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Preoperative visual acuity and contrast sensitivity in children with small, partial, or non-central cataracts

Eileen E. Birch, PhD^{a,b}, Vidhya Subramanian, PhD^a, Christina Cheng Patel, BS^a, and David Stager Jr, MD^{b,c}

^aRetina Foundation of the Southwest, Dallas, Texas

^bDepartment of Ophthalmology, UT Southwestern Medical Center, Dallas, Texas

^cPediatric Ophthalmology & Adult Strabismus, Plano, Texas

Abstract

Background—While there is clear evidence that dense central cataracts >3 mm diameter warrant prompt intervention to prevent deprivation amblyopia, guidelines for management of small, partial and non-central cataracts are not evidence-based. Consensus guidelines advise managing affected infants and preschool children by monitoring for possible progression in size or density and pharmacologic pupillary dilation, if indicated. Extraction is deemed necessary if the cataract progresses sufficiently to obscure the visual axis or if strabismus or nystagmus develops. We evaluated the long-term visual acuity outcome of the consensus guidelines and whether sensory testing during infancy and early childhood may be helpful in guiding treatment.

Methods—A total of 40 consecutive children diagnosed with small, partial, or non-central cataracts at 1 week to 2.5 years of age were enrolled in a prospective study of forced-choice preferential looking visual acuity and contrast sensitivity development. Long-term visual acuity outcome was assessed by crowded HOTV or ETDRS at 4–11 years.

Results—Of the 40 children managed by consensus guidelines, 24 (60%) had abnormal visual acuity at 4–11 years of age, including 9 (23%) who had moderate to severe visual impairment (20/80 or worse) in one or both eyes. Abnormal visual acuity and contrast sensitivity during development were predictive of abnormal long-term visual acuity outcomes.

Conclusions—Visual acuity and contrast sensitivity tests were sensitive to the effects of visual deprivation on the developing visual system and may be useful in early identification of children with small, partial, or non-central cataracts who could benefit from cataract extraction.

Congenital and infantile cataracts interfere with the normal visual experience essential for development of central visual pathways and vision. Visual deprivation during development results in profound anatomical and physiological changes in the primary visual cortex and results in visual impairment.¹ The severity of visual impairment depends on age at the onset of visual deprivation and its duration; early (infantile) onset and prolonged deprivation are associated with profound and irreversible visual impairment.^{2–9}

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Correspondence: Eileen E. Birch, PhD, Retina Foundation of the Southwest, 9600 North Central Expressway, Dallas, TX 75231 (ebirch@retinafoundation.org).

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To prevent or minimize the effects of visual deprivation on the developing visual system, the current standard of care is to extract cataracts that are visually significant within weeks or days of the initial diagnosis.^{10–12} Currently the consensus is that dense central or posterior lens opacities ≥ 3 mm in diameter are visually significant, and evidence-based guidelines support the benefit of prompt extraction, optical correction and, if indicated, amblyopia therapy.^{10–12} On the other hand, lens opacities <3 mm in diameter and opacities with clear areas (small, partial, non-central) such that they do not obscure the visual axis are not considered visually significant.^{10–12} Current American Academy of Ophthalmology (AAO) Preferred Practice Pattern guidelines advise managing affected infants and preschool children with observation to monitor possible progression in size or density and pharmacological pupillary dilation, if indicated.¹⁰ There are no evidence-based guidelines for cataracts that are initially considered to be not visually significant. There is general agreement that extraction is needed if the cataract progresses sufficiently to obscure the visual axis, if strabismus or nystagmus develops, or if there is a loss of visual acuity.^{10–12} In practice, most affected infants and preschool children cannot cooperate with standard visual acuity testing and most pediatric ophthalmology practices do not perform eye movement testing to monitor the onset of nystagmus, so progression of cataract size and/or density and onset of strabismus are the primary indications for cataract extraction in this age range.^{10–12}

One aim of the current study was to determine the long-term visual acuity outcomes of infants and preschool children with small or partial lens opacities (not considered visually significant) who were managed according to the American Academy of Ophthalmology Preferred Practice Pattern.¹⁰ A second aim was to determine whether sensory testing during infancy and early childhood provided information that might be useful in guiding treatment decisions.

Subjects and Methods

A total of 40 consecutive children diagnosed with small, partial, or non-central cataracts at 1 week to 2.5 years of age were enrolled in a prospective study of visual acuity development. A fellowship-trained pediatric ophthalmologist diagnosed all children. All children were otherwise healthy and developmentally normal. Children who experienced sight-threatening intraoperative, perioperative, or postoperative (eg, endophthalmitis, retinal detachment, glaucoma) adverse events were excluded from the study.

