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Clinical characteristics of children with severe visual impairment but favorable retinal structural outcomes from the Early Treatment for Retinopathy of Prematurity (ETROP) study

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Abstract

Purpose—To describe visual function and associated characteristics at the 6-year examination in children enrolled in the Early Treatment for Retinopathy of Prematurity Study who had unfavorable visual outcomes despite favorable structural outcomes in one or both eyes.

Methods—The clinical examination records of children completing the 6-year follow-up examination were retrospectively reviewed. Eligible subjects were those with visual acuity of ≤20/200 in each eye (where recordable) and a normal fundus or straightening of the temporal retinal vessels with or without macular ectopia in at least one eye. Data regarding visual function, retinal structure, presence of nystagmus, optic atrophy, optic disk cupping, seizures/shunts, and WeeFIM (pediatric functional independence measure) developmental test scores were reviewed.

Results—Of 342 participants who completed the 6-year examination, 39 (11%) met inclusion criteria. Of these, 29 (74%) had normal retinal structure, 18 (46%) had optic atrophy, and 3 (8%) had increased cupping of the optic disk in at least one eye. Latent and/or manifest nystagmus occurred in 30 children (77%). The presence of nystagmus was not related to the presence of optic atrophy. Of the 39 children, 28 (72%) had a below-normal WeeFIM score.

Conclusions—In 25 participants (7%) completing the 6-year examination, cortical visual impairment was considered the primary cause of visual loss. The remainder likely had components

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of both anterior and posterior visual pathway disease. Clinical synthesis of ocular anatomy and visual and neurologic function is required to determine the etiology of poor vision in these children.

Over the last several decades, improved perinatal care has led to a higher survival rate for increasingly younger preterm infants; concurrently, the frequency of complications associated with infants born prematurely has increased. Visual impairment is an important sequela of prematurity, affecting both the anterior (retinopathy of prematurity [ROP], optic atrophy) and the posterior (damage to the primary visual or peristriate cortex, lateral geniculate nuclei) visual pathways.[1–3]

Factors associated with the development of ROP, such as birth weight, gestational age, and ischemia, are also associated with diseases of the posterior visual pathways[1]; intraventricular hemorrhage, post-hemorrhagic ventricular dilatation, periventricular leukomalacia, and hydrocephalus can lead to isolated cerebral damage and posterior visual pathway dysfunction.[4] The resulting effects on the post-geniculate visual pathways may progress over time and are often not identified until the child is older or until after any anterior pathway pathology is treated.

The classic definition of the term *cortical blindness* or *cortical visual impairment* (CVI) refers to bilateral loss of vision with normal pupils and a normal structural eye examination. This designation, however, is incomplete, in that many subcortical white matter insults (eg, periventricular leukomalacia), as well as those involving the lateral geniculate nuclei, can affect visual function; moreover, this definition does not capture additional findings, such as ocular motor apraxia or visual inattention, which may also occur in these patients. Finally, the diagnosis may become obscured when concomitant ocular disease, such as ROP residua or optic atrophy is present, as the clinician must often qualitatively assess the relative contribution of each to the patients' visual deficits.

The terms *cerebral visual impairment*, *post-geniculate visual impairment*, and *posterior visual pathway disease* have been suggested to convey a more complete clinical picture of patients in whom ocular disease is not the sole or primary cause of visual loss; however, because damage to the visual cortex nearly always results when the white matter is damaged, due to lack of appropriate cellular connections and input to the cortex, these terms are frequently used interchangeably.

Siatkowski and colleagues[5] observed posterior visual pathway disease in a subset of prematurely born children enrolled in the Cryotherapy for Retinopathy of Prematurity (CRYOROP) study who had birth weights <1251 g and poor visual outcomes despite normal or almostnormal retinal structure. Of the 247 children completing the 10-year follow-up examination, 16 were identified as having an unfavorable visual outcome (<20/200) in both eyes even though at least one eye had a favorable structural outcome (normal retinal structure, or straightening of the temporal retinal vessels with or without macular ectopia).[5] They concluded that the poor visual outcome was likely the result of post-geniculate disease in 5 patients (31%), whereas the poor visual outcome was likely due to ROP in 6 (38%), and combined anterior and posterior visual pathway disease in 2 (13%).

