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All India Ophthalmological Society Guidelines

- ▶ **National, Expert-Based Consensus Statement Regarding Paediatric Eye Examination, Refraction And Amblyopia Management**



AIOS Focus Group Meeting
22nd October, 2017

AIOS Thanks

All the experts who participated and Cipla



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National, Expert-Based Consensus Statement Regarding Paediatric Eye Examination, Refraction And Amblyopia Management

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Word from your Secretary



Prof. (Dr.) Namrata Sharma

Hony. General Secretary, AIOS

Dear Friends,

Paediatric ophthalmology is not only a challenging field but also requires a proper understanding of the developing eye in children. The ability to get the maximum information from a brief encounter with a child is a skill honed after years of practice and hard work. Therefore, any text that gives the secrets of successful examination and managing a child would be very useful to all ophthalmologists. It gives me great pleasure to bring for all AIOS members the National, Expert-Based Consensus Statement Regarding Paediatric Eye Examination, Refraction And Amblyopia Management.

This is a document that has evolved after some of the most experienced and learned in this field have got together and evolved a consensus on this topic. It contains essential information and practice patterns that every ophthalmologist must be aware of whenever examining and managing a child.

I wish to thank Cipla for funding this extremely important endeavour and providing the logistics for holding the meeting. I also wish to thank all the participants who have taken out time to provide their knowledge and experience that has enabled this document to become an essential tool for every ophthalmologist. I must also express my gratitude to Prof. Rohit Saxena who helped us to organise this meeting and put the document in place.

We hope that you all will find this a useful and practical guide and use it in your day-to-day clinical practice.

A handwritten signature in black ink that reads "Namrata Sharma". The signature is written in a cursive style.

Prof. (Dr.) Namrata Sharma

Hony. General Secretary, AIOS

Editorial



Prof. (Dr.) Rohit Saxena

RP Centre, AIIMS, New Delhi

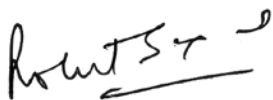
It is indeed a proud privilege to bring out this National, Expert-Based Consensus Statement regarding Paediatric Eye Examination, Refraction and Amblyopia Management, which contains the nitty-gritty of various aspects of examining a child and prescribing spectacles. While there have been many guidelines published by paediatric ophthalmologists from the west, there has been no such practice patterns available for Indian ophthalmologists and what would be the preferred clinical practice for our country. This document hopes to fill this void and provide guidelines best suited in the Indian scenario.

The process of developing the document involved creating a draft document using the best available scientific publications. The Scientific data included the best suited management options, particularly in the Indian scenario. These were discussed amongst a panel of Paediatric ophthalmologists and super specialists in the field, from across India under the aegis of the All India Ophthalmological Society. Finally a consensus statement for paediatric eye examination, paediatric refraction and amblyopia management in the Indian scenario was developed. The aim of the document is to provide an outline for the management of a disease in general and to help the ophthalmologists in paediatric eye care.

I wish to thank all the participants of the meeting who have provided great insights during the creation of this document as also my senior residents, Dr. Aswini Behra and Dr. Kishen Azmera who have help me in compiling all the information discussed in the meeting and put it together in a simple and easy to understand text.

I also wish to thank AIOS and particularly Dr. Santhan Gopal, the President, Dr. Namrata Sharma, the Secretary and Dr. Rajesh Sinha, Treasurer, who gave us the opportunity and support without which this document would not have been possible.

I hope all the members find this a useful document and use the information provided in their routine clinical practice.



Prof. Rohit Saxena

RP Centre, AIIMS, New Delhi

Word from your Treasurer



Prof. (Dr.) Rajesh Sinha
Hony. Treasurer, AIOS

Dear Friends,

The All India Ophthalmological Society is an academic body meant for dissemination of scientific knowledge amongst all its members. For the same purpose, various conferences have been organised over the last so many years. However, there was always the need for a consensus on the management protocol of various clinical disorders. For the same reasons, we started the Focussed group meetings to have a discussion at length amongst the leaders of various subspecialties to arrive at a consensus regarding the management protocol of a particular ocular condition. The basic idea is to enable the practicing ophthalmologists to understand the main ideas and concepts of various clinical entities and to apply them to solve problems in familiar and unfamiliar situations. This consensus can serve as a guideline for the general ophthalmologists not only to manage the patients in their clinical practice but also to explain to the patients that their management has been done as per a set guidelines by the experts in that particular field so that in case of suboptimal response to treatment they can defend themselves. Although such guidelines may not be useful as a document to protect anyone in a court of law, however they can always be produced to support the point of the ophthalmologists that he/she has followed a defined guidelines made by the experts in that specialty and hence no error was done. Apart from all these, it will definitely have a positive impact on the eye care facilities provided to our patients.

To create such guidelines on Refractive Error Correction and Amblyopia Therapy, notable experts in the field of Paediatric Ophthalmology from the entire country were invited. A detailed questionnaire was prepared related to the understanding of the clinical condition and the management protocol of the same. It was then put forward to each expert present in the meeting. There was a detailed discussion on each issue and a consensus was arrived at every point. The whole matter was finally read out and discussed again to fine tune the matter. This was then converted in the form of guidelines and sent for publication. I would like to appreciate and thank Prof. Rohit Saxena for all the efforts that he has made in compiling and finalising the whole guideline.