Informed consent was obtained from a parent before the child was enrolled in the study. The research protocol observed the tenets of the Declaration of Helsinki and was approved by the Institutional Review Board of UT Southwestern Medical Center and conformed to the requirements of the US Health Insurance Portability and Accountability Act of 1996. Written permission to obtain medical records from the referring pediatric ophthalmologist was obtained annually, and information about diagnosis, ocular alignment, cycloplegic refraction, sensory test results, treatment, and surgery was extracted for each visit. The study was conducted at Retina Foundation of the Southwest. With one exception (details provided below and Figure 1C), Teller Acuity Cards (Stereo Optical Corporation Inc, Chicago, IL) and contrast sensitivity card data were collected for research purposes only and did not contribute to clinical management decisions.

Children attended regular visits at 3–6 month intervals until they were at least 4 years old and able to cooperate with standard visual acuity testing. All sensory testing was monocular, using an opaque patch or a translucent occluder (for children with clinically apparent latent nystagmus), with best optical correction in place. For children with unilateral cataracts, data from the affected eye was analyzed. For children with bilateral cataracts, data from the right eye was analyzed.

Long-term Visual Acuity Outcome

Either the EVA-HOTV and ATS testing protocol (for children 4–6 years of age) or E-ETDRS (for children > 7 years of age) developed by the Pediatric Eye Disease Investigator Group^{13–15} was used to assess visual acuity on the most recent visit. Visual acuity outcome was categorized using the World Health Organization visual impairment definitions in the International Classification of Diseases as follows:¹⁶ normal vision (20/15–20/30), mild visual impairment (WHO category 1: 20/40–20/60), moderate visual impairment (WHO category 2: 20/80–20/200), or severe visual impairment (WHO categories 3–4: 20/400–CF).

Infant and Preschool Visual Acuity and Contrast Sensitivity

In a convenience sample of half of the cohort ($n = 20$), Teller Acuity Cards and contrast sensitivity cards were used to evaluate sensory development during infancy and preschool years, using standard methods.^{17–22} Test results were classified as normal if they fell within 95% tolerance limits for age, borderline if they fell outside the 95% tolerance limit but within the 99% tolerance limit, and abnormal if they fell outside of the 99% tolerance limit^{18,21,22}

Results

Long-term Visual Acuity Outcome

Clinical characteristics, age at first visit and outcome visit, and visual acuity outcome for all 40 participants are presented in Table 1. Cataract types included anterior polar ($n = 4$), anterior polar with persistent pupillary membrane ($n = 4$), cortical ($n = 7$), posterior lentiglobus ($n = 13$), posterior subcapsular ($n = 10$), sutural ($n = 1$), and lamellar ($n = 1$). Cataracts were unilateral in 22 children and bilateral in 18 children. Long-term visual acuity outcome for the affected eyes are summarized in Table 2. The mean ($\pm$ SD) age at the time of outcome visual acuity testing was 6.8 ± 2.4 years. Overall, 24 (60 %) had an abnormal visual acuity outcome in affected eye(s), including 9 (23%) that were classified as moderate or severe visual impairment (6 with unilateral and 3 with bilateral moderate or severe visual impairment). The prevalence of moderate or severe impairment among children with unilateral versus bilateral cataracts was not significantly different (27% vs 17%) $z = 0.80$; $P = 0.42$).

Among the 40 children enrolled, 26 (65%) had surgery prior to the outcome visit: 10 during infancy (< 12 months of age), 14 as preschoolers (1–4 years of age), and 2 at school age (6–11 years of age). In all but 2 cases, the decision to proceed with cataract extraction was made on the basis of cataract progression in size and/or density or the development of strabismus or clinically detectable nystagmus; that is, without visual acuity assessment. Only in the 2 school-age children was visual acuity noted in the chart as a factor contributing to the decision to proceed with surgery.

Infant and Preschool Visual Acuity Development

Figure 1 provides three examples of the wide range of patterns of visual acuity development observed among the 20 children, some with cataracts that remained stable and others with cataracts that progressed who underwent extraction. Figure 1A contains visual acuity data from a child who failed pediatric red reflex screening at 14 months of age; unilateral posterior lentiglobus was diagnosed by a pediatric ophthalmologist at 15 months. On his initial Teller Acuity Card test at 16 months, visual acuities of the affected and fellow eyes were normal for his age and visual acuities remained normal throughout follow-up. HOTV visual acuity at 48 months was 20/25 for the affected eye and 20/20 for the fellow eye, both normal for his age. This pattern of normal visual development was present in 8 of 20 children (40%) studied.