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There were insufficient data to determine the cause of visual loss in the remaining 3 patients.[5]

The purpose of the present study is to describe visual function abnormalities in children with visual impairment despite relatively normal ocular structure who were enrolled in the Early Treatment for Retinopathy of Prematurity (ETROP) study.

Methods

A detailed description of the ETROP study has been previously published.[6] Institutional review board approval was obtained at each participating institution, and guidelines of the Health Insurance Portability and Accountability Act were followed throughout the course of the study at each site.

The cohort of subjects in this trial was selected from those children who completed the 6-year examination in which best-corrected visual acuity was measured by study-certified vision testers and dilated fundus examination was performed by study-certified ophthalmologists. Recognition visual acuity testing was conducted using ETDRS optotypes, and when not possible, using the low vision Teller acuity card (0.23 cy/cm) or detection of light.

Children with neurological impairment were screened for ability to perform the test by being shown large individual copies of the 10 optotypes at a distance of 1 meter. The child continued through visual acuity testing only if he or she could correctly identify or match 9 of 10 consecutive letters. If this was not possible due to cognitive or developmental reasons, testing with an HOTV chart was attempted. Children who could not perform the HOTV task were coded as failing the screening due to neurological impairment.

For testing of nystagmus, the child was required to be fully awake; to be recorded, manifest nystagmus had to be always or nearly always present without occlusion, while latent nystagmus was diagnosed with occlusion of one eye. Searching or roving eye movements did not qualify as nystagmus.

Optic atrophy was diagnosed as present if the examiner felt that it was definitely present; although examiners could check whether the atrophy was partial or severe, optic atrophy was not formally graded or further characterized in the present study. Classification of all optic nerve and retinal structural data was determined by the independent clinical judgment of the certified study physicians.

Inclusion criteria included poor visual acuity (≤20/200) in both eyes, with a favorable structural outcome (normal fundus or straightening of temporal retinal vessels with or without macular ectopia) in at least one eye. Subjects were excluded if they had undergone vitrectomy or scleral buckling, or had a history of cataract or glaucoma. The following data were extracted from review of examination data: visual acuity and retinal structural appearance in each eye, presence or absence of manifest and/or latent nystagmus, presence or absence of optic atrophy and/or non-glaucomatous cupping >0.5, and WeeFIM developmental test scores.[7] The WeeFIM is designed to measure functional performance in

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self-care, mobility, and cognition in children from age 6 months to 7 years, and to assess the need for assistance and severity of disability; it is, in essence, a pediatric version of a functional assessment outcome evaluation. It does not quantify impairment or correlate disabilities with their etiologies. Although the WeeFIM assesses the overall effects of neuromotor impairment on functional skills, it does not replace formal comprehensive cognitive or neurodevelopmental assessments. While disability is often a result of or associated with global delays in cognitive function, there can be dissonance between the two; for example, a child with a high score on the WeeFIM may have serious neurologic disease (eg, in cerebral palsy), and a child with a low WeeFIM score may be cognitively intact (eg, in spina bifida).[7]

Subjects were classified as having definite versus possible cerebral visual impairment after review of their composite clinical data in all but 2 cases, in whom such a determination was not possible (see below). Neuroimaging was not part of the study protocol, nor was review of any imaging studies obtained by either study-certified or other treating physicians. Details of the clinical thought processes involved in these assignments are discussed below.

Results

At the 6-year examination, 342 of the surviving 370 children (92.4%) were evaluated. Of these, 39 (11%) met our inclusion criteria (see Table 1).