I hope that the efforts of all the experts in the field of Paediatric Ophthalmology and AIOS will be of immense use for the practicing ophthalmologists in managing refractive error and amblyopia. We are ready with guidelines related to other ophthalmic conditions as well and we have already planned for more focussed group meetings and consensus guidelines on various ophthalmic disorders in future.

Best regards,

A handwritten signature in black ink that reads "Rajesh Sinha". The signature is written in a cursive style.

Prof. (Dr.) Rajesh Sinha
Hony. Treasurer, AIOS

Preface

All India Ophthalmological Society, the largest association of life-member ophthalmologists in the world, has developed consensus on the management for a few common disorders, as a duty to serve its members, eye care professionals and general public at large.

The best available scientific publications were discussed amongst a panel of Paediatric ophthalmologists and super-specialists in the field, from across India, under the aegis of the All India Ophthalmological Society, to formulate a consensus statement for paediatric eye examination, paediatric refraction and amblyopia management in the Indian scenario. The scientific data was discussed and the best suited management option/ options, particularly in the Indian scenario, were included.

The document provides an outline for the management of a disease in general and to help the ophthalmologist in paediatric eye care; however, this cannot be applied to a particular case or individual, nor does it guarantee that strict adherence to treatment protocol thus outlined would lead to success in each and every case.

The attending qualified eye care professional can use the consensus statement for the management of disease/ situation; however, the discretion to deviate is his/her domain. There is no legal boundation to practice strictly as per the document; the attending qualified eye care professional can decide the best course of treatment for the benefit of patient depending upon the cultural/social/economical/geographical or any other diversity. The recommended pattern of practice is a guideline and not a medical standard, which needs to be adhered to in each and every situation. All India Ophthalmological Society is accessible to help the members and eye care professionals, in case needed, to resolve the issues arising out of pattern of practice.

The drugs, instruments and any other product mentioned in the consensus document are only for descriptive purpose, the society does not endorse any of them.

The panel of ophthalmologists and super-specialists in the field would meet again to add/delete/modify the consensus document as the need may arise; the present consensus document is valid unless it is revised/ modified.

Background and Need

Childhood blindness has received priority status by VISION 2020- Right to Sight initiative, as it results in significant social and economic burden.¹ Preventing or treating childhood blindness has a significant impact in terms of “blind years” avoided, gained productivity, and overall savings to the society. Common causes of childhood blindness in India are refractive errors, paediatric cataract, glaucoma, corneal blindness, congenital anomalies, ocular trauma and retinopathy of prematurity.^{2,3,4,5,6} Most of these diseases can be easily avoided or treated by reliable eye examination and timely management.^{1,7}

Despite being prioritised, paediatric eye care services in India are grossly inadequate. According to the World Health Organization (WHO), there should be one paediatric ophthalmology service center for every 10 million population. In India, this ratio is estimated to be 0.63 per 10 million population.⁸ As shown in a recent study, children form a significant proportion of patients at a tertiary eye care hospital, which has been attributed to the lack of paediatric specific services at primary and secondary healthcare level.⁹ While there is a need for trained paediatric ophthalmologists at tertiary eye care institutions, there is a greater need to impart education regarding paediatric eye examination to general ophthalmologists and optometrists around the country.¹⁰ Guidelines and preferred practice patterns for paediatric eye examination have been formulated by organisations such as the American Academy of Ophthalmology (AAO), American Academy of Paediatric Ophthalmology and Strabismus (AAPOS), and World Society of Paediatric Ophthalmology and Strabismus (WSPOS); however, they may not be applicable to India given its limited resources and lack of trained manpower.

The objective of the meeting was to generate a national level expert consensus statement on paediatric eye examination and refraction in order to enhance paediatric eye evaluation.

Paediatric ophthalmologists from across India met in New Delhi on 22nd October, 2017 to discuss the current practice patterns for paediatric eye examination, paediatric vision screening, refraction and management of amblyopia. The experts shared their experiences and discussed challenges currently faced in examining a child’s eye and performing refraction. The expert panel deliberated on different aspects of the paediatric eye examination and management of amblyopia and reached a consensus regarding the practice patterns in the Indian scenario.

All expert panelists and key opinion leaders in the field of paediatric ophthalmology concurred with regards to the lack of clear recommendations as to the timing of vision screening, components and methods of comprehensive paediatric eye examination, prescription of refractive errors based on age appropriate use of cycloplegia and indications for referral to paediatric ophthalmologist in the Indian scenario. The final consensus statement derived is below.

Paediatric Eye Examination

Paediatric eye examination should include the following:

History

A carefully taken history is an essential component of paediatric eye examination. It is often sufficient to reach a diagnosis, guide investigations, highlight warning signs of a severe pathology, and save time and resources. The informant should preferably be the person with whom the child spends maximum time (generally, the mother). These are some important points to be considered in paediatric history:

- Antenatal history – directed to rule out TORCH (rashes, fever etc) infections, immunisation status
- Perinatal history – details of delivery, whether pre-term, birth weight, cry at birth, institutional care (NICU stay)
- Systemic history – developmental milestones, history of seizures, neuropsychological symptoms, history of cerebral palsy, Down's syndrome, etc.
- Ocular history – history of misalignment of the eye, rubbing of the eyes, white reflex in the eye or any other ocular complaints
- Family history – history of consanguinity, history of ocular illness
- Other history – any known allergy, any medication use, immunisation status

All the points may not be relevant in every case and the attending ophthalmologist must decide based on individual case scenario.

