Pediatric eye screening – Why, when, and how

The World Health Organization (WHO) reports that there are approximately 19 million visually impaired children in the world, and 1.4 million are blind.^[1] In India, 0.8 per 1000 children are estimated to be blind.^[2] This is bound to be an underestimation of the overall problem because it excludes children with visual impairment that does not conform to the WHO definition of blindness. About half of the causes of blindness and visual impairment are potentially preventable or treatable.^[2] Childhood blindness is second only to adult cataract in terms of the number of blind person years lived and the consequent overall economic impact on the society.^[2]

Timely and periodic screening is critical for the detection of visual impairment and its etiology and to plan early intervention. Appropriate estimation of the visual function, and detection of refractive error, retinopathy of prematurity, congenital structural anomalies, congenital dacryocystitis, corneal scar, glaucoma, cataract, retinal abnormalities, retinoblastoma, strabismus, and amblyopia are the crucial components of screening in children. Protocols vary from country to country, with limited agreement on the need, modality, timing and periodicity of screening.^[3-6] While some countries and organizations have mandated screening at birth and thereafter periodically at every pre-scheduled point of contact with the pediatrician,^[3-6] recent recommendations by the United States Preventive Services Task Force (USPSTF) limit screening to children aged 3-5 years to detect amblyopia or its risk factors.^[7] The USPSTF advises that the current evidence is insufficient to recommend vision screening in children <3 years of age.^[7] However, a Joint Policy Statement by the American Academy of Pediatrics, American Academy of Ophthalmology, American Association for Pediatric Ophthalmology and Strabismus, and American Association of Certified Orthoptists emphasises that vision assessments and screening eye examinations are critical for the detection of conditions that result in visual impairment, lead to problems with school performance, harbinger serious systemic disease, and, in some cases, threaten the child's life.^[8,9] The incidence and prevalence of conditions causing visual morbidity varies widely across the world [Table 1].^[10] Unfortunately, the available data on pediatric vision impairment and blindness in India are not broad-based or robust enough to generalize and make firm recommendations.^[2] Glaringly, there are no formal Indian national guidelines for vision and eye screening in children.

Table 1: Estimated burden of visually significant ophthalmic conditions in children	

Condition	Frequency*
Refractive errors Myopia (-0.75 D or more in eye with lesser refractive error) Myopia (more than -2.0 D) Hyperopia (+3.0 D or more in eye with lesser refractive error) Hyperopia (more than +3.25 D) Astigmatism (worse eye cylinder power 3.0 D or more) Astigmatism (cylinder power more than 1.5 D)	0.7%-9% (prevalence, age 5-17 years) 0.2%-2% (prevalence, age 3-5 years) 4%-9% (prevalence, age 5-17 years) 6%-7% (prevalence, age 3-5 years) 0.5%-3% (prevalence, age 5-17 years) 4%-11% (prevalence, age 3-5 years)
Amblyopia	0.8%-3% (prevalence, age 6-72 months)
Strabismus	0.08%-4.6% (prevalence, age 6-72 months) 1.2%-6.8% (prevalence, age 6-17 years)
Cerebral visual impairment	Accurate data is lacking
Congenital cataract	0.02% (prevalence, age 0 to 1 year) 0.1% (prevalence, age 6 months to 6 years) 0.42% (prevalence, age 6 to 15 years)
Retinopathy of prematurity	8.6%-9.2% (incidence, 1000-1250 g at birth) 15.2%-18.3% (incidence, 800-999 g at birth)
Congenital glaucoma	0.0015%-0.0054% (prevalence, new born)
Retinoblastoma	0.0011%-0.0013% (yearly incidence, age <5 years)
Pediatric uveitis	0.004% (yearly incidence, age <16 years)

D: Diopter. *Please refer to the source document for references that support the data. Source: Wallace DK, Morse CL, Melia M, Sprunger DT, Repka MX, Lee KA, *et al.*, American Academy of Ophthalmology Preferred Practice Pattern Pediatric Ophthalmology/Strabismus Panel. Pediatric Eye Evaluations Preferred Practice Pattern: I. Vision Screening in the Primary Care and Community Setting; II. Comprehensive Ophthalmic Examination. Ophthalmology 2018;125:P184-P227.

Guidelines for pediatric eye screening continue to evolve as timing and methods have not been definitively established. Current guidelines are based on the available evidence and preferred practice recommendations of expert committees.^[8-10] Primary care providers (pediatricians) should perform a basic eye screening of newborns.^[8-10] Risk-based screening for retinopathy of prematurity, congenital anomalies, and retinoblastoma in the immediate post-natal period should be conducted by an ophthalmologist.^[8-10] Pre-screening history should include the following questions: (1) Do your child's eyes appear unusual, (2) Does your child seem to see well, (3) Does your child exhibit difficulty with near or distance vision, (4) Do your child's eyes appear straight or do they seem to cross, (5) Do your child's eyelids droop or does one eyelid tend to close, and (6) Has your child ever had an eye injury.^[8-10] Screening of infants under 6 months of age comprises of red reflex testing to detect abnormalities of the ocular media, external inspection of ocular and periocular structures, pupillary examination, and assessment of fixation and following screening are listed in Table 2.^[10,11] Screening from 6 months to 1 year includes binocular alignment.^[10,11] Between 1 year to 2 years and 2 years to 3 years, instrument-based screening with photoscreening or autorefraction devices can be valuable in detecting amblyopia risk factors.^[10,11] These tests are rapid and non-invasive, and minimal cooperation is required on the part of the child.^[10,11] Between ages 3 and 4 years, visual acuity screening with LEA symbols or HOTV letter chart become possible.^[10,11] Older children may

