Published in final edited form as:

J AAPOS. 2012 April ; 16(2): 182–184. doi:10.1016/j.jaapos.2011.12.156.

The prevalence of ocular structural disorders and nystagmus among preschool-aged children

Michael X. Repka, MD, MBA^{a,b}, David S. Friedman, MD, PhD^{c,d}, Joanne Katz, PhD^{c,d}, Josephine Ibironke, OD, MPH^a, Lydia Giordano, OD^c, and James M Tielsch, PhD^{c,d}

^aZanvyl Krieger Children's Eye Center and Adult Strabismus Service, Wilmer Eye Institute, Johns Hopkins University School of Medicine, Baltimore, Maryland ^bDepartment of Pediatrics, Johns Hopkins University School of Medicine ^cDana Center for Prevention Ophthalmology, Wilmer Eye Institute ^dDepartment of International Health, Bloomberg School of Public Health, Johns Hopkins University

Abstract

Purpose—To describe the prevalence of structural disorders of the eye and nystagmus in preschool-aged children.

Methods—Population-based evaluation of children 6 months through 71 months of age in Baltimore, Maryland, United States.

Results—Among 4,132 children identified from 54 census tracts, 3,990 eligible children (97%) were enrolled and 2,546 children (62%) were examined. Structural disorders were found in 41 children and nystagmus in 9 children for an overall prevalence of 1.96% (95% CI, 1.46%–2.59%). Only 11 (0.43%; 95% CI, 0.22%–0.77%) had vision loss in at least one eye, most often due to posterior segment disease.

Conclusions—Structural ocular abnormalities and nystagmus combined are present in nearly two percent of preschool-aged children in this population-based study. Vision loss due to these abnormalities is uncommon.

The Baltimore Pediatric Eye Disease Survey (BPEDS) was conducted to determine the prevalence of decreased visual acuity, strabismus, amblyopia, refractive error, and “other ocular disorders”—defined in this study as nystagmus or structural abnormalities of the ocular adnexa, the anterior segment, or the posterior segment—in a population-based sample of African American and non-Hispanic white children 6 months through 71 months of age living in Baltimore, Maryland. Prevalence information concerning the population-based disease burden associated with specific eye and vision problems can aid policy makers developing screening and treatment programs for eye problems. The purpose of this study was to describe the prevalence of other ocular disorders in the BPEDS cohort.

© 2012 American Association for Pediatric Ophthalmology and Strabismus. Published by Mosby, Inc. All rights reserved.

Correspondence: Michael X. Repka, Wilmer 233, Johns Hopkins Hospital, 600 North Wolfe Street, Baltimore, MD 21287-9028 (mrepka@jhmi.edu).

Conflicts of interest: The authors have no proprietary or commercial interest in any materials discussed in the manuscript.

Publisher's Disclaimer: This is a PDF file of an unedited manuscript that has been accepted for publication. As a service to our customers we are providing this early version of the manuscript. The manuscript will undergo copyediting, typesetting, and review of the resulting proof before it is published in its final citable form. Please note that during the production process errors may be discovered which could affect the content, and all legal disclaimers that apply to the journal pertain.

Methods

The protocol was approved by the institutional review board (IRB) of the Johns Hopkins Bloomberg School of Public Health as well as the Battelle Centers for Public Health Research and Evaluation IRB and the IRB of the Maryland Department of Health and Mental Hygiene. Parents or legal guardians provided written, informed consent for their child's participation. A detailed description of the BPEDS protocol has been published.¹

The study enrolled subjects from 54 contiguous census tracts in northeastern and eastern Baltimore City and adjacent portions of eastern Baltimore County. Parents or guardians of all enrolled subjects were invited to bring their child to the study clinic for a detailed interview and ophthalmological examination, including optotype visual acuity testing using the Amblyopia Treatment Study (ATS) visual acuity protocol for children 30 to 71 months of age (if possible)²; fixation preference testing at near; testing of ocular alignment at distance and near fixation; and examinations of the ocular adnexa, anterior segment, and posterior segment with pupillary dilation. Data were entered in real time into an electronic study record. Decreased visual acuity was defined as worse than 20/50 for children <4 years of age and worse than 20/40 for those ≥4 years among those subjects who were testable with appropriate refractive correction.¹ Two children could not complete optotype testing; in these, reduced vision was diagnosed following the study team's review of the records for clinical findings that would likely cause visual loss (one subject had optic nerve hypoplasia and the other had nystagmus).