Ocular examination

A torch, direct and indirect ophthalmoscope are the minimum necessary instruments for examining a child. The child should ideally be examined when he/she is comfortable, awake and alert, preferably on the mother's lap or shoulder (for small children). Diffuse light should be used for examination as bright light can startle the child and make any further evaluation difficult. Toys, pictures, candies, mobile phones or anything that catches the child's attention can be used for examination (Figure 1). Fundus and anterior segment examination can be best performed when the child is asleep or sedated. Examination under anaesthesia may also be needed in selected cases.



Figure 1: Paediatric eye examination using fixation toys

Comprehensive eye examination should include:

- Vision assessment (Unocular and binocular) by age-appropriate methods of vision screening. Assessment should be performed binocularly as well as unocularly. Methods of vision assessment in a child have been discussed later in the article.

- External examination includes looking for oculofacial anomalies, head posture and a torch-light examination of the external ocular structures like lids, lacrimal system, cornea, conjunctiva, sclera, and iris. Head posture can be best assessed by observing the child and his/her interaction with parents.
- Ocular motility – Ocular motility is assessed using toys and objects that will get the child's attention. Hirschberg test or a cover test may be performed in suspected cases of ocular misalignment. It is important to differentiate pseudo-strabismus from true strabismus. Presence of nystagmus is an indicator of poor vision possibly due to some congenital pathology.
- Pupils – Pupillary examination includes the assessment of size, shape, colour of iris, inter-eye asymmetry in pupil size, and pupillary reactions (direct and consensual light reflex and relative afferent pupillary reflex). Asymmetry in pupil size or abnormal pupillary reflexes may be suggestive of an underlying CNS pathology, thus necessitating an urgent referral.
- Distance Direct Ophthalmoscopy – Distance direct ophthalmoscopy to look for the red reflex should be performed in a darkened room. Absence of red reflex, a markedly diminished reflex, presence of a white or yellow reflex (leucocoria), or asymmetry of the red reflexes are all considered abnormal and are important indications for referral.
- Slit-lamp biomicroscopy (whenever applicable) should be performed for detailed ocular examination.
- Dilated fundus examination should be done for every child to evaluate the posterior segment.
- Age-appropriate cycloplegic refraction using retinoscopy should be performed in all cases.
- Stereoacuity assessment (optional) can be done atleast once in cooperative patients.

Methods of paediatric vision screening and criteria for referral

Vision screening is the most important aspect of paediatric eye examination. It can be challenging in pre-verbal children and ophthalmologists often have to rely on indirect means of assessing vision. These include fixation patterns, pupillary light reflexes and attitude towards the surroundings. Fixation reflex is present at birth in most infants, and develops in all by 3 months of age. Central, steady and maintained fixation with free alternation is suggestive of good and equal vision in both eyes. Fixation preference or resistance in occluding one eye implies poor vision in the fellow eye. Visually evoked response (VER) is an objective method of assessing the visual pathway and is helpful in infants or uncooperative children with developmental delay. Teller's (Figure 2) and Cardiff acuity cards (Figure 3), HOTV and Lea symbols may also be used, if available. Subjective visual assessment is preferred over instrument-based screening (VER), wherever possible.

In school-going children, vision assessment is simple and can be performed by using Snellen's chart at 6-metre distance or LogMAR charts at 4-metre distance.

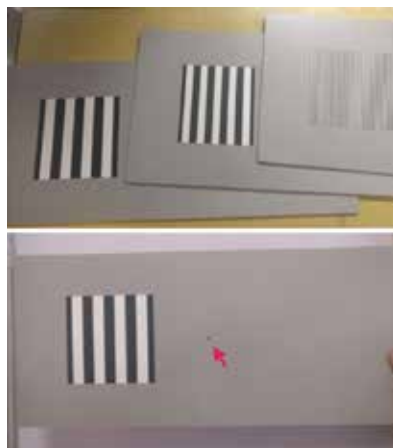


Figure 2: Teller's visual acuity cards showing the peeping hole for the examiner

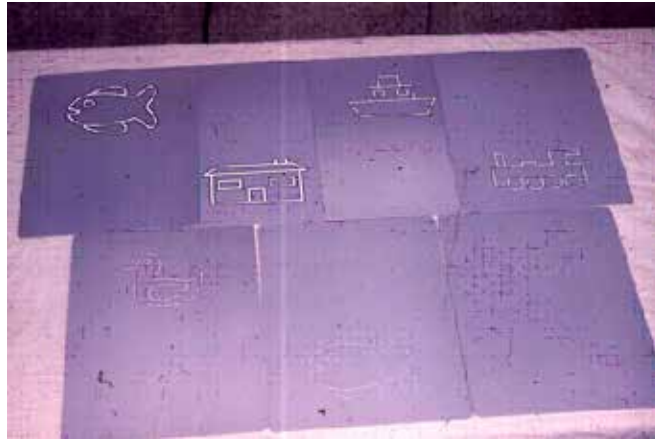


Figure 3: Cardiff visual acuity cards with vanishing optotypes

Table 1 summarises the various indications for referral to a paediatric ophthalmologist for comprehensive eye examination at a higher centre.