The visual acuity data in Figure 1B is from a girl who was first noted to have a posterior subcapsular cataract when she was 24 months of age. Like the first example, visual acuity was normal on her initial test (24 months), but visual acuity of the affected eye began to lag behind the fellow eye during follow-up. By 48 months, the affected eye visual acuity was borderline and, by 60 months, was abnormal. She has had no surgery. At 6 years, visual acuity in the affected eye was 20/40, indicating mild visual impairment was present. Among the 20 children studied, 9 (45%) were similar to this child, with normal visual acuity but abnormal visual acuity at later visits.

The final example is from a girl who was noted to have a congenital posterior lens opacity when examined by a pediatric ophthalmologist at 5 days of age following a failed neonatal red reflex screening by her pediatrician (Figure 1C). The pediatric ophthalmologist initially managed her with pupillary dilation from 3 weeks of age to 4 months. At 4 months, she was referred to us for Teller Acuity Card testing and was found to have a severe visual acuity deficit in the affected eye. This was the single case in the series of 20 children in which visual acuity card test results influenced clinical care. The patient had cataract extraction surgery 2 weeks after the initial visual acuity test and recovered some visual acuity over the next 6 months, but visual acuity of the affected eye only achieved 20/400 at 63 months (5 years) and 78 months (6 years). This pattern of early and persistent visual impairment was present in 3 of 20 children (15%) studied.

Infant and Preschool Contrast Sensitivity Development

Figure 2A shows the contrast sensitivity data from affected eye at the initial (16 months) and outcome (4 years) visits from the child whose visual acuity data are shown in Figure 1A. As expected for a 16-month-old child, his initial visit contrast sensitivity was above the 12-month norm for most spatial frequencies and the high spatial frequency cutoff was also slightly above the 12-month norm. At 4 years, contrast sensitivity at all spatial frequencies and the high spatial frequency cutoff cut-off were normal for his age.

Figure 2B shows the contrast sensitivity data from the affected eye at the initial (24 months) and 4 follow-up visits from the child whose visual acuity data are presented in Figure 1B. Her contrast sensitivity was normal for most spatial frequencies on all visits and, on the initial 24-month and the 27-month visits, her high spatial frequency cutoff was also normal for her age. However, at the later visits, she began to lose sensitivity at 6 cycles per degree. Note also that the high spatial frequency cut-off failed to improve with age and falls below normal at these later visits, so that it was well below normal by 48 months.

Figure 2C shows the contrast sensitivity data from affected eye at the initial (3 months) and the outcome (4 years) visits from the child whose visual acuity data are shown in Figure 1C. Consistent with the severely reduced visual acuity at 3 and 48 months, contrast sensitivity was also severely reduced at both time points.

Predictive value of Early Visual Acuity and Contrast Sensitivity Testing

Most children (17/20; 85%) had normal visual acuity and contrast sensitivity on the initial visit. Visual acuity and contrast sensitivity developed normally in both eyes of 8 children; visual acuity of all affected eye(s) at the outcome visit was normal for age, ranging from 20/15 to 20/30. The other 9 children who initially had normal visual acuity failed to keep pace with normal development during follow-up: during follow-up abnormal visual acuity or borderline visual acuity and abnormal contrast sensitivity was documented. Of the 9 children, 8 subsequently had cataract extractions and postoperative refractive and amblyopia treatment. All but 1 had abnormal visual acuity when the affected eye(s) were tested at the

outcome visit, ranging from 20/40 to 20/200. One child who had surgery had normal visual acuity in both eyes at the final visit.

Three children (15%) had abnormal visual acuity and contrast sensitivity on their initial visit. All 3 subsequently had cataract extractions and postoperative refractive and amblyopia treatment. Nonetheless, all 3 had moderate to severe visual impairment of the affected eye(s) at the outcome visit, with visual acuity ranging from 20/80 to 20/400.

Overall, 8 of 8 children who had normal visual acuity and contrast sensitivity development had normal long-term visual acuity outcomes; 11 of 12 children who had abnormal visual acuity or borderline visual acuity and abnormal contrast sensitivity had abnormal long-term visual acuity outcomes (Fisher exact test: $P = 0.00007$). Among the 11 who had subsequently had cataract extractions, abnormal visual acuity or borderline visual acuity and abnormal contrast sensitivity was detected 0.2–2.3 years prior to the clinical decision to proceed with surgery.