Of these 39 subjects, 25 (64%) had a normal-appearing fundus in each eye; 4 (10%) had a normal fundus in one eye but straightening of the temporal vessels or macular ectopia in the fellow eye. Another 7 (18%) had straightening of the temporal vessels in both eyes. One patient had a stage 4B detachment in one eye and straightening of the temporal vessels in the fellow eye.

Eighteen children (46%) had optic atrophy in one or both eyes. Increased cupping of the disk with an otherwise normal optic nerve and retinal examination was noted in 3 patients (8%) and was present in 9 with optic atrophy (23%).

Nystagmus was present in 30 children (77%): 4 (10%) had manifest nystagmus alone, 6 (15%) had latent nystagmus alone, and 20 (51%) had manifest and latent nystagmus. Sixteen patients presented with nystagmus (latent, manifest, or both) associated with optic atrophy, and/or non-glaucomatous cupping of the disk. Of the children who had nystagmus, 14 also had optic atrophy and 16 did not; of those without nystagmus, 5 had optic atrophy and 4 did not.

Developmental status was assessed via WeeFIM questionnaires administered by certified staff.[7] Of the 39 subjects in the study cohort, 7 (18%) had a normal WeeFIM score (>95); 4 (10%), an intermediate score (77–95), indicating below normal development; and 28 (72%), a score indicating severe disability (<77).Shunts were present in 8 patients (21%) and seizures in 8 (21%).

Our review of the data (Table 1) yielded the following results and clinical inferences . Patients 1 through 25 (group 1) were classified as having definite CVI. Patients 26–37

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(group 2) were felt to have either probable CVI or combined anterior and posterior visual pathway disease.

Although all group 1 subjects had a normal retinal appearance in both eyes, all also had poor visual acuity (≤20/200) bilaterally or visual acuity that could not be assessed due to neurodevelopmental delay (15 subjects [60%]). Each of the 12 subjects in group 2 had either normal retinal appearance or straightening of the vessels without macular ectopia in at least one eye; none had normal fundus findings bilaterally. In group 2, 4 subjects had a normal retinal appearance in one eye; in 2 subjects, the fellow eyes showed straightening of the temporal vessels; in 2, macular ectopia. Of the 12 subjects, 7 (58%) had straightening of the temporal vessels bilaterally. One patient had straightening of temporal vessels in one eye and had a stage 4B retinal detachment in the other.

Neurodevelopment varied but was generally impaired across group 1, indicating global cerebral impairment. Of the 25, 19 (76%) had a WeeFIM score suggestive of severe disability, 3 (12%) had below normal development, and 3 (12%) had low normal developmental scores at 99, 101, and 103. In group 2, neurodevelopmental delay was present in 6 (50%) of the 12 subjects. All but 3 had WeeFIM scores consistent with severe disability, and only 2 had scores in the normal range.

In group 1, 6 subjects (24%) had optic atrophy, 2 (8%) had increased cupping of the disk (>0.5), and 7 (28%) had optic atrophy and increased cupping of the disk. In group 2, optic atrophy, increased cupping of the disk, or manifest nystagmus was noted in 7 subjects (58%). The lack of ophthalmoscopic findings in these patients to explain the severity of the visual loss is consistent with CVI, although a component of anterior pathway disease is possible given the fact that no subjects had completely normal fundus findings (via indirect ophthalmoscopy) bilaterally.

In group 1, 5 (20%) had latent nystagmus alone, 1 (4%) had manifest nystagmus alone, and 14 (56%) had latent and manifest nystagmus. Nystagmus was prevalent within group 2 as well: 1 subject (8%) had latent nystamgus alone, 2 (17%) had manifest nystagmus alone, and 5 (42%) had combined latent and manifest nystagmus.

In group 1, 4 subjects (16%) had seizures and 6 (24%) had shunt surgery, compared to 4 subjects (33%) and 2 subjects (17%), respectively, in group 2.

