SCIENTIFIC REPORT

Cryptic cerebral visual impairment in children

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Aim: To review the medical records of seven children with a delayed diagnosis of cerebral visual impairment.

Methods: The charts of a consecutive series of children examined in a university based ophthalmology clinic with a delayed diagnosis of cerebral visual impairment were reviewed. Their neuroimaging studies were evaluated by a paediatric neuroradiologist.

Results: The seven patients ranged in age from 2 years to 10 years at the time of presentation and had acuities ranging from 20/40 to 20/400. Their visual fields ranged from full visual fields to homonymous hemianopias. Neuroimaging findings ranged from subtle occipital lobe abnormalities to obvious cerebral volume loss.

Conclusions: Cerebral visual impairment can be cryptic in children with mild visual impairment. Neuroimaging studies and visual field testing can help to differentiate this condition from other causes of visual impairment.

Cerebral visual impairment (CVI), also sometimes referred to as cortical visual impairment, is one of the most common causes of blindness in children.^{1–3} It usually occurs secondary to a perinatal insult, and in most cases these children have other neurological problems.⁴ CVI is a term used to describe a visual disorder due to abnormalities in the retrochiasmatic visual pathways. The findings and presentation of patients with CVI can be highly variable, and because of the difficulty in determining visual function in young children, the diagnosis may be missed initially. Complete cortical blindness can be caused by destruction of the optic radiations and/or the striate cortex, but this is rare.⁵ CVI most commonly occurs in children as a result of hypoxic-ischaemic brain injuries, especially during the perinatal period. The gestational age of the child at the time of the insult largely determines the type of injury that will develop.^{6,7} Full term infants usually develop watershed infarcts of the cerebral cortex, whereas premature infants more commonly sustain injuries to the periventricular white matter resulting in periventricular leucomalacia (PVL).⁸ Visual impairment in children with PVL is due to injuries to the optic radiations, whereas visual impairment in full term children is usually secondary to infarctions of the striate cortex.^{9,10} In most cases, children with CVI have other neurological problems that raise the suspicion of CVI; however, in some cases their neurological problems may be fairly subtle and the diagnosis of CVI may not be suspected.¹¹ In addition, some children with CVI have abnormalities of the anterior visual pathways such as optic atrophy, which may further obfuscate the diagnosis of CVI. We report the clinical and neuroimaging findings of seven children in whom the diagnosis of CVI was initially missed by another ophthalmologist.

Patients and methods

After obtaining institutional review board approval, the charts of seven consecutive children in whom the diagnosis

of CVI was initially missed by one or more ophthalmologists were reviewed (table 1).

The children ranged in age from 2 years to 10 years at the time the diagnosis of CVI was established. Six of the seven patients were male. All neuroimaging studies were reviewed by a paediatric neuroradiologist. The patient's vision at presentation was compared with their vision at subsequent follow up examinations.

Case presentations

Patient 1

A 10 year old boy with unexplained decreased vision was referred for evaluation. He was born after a full term pregnancy complicated by sepsis secondary to premature rupture of the amniotic membranes at the time of birth. His best corrected visual acuity (BCVA) was 20/200 in both eyes. His refraction was plano in both eyes. An electroretinogram (ERG) was normal. Pattern reversal visually evoked potentials (VEPs) were highly attenuated in both eyes. A computed tomography (CT) scan (fig 1) revealed old bilateral frontal and occipital lobe infarcts that were probably related to his perinatal sepsis.

Patient 2

A 6 year old boy was referred for electrophysiological testing to rule out a retinal dystrophy. He had been noted to hold objects close to his face since infancy. He had *Haemophilus influenzae* meningitis when 2 months of age, but his subsequent development had been normal. His BCVA was 20/60 in the right eye and 20/100 in the left eye. His cycloplegic refraction was +0.75D, in both eyes. He had slightly constricted visual fields, the right greater than the left. An ERG was normal. A magnetic resonance imaging (MRI) scan revealed porencephalic dilatation of the left occipital horn and pitting of the left occipital cortex.