Discussion

Current Preferred Practice Pattern guidelines¹⁰ are not sufficient to prevent visual deprivation and long-term visual impairment in infants and preschool children with small, partial, or non-central cataracts. Among our cohort of 40 children managed with these AAO guidelines, 60% had abnormal visual acuity outcomes at a mean age of 6.8 years, including 23% who had moderate to severe visual impairment (visual acuity 20/80) in one or both eyes. Abnormal visual acuity and contrast sensitivity testing during infancy and preschool years was associated with poor long-term visual acuity outcome. A number of prior studies have shown poor predictive value of visual acuity testing during the first years of life and long-term visual acuity outcome for a variety of pediatric ophthalmic conditions,^{23–25} and in cohorts with high risk for visual deficits due to very low birth weight.^{26,27} On the other hand, the predictive value of early visual acuity testing was reported to be excellent for children at risk for deprivation amblyopia due to congenital cataracts; visual acuity at 2 years of age was highly correlated with long-term visual acuity outcome at 5–8 years of age ($r = 0.88$, $P < 0.004$).⁵

Visual acuity and contrast sensitivity tests were sensitive to the effects of visual deprivation on the developing visual system and may be useful in early identification of children with small, partial, or non-central cataracts who could benefit from cataract extraction. However, additional data are needed to determine whether the availability of early visual acuity and contrast sensitivity testing results in better long-term visual acuity outcomes.

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FIG 1.

Teller card visual acuity as a function of age for 3 children with cataracts that were initially classified as visually not significant. A, A boy who failed the red reflex examination at a routine 15-month well-child pediatric visit and found to have unilateral posterior lentiglobus at 16 months of age. B, A girl whose mother noted a difference between her eyes and found to have a posterior subcapsular cataract at 24 months of age. C, A girl noted to have a congenital posterior lens opacity at 5 days old following a failed neonatal red reflex screening; initially managed with pupillary dilation, the cataract was extracted when she was 4 months old. Data from affected eyes are plotted as solid squares; data from fellow eyes are plotted with white squares. Circles show visual acuity obtained with EVA-HOTV at 3–6 years of age and/or E-ETDRS at 7 years of age. Mean normal visual acuity is shown as the gray points and line; on this log-log scale, normal visual acuity development is approximately linear. Also shown is the 95% tolerance range for normal visual acuity at each age²² as the pale gray area.

FIG 2. Contrast sensitivity as a function of spatial frequency for the children in Figure 1 (only data from affected eyes are shown). For 12-month-olds and for 4-year-olds 95% tolerance ranges for normal contrast sensitivity^{18,21} are shown as the pale gray areas.

Table 1

Clinical characteristics and visual acuity outcome

Laterality ^a	Type ^a	Age at diagnosis, years	Initial therapy	Progression	Age at surgery, years	Optical correction ^b		Other potentially sight-threatening conditions	Visual acuity outcome		
						Initial	Secondary		Age, years	Fellow eye, logMAR	Outcome, logMAR
U	AP	2.1	Monitor	no	NA	G	NA	spherical anisometropia ^c	6.1	0.0	0.0
U	AP+PM	0.4	Monitor	no	NA	NA	NA	NA	9.4	0.0	0.0
U	PSC	2.3	Monitor	no	NA	NA	NA	NA	4.6	0.1	0.1
U	PL	0.9	Monitor	no	NA	NA	NA	NA	4.5	0.1	0.1
U	PL	1.3	Monitor	no	NA	NA	NA	NA	4.0	0.0	0.1
U	AP	0.1	Monitor	no	NA	NA	NA	NA	5.7	0.2	0.1
U	PSC	2.1	Monitor	no	NA	G	NA	spherical anisometropia ^c	8.3	0.0	0.2
U	AP+PM	0.2	Monitor	no	NA	NA	NA	NA	4.6	0.2	0.2
U	PSC	0.2	pupillary dilation	yes	2.5	IOL	NA	NA	4.3	0.0	0.2
U	AP+PM	0.1	Monitor	no	NA	G	NA	aniso-astigmatism ^c	6.5	0.1	0.2
U	PL	1.0	Monitor	yes	1.3	IOL	NA	NA	6.8	0.1	0.3
U	PSC	2.0	Monitor	no	NA	NA	NA	NA	6.5	-0.1	0.3
U	PL	0.9	Monitor	yes	1.4	IOL	NA	NA	5.0	0.1	0.3
U	PSC	2.8	Pupillary dilation	Yes	3.5	IOL	NA	NA	7.3	-0.1	0.4
U	PL	3.0	Monitor	yes	4.3	IOL	NA	NA	11.8	0.1	0.4
U	PL	0.6	Monitor	yes	0.8	CL	IOL	NA	6.7	0.1	0.4
U	PL	0.2	Monitor	yes	0.4	IOL	NA	NA	9.6	-0.1	0.6
U	PL	0.1	Monitor	yes	2.5	IOL	NA	NA	4.8	0.2	0.6
U	PL	1.0	Monitor	yes	1.5	CL	IOL	NA	6.1	0.0	0.6
U	PSC	1.2	Monitor	yes	1.4	IOL	NA	NA	4.0	-0.1	0.7
U	PL	0.7	Monitor	yes	1.3	IOL	NA	NA	4.8	0.0	1.0
	PSC	0.0	Pupillary dilation	Yes	0.3	CL	IOL	NA	6.5	0.1	1.3
B	C	1.8	Monitor	yes; LE more rapidly	3.4/2.5	G	NA/NA	NA/NA	10.5	0.2	-0.1
B	AP+PM	0.3	Monitor	no; improved by 7 m	NA/NA	NA/NA	NA/NA	NA/NA	5.8	0.0	0.0
B	PL	0.3	Monitor	Yes	0.6/0.6	CL/CL	IOL/IOL	NA/NA	10.1	0.1	0.1