TABLE-1: Indications for referral to a paediatric ophthalmologist or higher center							
Method	Indication for referral	Recommended age					
		New born-6 months	6-12 months	1-3 years	3-4 years	4-5 years	Every 1-2 years after 5 years of age
Red reflex test	Absent, white, dull, opacified, or asymmetric	+	+	+	+	+	+
External inspection	Structural abnormality (e.g., ptosis)	+	+	+	+	+	+
Pupillary examination	Irregular shape, unequal size, poor or unequal reaction to light	+	+	+	+	+	+
Fix and follow	Failure to fix and follow	Cooperative infant ≥ 3 months	+	+			
Corneal light reflection	Asymmetric or displaced		+	+	+	+	+
Instrument-based vision screening	Failure to meet screening criteria			+	+	+	+
Cover test	Re-fixation movement				+	+	+
	Worse than 20/50 either eye or 2 lines of differences between the eyes				+	+	+
Distance visual acuity (monocular)	Worse than 20/40 either eye					+	+
	Worse than 3 of 5 optotypes on 20/30 line, or 2 lines of difference between the eyes						+

Adapted from American Academy of Ophthalmology Preferred Practice Patterns 2018¹¹

Timing of examination and screening

Paediatric vision screening and eye examination should be timed in such a way so as to maximise the detection of ocular morbidities and be able to provide timely intervention.

Table 2 gives the guidelines for vision screening and comprehensive eye examination in children.

TABLE-2: Timings for Vision Screening	
When	How
Neonates high-risk cases*	At discharge within 1 month
Birth-3 years	Vision screening by an ophthalmologist atleast once
3-5 years	Comprehensive eye examination by an ophthalmologist atleast once
5-8 years	Comprehensive eye examination by an ophthalmologist atleast once

*High-risk cases- Premature, low birth weight, Down's syndrome, etc.

Other tests

Intraocular pressure (IOP)

IOP assessment is not necessary for every child. It should be considered only in cases with high-risk features of buphthalmos, epiphora, photosensitivity and corneal clouding. Instruments which may be used in the OPD for IOP assessment in infants and toddlers are Tonopen or ICare. Older children may be able to cooperate with the non-contact tonometer or Perkin's/ Goldmann Applanation tonometer. Examination under anaesthesia may be needed in uncooperative children.

Visual fields

Visual field testing is not routinely required and if needed, confrontation visual field testing may be performed in children. The peripheral visual field of younger children can be assessed by observation for re-fixation to the field of gaze in which an object of interest has been presented. Quantitative visual field testing should be attempted only when indicated as reliability of the test may be poor.

Imaging

Imaging in paediatric age group is indicated only in certain special situations.

- Face photography may be needed to document and follow changes of facial or ocular structural abnormalities.
- Anterior segment photography through a slit-lamp, for cataract and other anomalies.
- Corneal topography to detect early changes related to keratoconus.
- Image analysis for optic nerve head assessment or abnormal retinal pathology.

Paediatric Refraction

Paediatric refraction is challenging since children have strong accommodation, are often uncooperative and have a dynamic ocular system due to growth. Important questions in paediatric refraction are, “how to refract”, “what cycloplegic to use”, and “when to prescribe”. As there are no standard guidelines for refraction and prescription for paediatric patients in India, most ophthalmologists rely on their clinical experience. The consensus on paediatric refraction is summarised below.

When to refract

All children should undergo vision assessment and refraction (if vision is suspected to be sub-normal on screening), at least once before the child starts going to school i.e., at 3-4 years of age. Early refraction is indicated in symptomatic cases i.e., difficulty in identifying faces from a distance, history of frequent falls, watching television or mobile closely, etc. and in certain clinical settings like history of any ocular surgery particularly congenital cataract, premature birth, peri-natal insult or developmental delay. Pediatricians should be sensitised for timely referral to an ophthalmologist for the same as a high refractive error may lead to squint or amblyopia besides adversely affecting normal growth and development of the child.

What Cycloplegic to be used

Atropine, homatropine and cyclopentolate are commonly used cycloplegics in children and are often used interchangeably.

Table 3 describes the age-appropriate use of these cycloplegics agents.

TABLE-3: Age-appropriate cycloplegia	
Condition	Cycloplegic
Presence of esotropia <ul style="list-style-type: none">• Till 5 years• >5 years	Atropine 1% eye ointment Atropine 1% eye ointment/ Cyclopentolate 1% eye drop/ Homatropine 2% eye drop
No strabismus	Atropine 1% eye ointment/ Cyclopentolate 1% eye drop/ Homatropine 2% eye drop

Atropine is the cycloplegic of choice in case of esotropes less than 5 years of age as it is essential to eliminate the accommodative component of esotropia. Atropine sulphate ointment, in a concentration of 1%, should be applied in rice-grain size, three-times a day for three days prior to the day of refraction. Atropine drops are usually avoided as the risk of systemic absorption and consecutive systemic side-effects are higher. In children older than 5 years and in orthophoric patients, any one of the three cycloplegics could be used. However, the side-effects of these drugs must be kept in mind and the parents should be forewarned before prescribing them.

Ocular side-effects seen with atropine eye ointment are contact dermatitis of the lids, allergic conjunctivitis, keratitis and raised intraocular pressure. Systemic adverse effects are dryness of secretions, fever, skin rash, irritability, tachycardia and convulsions. Side-effects of homatropine are similar to but less severe than those of atropine. Cyclopentolate drops can cause ocular irritation, lacrimation, allergic blepharoconjunctivitis, conjunctival hyperemia, and increase in intraocular pressure. Systemic side-effects include irritability or drowsiness, ataxia, disorientation, incoherent speech, restlessness and visual hallucinations.