Records coded with strabismus, decreased vision, nystagmus, and structural disorders were retrieved and reviewed for visual outcomes. These data vary slightly from prior BPEDS reports because we have included the entire study population sample here rather than just the two races reported in the primary study reports.^{1,3,4} Data for refractive error, amblyopia, and vision loss have been previously reported.^{1,3,4} Prevalence of other disorders was calculated as the number affected divided by the number examined. For these estimates, 95% confidence intervals were calculated using exact Poisson estimates around the mean event rate.

Results

We identified 63,737 occupied dwelling units in 54 census tracts. Among 4,132 children identified as eligible, 3,990 (97%) were enrolled and 2,546 (62%) were examined (1,309 boys). Study participants included 1,030 white children, 1,268 African Americans, 218 "others" and 30 with missing racial/ethnic information.

A total of 159 children (6.2% [95% CI, 5.3%–7.3%]: 68 white, 73 African American, 18 other) had abnormalities of alignment, vision, nystagmus, or structural abnormalities; of these, 71 (45%) were girls. Children not examined were similar to those examined in terms of race or ethnicity, gender, parent-rated eye health of the child, the proportion of parents reporting that the child had difficulty seeing in the past year, the proportion of parents reporting that the child had a prior diagnosis of an eye problem, and parent-rated general health of the child. Younger children were less likely to be examined, while those with health problems, those with stay-at-home mothers and those with parents with relatively more education were more likely to be examined.

Other ocular disorders were identified in 50 children (1.96% [95% CI, 1.46%–2.59%]: 23 white, 24 African American, 3 Hispanic). Prematurity was reported for 19 (38%), with 2 additional children of uncertain gestational age. Structural ocular disorders were found in 41 children, 8 of whom had vision loss; nystagmus was found in 9 children, 3 of whom had vision loss (Table 1).

Disorders of the posterior segment were observed in 16 subjects and disorders of the anterior segment in 14. Posterior segment disorders were often associated with vision loss (6 of 16 subjects), whereas disorders of the anterior segment (including most frequently iris anomalies and conjunctivitis) rarely caused vision loss (1 of 14 subjects).

By comparison, strabismus was found in 67 of 2,546 subjects (2.6%) of this BPEDS cohort, equally distributed between esotropia and exotropia. One case of Duane syndrome and 2 of dissociated vertical deviation were identified. Decreased vision after attempted refractive correction in at least one eye from any cause was found in 24 of 1,470 subjects (1.6%) subjects 36 months or older in BPEDS, about one-third each due to amblyopia, not amblyopia, and unclassified.

Discussion

In this population-based study, we identified at least one eye abnormality in 6.2% of our cohort of preschool children aged 6–71 months in Baltimore, Maryland. Most of these disorders were either decreased vision or strabismus. Other ocular disorders were encountered at a low rate in this population of children, with a prevalence of 1.96%. The majority of these disorders were incidental, as the prevalence of abnormalities significant enough to be associated with vision loss was very uncommon, found in only 0.43% of the cohort.

There are few population-based reports of preschool-aged children regarding this set of disorders with which we can compare our rate of visual impairment from ocular pathologies. Visual impairment was noted in 2 of 1,118 patients (0.2%) from ocular pathology in the Sydney Eye Study.⁵ The Multi-Ethnic Pediatric Eye Disease Study (MEPEDS) Group from Los Angeles, California, found 6 of 3,835 patients (0.2%) with vision impairment from ocular diseases.⁶

The cross-sectional Millennium Cohort Study, based solely on parental interviews at age 3 years, reported a prevalence of 5.7% for any eye condition and a prevalence of 0.24% visual loss.⁷ The prevalence of all conditions is similar to that in BPEDS, while the visual impairment rate is slightly lower. It is likely that because visual acuity was not tested in the Millennium Cohort Study some cases of vision loss may have been missed.

Recruiting subjects for BPEDS was challenging, and we examined only 62% of eligible children; nevertheless, data from the enrollment interview of parents and guardians completed for 97% of eligible children indicate that those who were examined were similar in terms of race/ethnicity, sex, quality of eyesight, and occurrence of visual problems in the prior year compared with children who were not examined. It is thus unlikely that the current estimates are subject to major nonresponse bias. Moreover, we did not include visual acuity data based on fixation preference testing for children unable to perform ATS single-surrounded HOTV optotypes because we have found it to be unreliable.^{8,9} This means that we may have underestimated the proportion of children with visual loss, since we did not have use of quantitative measures among the entire cohort. This could be particularly true for young children with nystagmus.