Laterality ^a	Type ^a	Age at diagnosis, years	Initial therapy	Progression	Age at surgery, years	Optical correction ^b		Other potentially sight-threatening conditions	Visual acuity outcome		
						Initial	Secondary		Age, years	Fellow eye, logMAR	Outcome, logMAR
B	AP	0.1	Monitor	No	NA/NA	NA/NA	NA/NA	NA/NA	4.8	0.1	0.1
B	L	0.2	Monitor	No	NA/NA	NA/NA	NA/NA	NA/NA	4.3	0.2	0.1
B	PL	0.2	Monitor	Yes	0.3/0.3	CL/CL	IOL/IOL	NA/NA	4.5	0.6	0.2
B	S	1.0	Monitor	No	1.3/1.3	CL/CL	NA/NA	NA/NA	5.8	0.3	0.3
B	PSC	0.7	Monitor	yes; RE only	0.9/NA	CL/NA	NA/NA	NA/NA	4.5	0.3	0.3
B	C	2.8	Monitor	yes	3.5	IOL/IOL	NA/NA	NA/NA	11.1	0.4	0.3
B	C	2.5	Monitor	yes	4.4/4.5	IOL/IOL	NA/NA	NA/NA	5.9	0.2	0.3
B	PL	0.3	Monitor	yes	0.4/0.4	CL/CL	NA/NA	NA/NA	12.7	0.3	0.4
B	AP	0.1	Monitor	no	NA/NA	NA/NA	NA/NA	NA/NA	4.6	0.3	0.4
B	C	1.7	Monitor	yes	2.2/2.2	CL/CL	NA/NA	NA/NA	8.2	0.5	0.4
B	PSC	2.5	Monitor	yes	7.1/7.1	IOL/IOL	NA/NA	NA/NA	7.7	0.4	0.5
B	C	0.3	Monitor	yes	0.6/0.6	CL/CL	IOL/IOL	NA/NA	9.1	0.5	0.5
B	PSC	0.2	Monitor	yes	0.4/0.4	CL/CL	NA/NA	NA/NA	6.9	0.6	0.6
B	C	1.25	Monitor	yes	7.5/7.5	G/G	NA/NA	NA/NA	10.8	0.7	0.6
B	C	0.5	Monitor	yes	0.8/0.8	G/G	NA/NA	NA/NA	5.1	0.7	0.8

B, bilateral; LE, left eye; NA, not applicable; RE, right eye; U, unilateral.

^a AP, anterior polar; C, cortical; L, lamellar; PL, posterior lentiglobus; PM, pupillary membrane; PSC, posterior subcapsular; S, sutural.

^b CL, aphakic contact lens; IOL, intraocular lens; G, glasses.

^c Treated with glasses.

Table 2

Long-term visual acuity outcome of children whose cataracts were initially considered not visually significant

Outcome visual acuity ^a	Definition	N (%)
Normal vision	20/15–20/30	16 (40)
Mild visual impairment	WHO Category 1 (20/40–20/60)	15 (37)
Moderate visual impairment	WHO Category 2 (20/80–20/200)	8 (20)
Severe visual impairment	WHO Category 3 and 4 (20/400-CF)	1 (3)

CF, counting fingers.

^aVisual acuity outcome category as defined by WHO International Statistical Classification of Diseases.¹⁶