited. In the preterm baby, the amount of oxygen delivered needs to be carefully regulated as it could affect other disease processes such as retinopathy of prematurity (ROP). Some children with CVI have epilepsy as a result of their structural brain abnormality. Caregivers sometimes believe that treating the epilepsy or treating the abnormal EEG will help vision, but this is usually not the case. If epileptic seizures are present, then they should be treated, but simply treating the EEG will not improve vision.

PROGNOSIS: OUTCOME OF TREATMENT

The prognosis is dependent on the age at which the insult was sustained and the extent of the insult. Associated neurological abnormalities are also an important factor to be considered. Rehabilitation improves outcome significantly, and it is important that the approach is multidisciplinary as the child rarely has purely an ocular disorder. The preterm infant who receives an hypoxic-ischemic insult tends not to improve as much as the term infant.11
In general, there is visual improvement over time with CVI. However, in prognosticating whether there will be significant improvement, it is important to note the cause of the CVI (some causes have a worse prognosis for improvement than others) and also the age of the child when the insult occurred. Involvement of the optic radiations has a worse prognosis than involvement of the striate cortex. If periventricular leukomalacia is found on neuroimaging, the prognosis is thus poorer, as it suggests optic radiation damage. Visual recovery from CVI caused by bacterial meningitis is known to be poor.

**FUTURE RESEARCH**

Research is focused on assessment of children with CVI as well as treatment. It is difficult to assess what a child with CVI sees. Electrophysiological tests (visual evoked potential) are helpful in indicating different qualities of sight. The use of functional magnetic resonance imaging (fMRI) in diagnosis of CVI is being investigated, but there are major limitations in children because the technique requires the subject to be alert, still, and cooperative.

**References**