Patients 38 and 39 were classified into a third group, with an indeterminate cause of visual loss. Both had high-normal developmental WeeFIM scores but vision in one was unable to be assessed due to neurodevelopmental delay, reflecting cases of dissonance between disability and global function as described earlier. One patient had a normal fundus in each eye, while the other had a normal appearing fundus in only one eye. There was no optic atrophy or cupping of the disk in either patient. Manifest nystagmus alone occurred in one patient, and combined manifest and latent nystagmus was noted in the other.

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Discussion

The growing population of surviving preterm infants faces unique issues such as CVI, cerebral palsy, and neuro-developmental delay. The incidence of CVI is strikingly increased in preterm infants (19/1000) compared to those born at term (2/1000).[8] CVI can arise from a variety of insults, such as vascular dysplasia, hypoxia, prematurity, and hydrocephalus, or from structural central nervous system abnormalities.[9]

Rudanko and colleagues[1] reviewed long-term data from 125 premature infants born from 1972 to 1989. Overall, 46% of the preterm infants had ROP. When segregated by birth weight, ROP occurred in 97% of infants with a birth weight <1000 g, 56% with birth weight 1000–1499 g, and 11% with birth weight >1500 g. They also reported that the prevalence of optic atrophy in preterm infants increased with greater birth weight. Overall, 28% of the preterm infants had optic atrophy. When segregated by birth weight, optic atrophy was present in 3% of preterm infants with birth weight <1000 g, 19% with birth weight 1000–1499 g, and 47% with birth weight >1500 g. The occurrence of CVI was not correlated with birth weight strata, although it was not present in any infant with birth weight <1000g; overall, 12% of all preterm infants had CVI, compared to 14% of preterm infants with birth weight 1000–1499 g, and 18% with birth weight >1500 g.[1] Because our diagnostic methods for ROP are similar today to those of the 1970s and 1980s (indirect ophthalmoscopy), these ocular structural data parallel what we continue to observe clinically; however, given the diagnostic advances in neuroimaging and neurodevelopmental assessment since that time, their data concerning CVI may not be as accurate as current assessments.

We sought to identify the cause of visual loss based on a clinical synthesis of fundus appearance, presence of nystagmus, and neurological development. Study protocol did not specify any predetermined guidelines for classifying the etiology of poor vision as anterior versus posterior pathway disease, and, as mentioned earlier, neuroimaging was not part of the study protocol. Although imaging may be helpful, there is often a poor correlation between brain anatomy and visual function, and the final diagnosis of cortical visual impairment remains one determined clinically after consideration of all available structural and functional data.

Although optic atrophy is not a specific finding for subcortical/cortical visual loss and is often present in ROP and other retinal disease, it may occur in association with hypoxic prenatal brain damage or damage to the lateral geniculate nucleus secondary to transsynaptic degeneration of fibers along the optic tract.[5, 10] Similarly, degeneration of the posterior visual pathways has been described in preterm infants with periventricular leukomalacia (PVL), and Fazzi and colleagues[11] observed quantitative volumetric reduction of the temporo-parietooccipital region in these infants. An associated ophthalmologic finding of PVL is nonglaucomatous increased cupping of the optic disk due to damage to the optic radiations and degeneration of axons anterior to the insult.[5]

Nystagmus has been reported to differentiate anterior or combined anterior-posterior visual pathway disease from isolated posterior pathway disease. Previously, nystagmus in children with poor visual acuity was thought to indicate anterior visual pathway disease because a

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functioning geniculostriate cortex was considered necessary to generate nystagmus.[5, 9] However, many patients with PVL also have nystagmus, which may indicate its presence in combined or isolated posterior visual pathway disease. Ricci[4] reported that the prevalence of nystagmus was 40% in infants with post-hemorrhagic ventricular dilatation. Khetpal and colleagues[9] stated that PVL might reduce the threshold for development of nystagmus. Hoyt[12] showed that in patients with post-geniculate visual loss of cortical origin versus those with PVL, the incidence of nystagmus was significantly higher in the PVL group (50%) than the cortical group (4.8%). Additionally, nystagmus may be present in children with no evidence of either anterior or posterior visual pathway dysfunction (previously referred to as “motor” nystagmus). Hence nystagmus as a surrogate for anterior visual pathway disease is not sufficiently reliable to be regarded as a sole, or even a primary, marker for the localization of visual deficits.