Patient 3

A 10 year old boy was referred for evaluation of decreased vision. He had an apnoeic episode when 6 weeks of age and subsequently developed a seizure disorder and strabismus. He was treated with seizure medications until 2 years of age, but had been seizure free since then. He underwent strabismus surgery to correct an esotropia when 15 months of age. He had great difficulty keeping up with the other students in his class at school. His BCVA was 20/400 in both eyes. He had a refractive error of +0.50D in each eye. An MRI revealed significant encephalomalacia of the right occipital lobe with mild volume loss and gliosis of the left occipital lobe (fig 2). He was subsequently taught to read using braille which resulted in a dramatic improvement in his school performance. At his last follow up examination when 20 years of age, his vision was 20/400 in the right eye and counting fingers (CF) at 4 feet in the left eye. He had a

Abbreviations: CVI, cerebral visual impairment; ERG, electroretinogram; MRI, magnetic resonance imaging; PVL, periventricular leucomalacia; VEPs, visually evoked potentials

Table 1 Clinical findings

Patient no	Age (years, at time of diagnosis)/sex	BCVA (right eye/left eye) at time of diagnosis	Age (years) at last follow up	Last measured acuity (BCVA)	Visual field findings	Neuroimaging	Suspected cause of CVI
1	10 M	20/200 20/200	10	20/200 20/200	Full	Bilateral frontal and occipital infarcts	Perinatal sepsis
2	7 M	20/60 20/100	9	20/200 20/200	Generalised constriction right eye > left eye Left homonymous hemianopsia	Porencephalic dilatation of the left occipital horn Right occipital lobe and posterior temporal lobe encephalomalacia	<i>Haemophilus influenzae</i> meningitis
3	10 M	20/400 20/400	20	20/400 CF 4 ft	Full	Left occipital encephalomalacia with ex vacuo dilatation of the left occipital horn	Anoxic injury during infancy
4	7 F	20/40-1 20/60-2	15	20/40 20/50-2	Full	Left hemispheric encephalomalacia Absent septum pellucidum and abnormalities of occipital lobes	Unknown
5	2 ½ M	CSM CSM	8	20/40 20/60	Right homonymous hemianopsia	Subtle, bilateral occipital lobe atrophy	Closed head injury at 8 months of age
6	5 M	CSM 20/40-2 20/40-3	7	20/50-2 20/50-1	Full		Hydrocephalus
7	4 M	CSM CSM	15	20/60-2 20/70-2	Full		Unknown

BCVA, best corrected visual acuity.

complete left homonymous hemianopsia using Goldmann perimetry and exotropia. His ocular examination was otherwise normal.

Patient 4

A 7 year old girl was referred for evaluation of decreased vision. She had undergone three previous strabismus surgeries. She was the product of a full term pregnancy and her medical history was unremarkable. Her BCVA was 20/40-1 in the right eye and 20/60-2 in the left eye. Her refraction was plano +1.00 × 075, in the right eye and plano +1.00 × 100 in the left eye. She had an A-pattern exotropia with bilateral superior oblique overaction and dissociated vertical

deviation. Her optic discs and retinas were normal in appearance. Goldmann visual fields were normal in both eyes. An MRI revealed a focal area of encephalomalacia in the left occipital lobe with ex vacuo dilatation of the occipital horn of the left lateral ventricle (fig 3).

Patient 5

A 2 ½ year old boy was evaluated for decreased vision. He had a history of a closed head injury when 8 months of age after falling out of bed. He developed a right hemiparesis and a seizure disorder after the accident. His visual acuity was

Figure 1 Patient 1. Computed tomography, brain, axial, demonstrates chronic, bilateral occipital infarcts with encephalomalacia and ex vacuo dilatation of the occipital horns of both ventricles.

Figure 2 Patient 3. Magnetic resonance imaging, T2, fast spin echo, brain, axial, shows an area of encephalomalacia of the right occipital lobes with evidence of contralateral injury, mild volume loss, and gliosis.

Figure 3 Patient 4. Magnetic resonance imaging, T1, three dimensional, FSPGR, brain, axial, showing focal area of encephalomalacia of the left occipital lobe, with ex vacuo dilatation of the occipital horn of the left lateral ventricle.

central, steady, and maintained in both eyes. He had bilateral overaction of the inferior oblique muscles, possibly secondary to bilateral fourth nerve palsies. His fundi were normal. When 8 years of age, his BCVA was 20/40 in the right eye and 20/60 in the left eye. An MRI revealed extensive left hemispheric encephalomalacia and milder abnormalities in the left parietal and occipital lobes (fig 4). He had a right homonymous hemianoptic defect on Goldmann visual field testing.