In conclusion, other ocular disorders were unusual in a predominantly African American and non-Hispanic white cohort of preschool-aged children. More than 80% of these disorders were not visually significant. Development of screening methods targeting these problems would not seem warranted as the large majority would be detected by current screening strategies based on determination of visual acuity and strabismus. Refractive error screening might miss some of the posterior segment abnormalities, but treatment options for these conditions are largely absent, reducing any adverse outcome from this strategy.

Acknowledgments

Financial support: Supported by the National Eye Institute, National Institutes of Health, Bethesda, MD (EY14483).

References

1. Friedman DS, Repka MX, Katz J, et al. Prevalence of decreased visual acuity among preschool-aged children in an american urban population: The Baltimore Pediatric Eye Disease Study, methods, and results. *Ophthalmology*. 2008; 115:1786–95. 1795.e1–4. [PubMed: 18538407]
2. Holmes JM, Beck RW, Repka MX, et al. The amblyopia treatment study visual acuity testing protocol. *Arch Ophthalmol*. 2001; 119:1345–53. [PubMed: 11545641]
3. Friedman DA, Repka MX, Katz J, et al. Prevalence of Amblyopia and Strabismus in White and African-American Children Aged 6 through 71 Months: The Baltimore Pediatric Eye Disease Study. *Ophthalmology*. 2009; 116:2128–34. [PubMed: 19762084]
4. Giordano L, Friedman DS, Repka MX, et al. Prevalence of refractive error among preschool children in an urban population: The Baltimore Pediatric Eye Disease Study. *Ophthalmology*. 2009; 116:739–46. [PubMed: 19243832]
5. Pai AS-I, Wang JJ, Samarawickrama C, et al. Prevalence and Risk Factors for Visual Impairment in Preschool Children: The Sydney Paediatric Eye Disease Study. *Ophthalmology*. 2011
6. Multi-Ethnic Pediatric Eye Disease Study (MEPEDS) Group. Prevalence and causes of visual impairment in African-American and Hispanic preschool children: the Multi-Ethnic Pediatric Eye Disease Study. *Ophthalmology*. 2009; 116:1990–2000. [PubMed: 19592106]
7. Cumberland PM, Pathai S, Rahi JS. Millennium Cohort Study Child Health Group. Prevalence of eye disease in early childhood and associated factors: findings from the millennium cohort study. *Ophthalmology*. 2010; 117:2184–90. [PubMed: 20561688]
8. Friedman DS, Katz J, Repka MX, et al. Lack of concordance between fixation preference and HOTV optotype visual acuity in preschool children: The Baltimore pediatric eye disease study. *Ophthalmology*. 2008; 115:1796–9. [PubMed: 18538405]
9. Cotter SA, Tarczy-Hornoch K, Song E, et al. Fixation preference and visual acuity testing in a population-based cohort of preschool children with amblyopia risk factors. *Ophthalmology*. 2009; 116:145–53. [PubMed: 18962921]

Table 1

Structural abnormalities and nystagmus recorded in the Baltimore Pediatric Eye Disease Survey

	Cohort (N = 2,546)	With vision loss
	Number (%)	Number (%)
Eyelids	9 (0.35)	1 (0.04)
Nasolacrimal duct obstruction	2 (0.08)	0
Anterior segment ^a		
Cornea	2 (0.08)	0
Iris ^b	6 (0.24)	0
Lens	2 (0.08)	1 (0.04)
Conjunctivitis	4 (0.16)	0
Posterior segment		
Retina	10 (0.39)	4 (0.16)
Optic nerve ^c	6 (0.24)	2 (0.08)
Nystagmus	9 (0.35)	3 (0.12)
Total ocular abnormalities excluding refractive error, decreased vision, and strabismus	50 (1.96)	11 (0.43)

^aOne subject had an abnormality of the lens and iris, categorized as lens.

^bIncluding coloboma (1), heterochromia (2), corectopia (1), iris stromal atrophy (1), and postsurgical changes (1).

^cOptic nerve cupping/atrophy (4), optic nerve pit (1), optic nerve hypoplasia (1).