Prescribing spectacles

There are different practices in different parts of the country about the minimum refractive error to be prescribed.

Table 4 provides the national consensus for minimum refractive correction in infants and young children. Adapted from American Academy of Ophthalmology Preferred Practice Patterns 2018¹¹ for Indian conditions.

TABLE - 4: Minimum refractive correction to be prescribed in infants and young children			
Condition	Refractive errors (diopters)		
	<1 year	1-2 years	2-3 years
Isometropia			
Myopia	≥-3D	≥-3D	As per refraction
Hyperopia (no manifest deviation)	≥+4D	≥+4D	≥+4D
Hyperopia with esotropia	≥+1.5	≥+1.5	≥+1.5
Astigmatism	≥3D	≥2D	≥2D
Anisometropia (without strabismus)			
Myopia	≥-3D	≥-3D	As per refraction
Hyperopia	≥+2 D	≥+1.5D	≥+1.5D
Astigmatism	≥2.5D	≥2D	≥2D

How to prescribe spectacles

After having assessed the refractive error in a child, the next step is to appropriately prescribe spectacles with a good acceptance.

Prescription in special conditions:

- Prescription in pseudophakes and aphakes – Refractive error as assessed by retinoscopy should be prescribed immediately irrespective of age in pseudophakes and aphakes. Only near spectacles are sufficient in children up to 2 years, and thereafter with the added demand for distance vision, bifocals are to be prescribed (Figure 5). Occlusion is needed in unilateral pseudophakes/aphakes or bilateral cases with unequal vision.
- Gross developmental delay, Down's syndrome or other cases of mental retardation – Refraction and prescription according to retinoscopy can be given as early as 6 months of age.
- Retinopathy of prematurity (ROP) – Myopia tends to progress in cases of ROP and prescription can be given according to retinoscopy as early as 6 months.
- Intermittent exotropia – Myopia correction can be given as per the recommended guidelines mentioned in Table 4. Overcorrection up to -0.5D can be tried in children >3 years.
- Esotropia – Hyperopia ≥+1.5D must be prescribed in children with esotropia. Overcorrection can be done if vision does not fall below 6/12. Bifocals are needed in cases with high AC/A ratio. Weaning of spectacles should start at the age of 7 years depending on the retinoscopy.

Table 5 gives the recommendation for the type of spectacles to be prescribed in children.

TABLE - 5: Spectacles specifications

Frame	Characteristic – Break-resistant, light, colourful, preferably with nose pads, elastic headbands for infants				
	Materials – titanium (ideal), plastic materials (cellulose acetate, polyamide), silicon-based rubber frames (light and pliable) Soft ear support, elastic band to prevent peeping over the frame				
Lens	Characteristic – Impact-resistant, thin, light, scratch free, UV protection, high index (preferably)				
	Materials	CR39	Polycarbonate	Trivex	High-index plastics
	Impact resistance	Good	High	High	Very High
	Refractive index	1.498	1.586	1.532	1.6-1.74



Figure 4: Prescription of full cycloplegic hyperopic correction in a case of accommodative esotropia



Figure 5: Executive style bifocal spectacles (transecting through the pupil) in an accommodative esotropia with high AC/A ratio

Amblyopia

Amblyopia is a disease of early childhood, and most of the children can be cured if diagnosed and treated in time. It is associated with significant morbidity in terms of life-long poor vision and its adverse effects on quality of life. The current prevalence of amblyopia ranges from 0.8% to 3.3%.

Amblyopia is defined as reduced best-corrected visual acuity (BCVA) in one or both the eyes which are structurally normal or the structural abnormality of the eye or the visual pathway does not attribute fully to the reduction in the BCVA. It is due to formed visual deprivation (due to media opacity or image blur) or abnormal binocular interaction. These abnormal visual experiences are usually in early childhood, typically before the age of visual maturation (which is around 7 years). Based upon the possible causative factor, amblyopia can be classified as strabismic, refractive (anisometropic or ametropic/ bilateral refractive error), visual deprivation or mixed type.

Strabismic Amblyopia

Presence or occurrence of strabismus early in life leads to abnormal binocular interactions. Both esodeviations and exodeviations can adversely affect the visual development of a child. However, unequal, constant and non-alternating esodeviations are supposed to be the most dangerous for visual development. While exodeviations may hamper development of stereoacuity, esodeviations may lead to reduced vision of the constantly deviated eye. With presence of esotropia, there is abnormal binocular interaction, as the images captured by each eye are separate and non-fusible. This causes binocular rivalry/competition, which leads to visual cortical domination of the fixing eye over the visual cortex of the deviating eye, resulting in suppression and amblyopia.

Refractive Amblyopia

Vision development in children depends on the formation of clear image on both retinae. Presence of significant refractive error (unilateral or bilateral), leads to blurred or defocused image.

In untreated anisometropia, where the refractive error is unequal, the image blur of one eye is greater than the other; therefore the eye with greater image blur is at risk of developing amblyopia due to direct effect of image blur on the development of vision. There is also a component of competitive inhibition or binocular rivalry which may contribute to amblyopia.

Bilateral refractive error leads to bilateral blur in retinal image formation and bilateral amblyopia. Significant hypermetropia, moderate-to-high myopia and significant astigmatism can cause amblyopia in children.

Refractive amblyopia, in presence of esotropia may be associated with strabismic amblyopia. In such cases, it is believed that the binocular rivalry/competition is stronger than pure refractive, and hence the prognosis may be poorer.