Our data were collected from a small subpopulation within the ETROP study; nevertheless, we feel that the following conclusions are justified. First, the prevalence of postgeniculate disease as the sole cause or main contributing factor of visual impairment in patients with a favorable retinal structural outcome from ROP is rare and occurred with high clinical certainty in only 25 of the 342 participants completing the 6-year examination (7%). Even when considering the patients in whom CVI is likely or possible, the combined prevalence in this cohort at age 6 years was still no more than 11%. One may be concerned that subjects with bilateral retinal vascular straightening had concurrent maculopathy that limits visual potential; however, none of these patients in our cohort had macular ectopia, and it is rare in our clinical experience for such children to have visual acuity of <20/200 on a retinal basis. If, however, one excludes the subjects in group 2 with retinal vessel straightening but no macular ectopia in each eye, the maximal prevalence of CVI decreases to only 9%. Regardless, this represents an increase from what was seen in 10-year-olds in the CRYO-ROP study, where similar figures were in the 2%–3% range.[5] The reasons for this difference remain unclear, but increased survival of younger infants may account for part of this change.

Second, although manifest nystagmus is common in anterior visual pathway disease, it may also occur in infants with PVL and related posterior visual pathway involvement. Within this study, the presence of manifest and/or latent nystagmus was actually higher in patients with definite CVI (80%) than those with probable CVI (67%). Study design and data collection do not allow interpretation of whether the nystagmus in patients with both latent and manifest nystagmus was a combination of latent and idiopathic infantile nystagmus versus manifest latent nystagmus alone; however, manifest nystagmus alone was less common in children considered to have definite CVI (4%) versus those with possible CVI or combined anterior-posterior pathway disease (17%). Thus, while helpful in the overall assessment of the child, the presence or absence of manifest nystagmus alone cannot be regarded as a reliable univariate marker to localize the etiology of visual loss.

Third, due to the plasticity of the developing central nervous system and the variation in damage due to CVI, the lack of optic atrophy and non-glaucomatous increased cupping of the disk does not rule out posterior pathway involvement in the setting of reduced visual acuity. Optic atrophy and non-glaucomatous optic nerve cupping were absent in only 40%

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of subjects felt to have definite CVI, most of whom had WeeFIM scores consistent with severe disability. When evaluating an infant for possible posterior pathway involvement, the examiner must consider neurological status and developmental status as well as visual function. The presence of neurodevelopmental delay, shunts, and/or seizures may give credence to the diagnosis of posterior pathway involvement regardless of the presence or absence of optic atrophy or increased cupping. Obviously, results of neuroimaging would be critical in this evaluation, but these data were not available as part of the trial.

In conclusion, although limited by study size and evaluation of visual function due to neuro-developmental delay, this report will be useful to clinicians and parents alike when evaluating premature children with bilateral visual loss, and provides further insight into the cause of long-term visual loss in children who have structurally favorable retinal outcomes following ROP.