Method	Screening Logistics	Indications for Referral	Recommended Age					
			0-6 m	6-12 m	1-3 y	3-4 y	4-5 y	>5 y
Comprehensive screening at birth	Primary screening at birth by the pediatrician. Risk-based primary screening for retinopathy of prematurity, congenital anomalies, and retinoblastoma by the ophthalmologist	Congenital structural anomalies of the eye and adnexa, congenital dacryocystitis, congenital corneal opacity, congenital cataract, congenital glaucoma, retinopathy of prematurity, and retinoblastoma						
Red reflex test	Ophthalmic technician or optometrist	Absent, white, dull, opacified, or asymmetric	Yes	Yes	Yes	Yes	Yes	Yes
External inspection	Ophthalmic technician or optometrist	Structural abnormality (e.g., ptosis, epiphora)	Yes	Yes	Yes	Yes	Yes	Yes
Pupillary examination	Ophthalmic technician or optometrist	Irregular shape, unequal size, and poor or unequal reaction to light	Yes	Yes	Yes	Yes	Yes	Yes
Fix and follow	Ophthalmic technician or optometrist	Failure to fix and follow	Yes*	Yes	Yes	Yes	Yes	Yes
Corneal light reflex	Ophthalmic technician or optometrist	Asymmetric or displaced		Yes	Yes	Yes	Yes	Yes
Instrument-based screening	Ophthalmic technician or optometrist	Failure to meet screening criteria			Yes	Yes	Yes	Yes
Cover test	Ophthalmic technician or optometrist	Refixation movement				Yes	Yes	Yes
Distance visual acuity (monocular)	Ophthalmic technician or optometrist	Worse than 20/50 either eye or 2 lines of differences between the eyes				Yes	Yes	Yes
		Worse than 20/40 either eye					Yes	Yes
		Worse than 3 of 5 optotypes on 20/30 line, or 2 lines of difference between the eyes						Yes

Table 2: Age-appropriate methods for pediatric vision screening and criteria for referral

*in a cooperative child >3 m. Source: Hagan JF, Shaw JS, Duncan PM, eds. 2017, Bright Futures: Guidelines for Health Supervision of Infants, Children and Adolescents. 4th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2017 and Wallace DK, Morse CL, Melia M, Sprunger DT, Repka MX, Lee KA, *et al.*, American Academy of Ophthalmology Preferred Practice Pattern Pediatric Ophthalmology/Strabismus Panel. Pediatric Eye Evaluations Preferred Practice Pattern: I. Vision Screening in the Primary Care and Community Setting; II. Comprehensive Ophthalmic Examination. Ophthalmology 2018;125:P184-P227

be tested with standard optotypes.^[10,11] Children who are untestable should be rescreened within 6 months or referred for a comprehensive eye examination.^[10,11] Children who are testable using the subjective visual acuity assessment and fail should be referred for a comprehensive eye examination after the first screening failure.^[10,11] Additional findings that would warrant referral of for a comprehensive ophthalmic examination are listed in Table 2.^[10,11] Children should continue to have annual school-based vision screening throughout the childhood and adolescence.^[10,11] In India, screening up to age 5 years could be integrated with the Universal Immunization Program of the Government of India and performed by a trained ophthalmic assistant or an optometrist. Beyond the age of 5 years, it should be a part of annual school health check-up and performed by a trained ophthalmic assistant or an optometrist.

This issue of the Indian Journal of Ophthalmology carries several articles that address various issues related to vision screening in children, which indicates that there is renewed enthusiasm in Indian caregivers and researchers to study this aspect.^[12-16] If India must relieve itself of the burden of avoidable pediatric blindness and provide the benefit of early rehabilitative intervention to those who are incurably blind, then it is imperative to accumulate reliable population-based data and use that as a base to craft a robust screening program, seamlessly linked to curative and rehabilitation facilities. A working group representing all the stakeholders seems to be an immediate primary need to prioritize this issue.

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Cover Page Illustration

Bietti's crystalline dystrophy

Bietti's crystalline dystrophy is a rare inherited retinal dystrophy, which usually presents with progressive visual loss in the third or fourth decade. The disease is characterized by multiple small intraretinal crystal deposits. This causes retinal atrophy leading to peripheral visual field and visual loss often causing legal blindness by the fifth or sixth decade. The gene responsible is cytochrome P450 4V2 (CYP4V2) which codes a protein involved in fatty acid metabolism.

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