Visual Deprivation Amblyopia

Visual deprivation is the least common but the most severe form of amblyopia. Complete or partial opacification of ocular media leads to image blur which causes amblyopia. Visual deprivation in unilateral settings is more severe than in bilateral conditions, as in unilateral cases competitive inhibition from the sound eye plays a role in addition to image blur.

The common causes are congenital or early cataracts, infectious or non-infectious intraocular inflammation, vitreous haemorrhage, ptosis, etc.

Cataracts developing early carry worse prognosis. Cataracts in < 6 years of children with > 3 mm of opacity, obstructing the pupil are much more harmful than those occurring in older children.

Small polar or lamellar opacities of the lens where refraction can be performed and corrective spectacles can be given, may lead to normal visual development.

In certain situations, where there is presence of structural abnormality like optic nerve hypoplasia, retinopathy of prematurity, uveitis etc, there may still be a component of treatable amblyopia.

All children irrespective of age of presentation and presence of co-existing ocular abnormality should be offered appropriate amblyopia therapy trial.

Work-up

The basic work-up should include a reliable history, physiological and anatomical assessment of the eye and the visual system as a whole.

1. Demographic data including gender, date of birth (age), informant's identity and relationship to the patient.
2. A detailed and comprehensive eye examination is essential.
3. Specific attempts should be made to find out presence of risk factors for the development of amblyopia including strabismus, anisometropia, positive family history for either strabismus or amblyopia, structural abnormalities including media opacity.
4. Present eye problems, history of previous ocular disorders, treatment taken, etc.
5. Detailed pre/perinatal history including gestational age, birth weight, presence of developmental delay or cerebral palsy.
6. If indicated, review by a paediatrician of the general health of the child and evaluation of the developmental status may be done.
7. Co-operation level of the child should be documented at each visit, so that the accuracy of the findings is taken into account for future comparisons.

Examination

a) Pupillary examination

A direct and consensual reflex is to be noted, abnormal reflexes may point towards structural or organic defects. Amblyopia may be associated with very mild relative afferent pupillary defect; a gross RAPD should be deeply investigated for the cause.

b) External/ Anterior Segment examination

A gross torch-light eye examination and a slit-lamp eye examination in older and co-operative children should be done. All the abnormalities which can predispose to amblyopia should be noted and the caregiver should be informed about the risk of amblyopia.

c) Cycloplegic retinoscopy/ auto-refractometer refraction and funduscopy examination

All children with suspicion of amblyopia or risk of amblyopia should be subjected to cycloplegic refraction and fundus assessment. As children have a strong accommodative tone, age-appropriate cycloplegic agent should be used. (Table 3).

All children with asthenopic symptoms, suspicion of moderate-to-high hypermetropia/myopia and significant astigmatism should be subjected to cycloplegic refraction.

The fundus of all the children subject to dilatation should be examined. If any peripheral retinal abnormality is suspected, then the child should be subjected to indentation indirect ophthalmoscopy and/or general anaesthesia/sedation as the need arises.

d) Visual acuity, assessment of fixation pattern (in very young children)

Quantitative assessment of visual acuity should be done on any of the optotypes (letters, numbers or symbols). The choice of optotypes depends upon the age and background of the child and also on the availability of the charts. Special attention to be paid to make the child comfortable. Care must be taken that the child does not peek through the other eye. Preferably a patch may be applied to the other eye or occlusion with the palm (not fingers) may be done. In children with nystagmus, blurring of other eye rather than occlusion is preferred which can be achieved with translucent occluders or high plus lenses. The choice of chart depends upon the age of the child and availability.

In pre-verbal children or children where the quantitative assessment is not possible, qualitative assessment of fixation and following movements is needed. For assessment of fixation and following movements, a hand-held light source (torch), toy or any other accommodative target may be used. Accommodative targets are preferred as they can elicit accommodation and thereby accommodative esotropia may be unmasked. It is preferable to note the fixation and followability of each eye separately. The fixation pattern is to be noted as “fixes and follows” or “central, steady and maintained”.

Fixation preference is to be noted in terms of resentment to closure of either eye, as stronger objection or resentment to closure of an eye may point towards poor vision in the contralateral eye (Figure 6).



Figure 6: Child with right amblyopia resisting the occlusion of the better (left) eye.

If the child has strabismus, then the fixation pattern needs to be assessed binocularly and the strength of fixation of non-preferred eye may be noted. The fixation can be graded as not fixing, not holding fixation, holding momentarily, holding for a few seconds, holding through a blink or freely alternating.

e) Binocular red reflex (Brückner) test

The direct ophthalmoscope is to be shone from a distance of approximately half meter, in such a manner that the red reflex is seen in both the eyes. An equal and symmetric red reflex in both eyes is considered normal; a diminished red reflex may point towards media opacity. Brighter red reflex superiorly may point towards myopia and inferiorly may point towards hypermetropia.

f) Ocular motility, alignment and head posture

A note of the head posture is to be made when the child focuses on an object of attention or when an older child reads the 6/12 - 6/6 optotypes of the visual acuity chart. In case of doubt, the parents/ caregivers should be asked to observe the child and if possible bring photographs, in cases with history of head posture or intermittent deviations. The ocular alignment and motility is to be tested in the straight head position before cycloplegia.

g) Sensory functions to be tested before any dissociating examination are:

1. The Worth 4-dot Test
2. Binocularity/stereoacuity testing: Randot/Titmus/TNO
3. Vergence testing with a prism bar (fusional vergence)

Binocularity implies that the child is able to perceive from both the eyes simultaneously and this is adversely affected in the presence of refractive error, amblyopia and strabismus. Presence of good stereoacuity may point towards normal alignment and a good sensory function of either eye.