Patient 6

A 5 year old boy with a history of spina bifida and hydrocephalus was referred for evaluation of an exotropia. He had previously undergone a ventriculo-peritoneal shunting procedure. His BCVA was 20/40-2, with a refraction of +2.50 sphere in the right eye, and 20/40-3, with a refraction of +2.50 +1.00 $\times$ 030 in the left eye. He had an intermittent left exotropia with mild, bilateral inferior oblique overaction. One year later, his BCVA was 20/60 in each eye. An MRI revealed an absent septum pellucidum but normal optic nerves. There were also multiple dysplastic areas, including both occipital lobes.

Patient 7

A 4 year old boy was referred for evaluation of a large angle exotropia. He also had a complex partial seizure disorder and delayed development. A MRI revealed subtle bilateral occipital lobe abnormalities (fig 5). When 6 years of age, his BCVA was 20/60 in both eyes. When 15 years of age, his BCVA was 20/60-2 in the right eye and 20/70-2 in the left eye. Goldmann visual fields were full in both eyes.

DISCUSSION

Cerebral visual impairment should be considered in the differential diagnosis of any child with unexplained decreased vision. In particular, it should be suspected in a child who is unable to achieve the expected visual acuity based on their ocular examination, while taking into account their mental or verbal abilities. Electrophysiological testing can be helpful in determining which patients may have an

Figure 4 Patient 5. Magnetic resonance imaging, T2, fast spin echo, brain, axial, shows extensive encephalomalacia of the left occipital and temporal lobes

Figure 5 Patient 7. Magnetic resonance imaging, T2, fast spin echo, brain, axial, shows bilateral atrophy and gliosis of the occipital lobes.

additional reason for their decreased vision. Ultimately, a neuroimaging study of the visual pathways is usually required to establish the diagnosis of CVI. Computed tomography may be adequate in patients with severe abnormalities, but MRI with T2, fast spin echo with gadolinium is necessary to diagnosis more subtle abnormalities.¹²

The diagnosis of CVI was probably missed in our patients because four had not undergone any previous neuroimaging studies, and all had relatively minor neurological problems. While two had a history of seizures, the seizures had resolved, and the children were no longer on seizure medication. One child also had hydrocephalus, but his visual disability was so minor that it had not been appreciated. All of the children were ambulatory, and none had cerebral palsy. All of the school aged children were attending schools in their local communities, although some were in special education classes. In contrast, Huo and co-workers⁴ reported that 75% of the children in their series with CVI had other neurological deficits and 50% had seizures. Rogers and co-workers¹³ reported cerebral palsy in 53% of the children in their study with CVI.

A probable aetiology for CVI was established in five of the seven patients. These aetiologies included perinatal sepsis, meningitis, a prolonged episode of apnoea, a closed head injury, and hydrocephalus.^{13–20} None of the children in our series had known hypoxic-ischaemic insults during the perinatal period. Hypoxic-ischaemic insults generally result in more catastrophic brain injuries with profound visual impairment and severe neurological handicaps.¹⁹ Given the relatively subtle visual and neurological problems of our patients, this was not unexpected. In most children, an aetiology for CVI can be elicited with a careful history. In a review of 170 children with CVI, Huo *et al*⁴ found 9.4% to be idiopathic. We could not establish an aetiology for the striate cortex abnormalities in two of the patients in our series.

The visual acuity was preserved much better than expected for the children in our series compared to other series of children with CVI. Six of our seven patients (86%) had a visual acuity of 20/200 or better. In contrast, Hoyt²¹ reported that only 10% of the children in his series with CVI had a visual acuity of 20/200 or better, although this was a much larger series including all patients with CVI, not just those presenting with a difficult diagnosis. CVI generally affects the visual acuity equally in both eyes. The only exception to this rule in our series was patient 3. The more severely reduced visual acuity in this patient's left eye was probably caused by strabismic amblyopia superimposed on his CVI. One patient had a complete homonymous hemianopsia suggesting that the occipital lobe injuries were asymmetrical. Patient 3 had a dramatic improvement in his school performance, once the extent of his visual difficulties was fully appreciated, and he was taught to read using braille. Children with CVI often have profound visual perceptive problems that make it difficult for them to read efficiently, even when their visual acuity would seem to be adequate for reading by sight.¹³

The small number of patients in our series makes description of trends and generalisations impossible, but these cases emphasise that the presentation of patients with CVI may be cryptic, later than expected, and visual function may be quite good. The diagnosis of CVI should be considered in any child in whom there are no ocular findings to account for their decreased vision. A high quality MR scan as well as a careful medical history is critical in establishing the diagnosis of CVI.

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