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Table 1

Clinical characteristics of subjects

Subject	WeeFIM score [a]	Summary diagnosis[b]	Visual acuity (OD, OS)[c]	Shunt	Seizure	Optic nerve appearance [d]	Nystagmus	
							Latent	Manifest
Group 1: patients with definite CVI								
1	29	1,1	ND	Yes	No	Normal	No	Yes
2	37	1,1	ND	No	No	Normal	No	No
3	53	1,1	20/252, 20/320	Yes	No	ONA, C/D >0.5	Yes	No
4	23	1,1	ND	Yes	No	ONA	Yes	No
5	48	1,1	ND	No	No	Normal	No	No
6	99	1,1	20/200, 20/1280	No	No	ONA	Yes	Yes
7	58	1,1	ND	No	No	ONA, C/D >0.5	Yes	Yes
8	44	1,1	20/252, 20/400	No	No	ONA	Yes	Yes
9	20	1,1	ND	No	Yes	ONA	No	No
10	25	1,1	LP	Yes	Yes	Normal	Yes	Yes
11	64	1,1	20/640, 20/252	No	Yes	ONA, C/D >0.5	Yes	Yes
12	88	1,1	20/400, 20/200	No	No	Normal	Yes	No
13	50	1,1	ND	No	No	Normal	Yes	Yes
14	103	1,1	20/200, 20/252	No	No	Normal	Yes	Yes
15	101	1,1	20/252, 20/252	Yes	No	C/D >0.5	Yes	Yes
16	27	1,1	ND	No	No	Normal	Yes	Yes
17	29	1,1	ND	No	No	ONA	Yes	Yes
18	27	1,1	ND	No	Yes	ONA, C/D >0.5	No	No
19	18	1,1	LV	No	No	ONA, C/D >0.5	Yes	Yes
20	47	1,1	20/500, 20/640	No	No	Normal	Yes	No
21	50	1,1	ND	Yes	No	ONA, C/D >0.5	Yes	Yes
22	26	1,1	ND	No	No	ONA, C/D >0.5	Yes	Yes
23	90	1,1	ND	No	No	C/D >0.5	No	No
24	18	1,1	ND	No	No	ONA	Yes	No

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Subject	WeeFIM score [a]	Summary diagnosis[b]	Visual acuity (OD, OS)[c]	Shunt	Seizure	Optic nerve appearance [d]	Nystagmus	
							Latent	Manifest
25	93	1,1	ND	No	No	Normal	Yes	Yes
Group 2: patients with probable CVI vs combined anterior and posterior pathway disease								
26	100	3,1	20/800, 20/400	No	No	Normal	Yes	Yes
27	32	2,2	ND	No	No	Normal	Yes	No
28	24	2,2	ND	No	Yes	Normal	Yes	Yes
29	55	2,2	ND	No	No	Normal	No	Yes
30	51	1,3	LV	No	Yes	Normal	No	No
31	38	2,2	LV	No	Yes	ONA, C/D >0.5	Yes	Yes
32	91	2,2	20/200, 20/800	No	No	ONA, C/D >0.5	Yes	Yes
33	19	2,2	LV	No	No	ONA	No	No
34	34	1,2	ND	Yes	No	Normal	No	No
35	64	1,2	ND	No	No	C/D >0.5	No	Yes
36	103	2,2	ND	Yes	Yes	ONA	No	No
37	71	4B,2	LP, 20/252	No	No	ONA	Yes	Yes
Group 3: patients with indeterminate cause of visual loss								
38	123	1/5B	20/200, LP	No	No	Normal	Yes	Yes
39	111	1,1	ND	No	No	Normal	No	Yes

C/D, cup/disc ratio; *CVI*, cortical visual impairment; *LP*, light perception; *LV*, low vision Teller card; *ND*, neuro-developmental delay; *OD:* right eye; *ONA*, optic nerve atrophy; *OS:* left eye; *WeeFIM*, pediatric functional independence measure.

[a] WeeFIM scores (out of maximum of 126): >95, normal; 77-95, below normal; <77, severe disability.

[b] Summary diagnosis (retinal appearance for right eye, left eye):*1*, normal-appearing fundus; *2*, straightening of temporal vessels; *3*, macular ectopia, 4B, incomplete retinal detachment involving the macula 5B, total retinal detachment.

[c] C/D >0.5 refers to nonglaucomatous cupping of optic nerve.

[d] Neuro-developmental delay prevented visual acuity assessment.