Sensory function (binocularity, stereoacuity) should be tested before examination involving dissociation.

TABLE-6: Minimum work-up, as per consensus at the meeting

Minimum work-up as per consensus meeting
1. Visual acuity of either eye (in case the child can read)
2. Fixation of either eye to be noted and recorded
3. Glow of either eye to look for gross refractive error and media clarity
4. Worth Four-Dot test
5. Cover and Cover-uncover test, with a note of presence of strabismus, if any
6. Bruckner's red reflex test
7. Bagolini's striated glasses test
8. Fundus examination

TABLE-7: Diagnostic criteria for Amblyopia

Criterion for unilateral amblyopia	Findings
Resentment to monocular occlusion	Asymmetric resentment - points toward poor vision in contralateral eye
Fixation preference	Failure to initiate or maintain the fixation through the blink may point towards poor vision in the eye
Preferential looking charts	> 2 octave inter-ocular difference
Best corrected visual acuity on optotypes	> 2 line inter-ocular difference
Criterion for Bilateral Amblyopia	Findings
Best corrected visual acuity on optotypes	Age < 4 years; BCVA <20/50 in either eye
	Age > 4 years BCVA < 20/40 in either eye

Prevention of Amblyopia

Screening of all the children who are at risk of amblyopia is important to identify children with a problem. It is well established that the children with refractive errors or strabismus, if identified and treated early, have better outcomes.

Table 4 mentions the refractive correction to be given in children in Indian scenario.

Amblyopia Therapy

As the child grows and attains visual maturation, the success of amblyopia therapy decreases. Also the success of amblyopia therapy is multifactorial, depending on the age of onset, cause, severity and outcome of previous treatment (if any). All children irrespective of their age should be offered the chance to be treated for amblyopia. Caregivers and older children should be made aware of the preferable treatment modality, the same to be discussed and should be encouraged to do so.

Treatment options for Amblyopia therapy

Depending upon the cause of amblyopia, the treatment should be planned. There are various modes of treatment published in the literature, some of the options include:

- Optical correction
- Occlusion therapy
- Pharmacological penalisation
- Optical penalisation (translucent or opaque lenses and filters)
- Dichoptics
- Non-conventional methods like Acupuncture
- Active Vision Therapy
- Surgery

The principle of occlusion therapy, pharmacological penalisation and optical penalisation is that they decrease the neuronal inputs from the better/normal eye thereby increasing the neuronal input from amblyopic eye and therefore partially obviating the problem of binocular rivalry/competition. They are all known to improve the visual acuity in the affected eye.

Optical correction and active vision therapy is known to improve the neuronal inputs and help in improving the visual acuity.

Optical Correction

Correction of refractive errors (if any) is the first step in the treatment of amblyopia and proper optical correction should be prescribed after an age-appropriate cycloplegic refraction.

It has been shown that correcting the image blur optically alone can improve the best corrected visual acuity in children with anisometropic and strabismic amblyopia. Spectacles are usually well-tolerated in children and it is important that the spectacles are of proper fitting, alignment and centration and, if needed, head straps and ear supports can be used to ensure a comfortable fit.

Occlusion Therapy

There are various methods of occlusion therapy being practiced. They include application of opaque patch over the skin directly, application of opaque patch over the spectacles, application of rubber patch on spectacles, pirate patch, occlusion contact lenses, etc.

The most accepted pattern of using amblyopia therapy is to apply opaque adhesive patch directly over the skin and to put on the spectacles (if any) over it. The choice of patch, whether to use the ones that are commercially available or homemade (with the help of micropore tape), is a matter of personal choice of the ophthalmologist and the caregivers/patient using it (Figure 7).



Figure 7: Occlusion therapy by application of commercially available patch over the skin with spectacles worn over the occlusion.

The various patterns of occlusion therapy administration are:

1. Full time occlusion: This means that the better/normal eye is patched for all the waking hours during the day of occlusion. To prevent occlusion amblyopia, the amblyopic eye is occluded for one day. The frequency of occlusion of the amblyopic eye depends upon the age of the patient, older the patient, greater the number of days of occlusion of the sound eye before alternating with the amblyopic eye for a day to prevent occlusion amblyopia. An example of how to advise occlusion is given in table 7. Full-time occlusion improves the visual acuity faster as compared to part-time occlusion, it may be a better mode of therapy in pre-school children; though it is associated with a greater risk of occlusion amblyopia than part-time occlusion. As the occlusion is advised for all the waking hours and either of the eye is always occluded, this may have a adverse psychological impact.

TABLE-8: Example of a full-time occlusion schedule.			
Age (completed in years) of patient at the beginning of therapy	Number of days normal/better eye occluded	Number of days amblyopic eye is occluded	One cycle of occlusion in number of days
1	1	1	2
2	2	1	3
3	3	1	4
4	4	1	5
5	5	1	6
6 and > 6 (≥6)	6	1	7

2. Part-time occlusion - This implies that the better/normal eye is patched for a given number of hours (usually 6 hours/ 4 hours or even 2 hours) during the wakeful hours of the child when they are involved with some fine visual task. In this pattern of occlusion, the child can be patch-free for rest of the day and may have lesser psychological impact on the child. Part-time occlusion is better accepted by school-going children. It is recommended that the caregivers/parents fix the number of hours at a single stretch of time so that there is no variation in proper administration of a pattern of part time occlusion.

Pharmacological penalisation

In this form of amblyopia treatment, the image input from the better/normal eye is blurred using cycloplegic agents. The basic principle here is that the image of the amblyopic eye should be clearer than the penalised eye so that the neuronal input from the amblyopic eye is better and becomes the fixing/dominant eye.

Atropine ointment 1% is the recommended agent with the dosage of twice a week; however, the dosing schedule may be as per the discretion of the treating ophthalmologist. Atropine drops may be associated with systemic absorption, hence should not be used. Although the systemic side-effects are more common in very young children, but can occur even in older children.

There have been reports of decrease in vision in the non-amblyopic eye with use of atropine, hence a constant vigil on the vision of the better/normal eye has to be kept. Besides this, there are chances of photosensitivity, allergy and anti-cholinergic side-effects with use of atropine.

Optical penalisation

Deliberately giving altered refractive correction to defocus the image in the normal/better eye has been tried; however, the results have not been promising in treatment of amblyopia.

Translucent filters, ground glass or stick on tapes on spectacles alone are not effective in treatment of amblyopia, however they can be used for maintenance and weaning off from amblyopia therapy.

Surgery

Operative procedure to clear media opacity like cataractous/ subluxated lens, non-clearing vitreous haemorrhage, corneal opacity, etc., which can hamper the amblyopia treatment needs to be done.

Refractive surgery to correct anisometropia in anisometropic amblyopia is not advocated by the panel, more so in Indian settings, however it may be of limited use in certain cases.

Active Vision Therapy

The child is encouraged to use the amblyopic eye for visually demanding tasks in the form of reading/writing, drawing, watching television/video games, along with occlusion of better/normal eye. This not only increases compliance as it acts as an encouragement to the child but may help as an adjuvant to occlusion. Minimum 1 hour daily activity should be prescribed; however, along with occlusion therapy as it has shown to be beneficial compared to occlusion alone¹².

Planning amblyopia treatment

Earlier the start of the treatment, better is the outcome. It is important to note the grade of amblyopia, type of amblyopia and age of the patient before planning the treatment.

All children irrespective of age, should be offered treatment, more so in the case of refractive amblyopia as even older children and young adults have been found to benefit from treatment in certain cases.

Proper and careful refractive correction is essential for all types of amblyopia therapy and the cycloplegic agent depending upon the age should be used to refract and appropriate refractive correction be prescribed. In case of large anisometropia or refractive error, contact lenses may be the choice of refractive rehabilitation.

Spectacle or optical adaptation time of around four weeks should be given before starting the occlusion. However occlusion can be started along with prescription of spectacles if amblyopia is dense or repeated visits are difficult. All children undergoing amblyopia therapy need to be re-assessed and re-refracted under proper cycloplegia periodically. For children less than 3 years of age, the repeat refraction needs to be done every 6 months and for older children it should be done on yearly basis. However, the cycloplegic refraction may be repeated earlier if indicated.

Occlusion is the primary modality of amblyopia treatment. Even in presence of nystagmus e.g. fusional maldevelopment nystagmus (manifest latent nystagmus), occlusion remains the mainstay of treatment. The caregiver and the child should be explained the pattern of occlusion with examples so that it is clearly understood and followed.

Pharmacologic penalisation may be used, when despite the efforts and explanation about the occlusion therapy, there is non-compliance or non-transient, non-resolving allergic issues associated with occlusion.

Dichoptics and optical penalisation may be used in mild amblyopia or in cases of maintenance of vision after amblyopia therapy e.g. in cases where the patient has completed the amblyopia therapy and is waiting for strabismus surgery or in cases where the ophthalmologist fears recurrence of amblyopia.

Strabismic amblyopia

Strabismic amblyopia may be commonly associated with refractive errors; hence proper cycloplegic refractive correction is an essential before initiation of amblyopia therapy.

The duration of occlusion therapy depends on the degree of amblyopia. In severe amblyopia, full time occlusion depending upon the age of the patient is planned. In case of non-compliance, the caregiver and the patient is re-assured, failing which part-time occlusion may be started. In moderate-to-mild amblyopia, part-time occlusion for 6 hours and 2 hours may be the choice of occlusion pattern.

Children less than one year of age, irrespective of pattern of occlusion therapy, needs to be followed up every 15 days. Older children on full-time occlusion should be followed up every 4-6 weeks, while those on part-time occlusion should be followed up in 6-8 weeks.

Children with infantile esotropia and in children less than one year of age, if full-time occlusion is advised then the follow-up should be fortnightly. In case the child is on 6 hours or 2 hours occlusion, the follow-up can be 4-6 weeks apart.

The strabismus correction is to be planned when the vision is equal in both eyes or the maximum possible visual acuity is achieved in the amblyopic eye.

Refractive amblyopia

All cases of refractive amblyopia should be given optical adaptation for 2 weeks, however occasionally; the occlusion may be started along with prescription of spectacles.

If the anisometropia is large, then contact lens may be prescribed for optical correction along with occlusion.

In moderate-to-severe amblyopia, part-time occlusion for 6 hours may be started and if there is no improvement, full-time occlusion therapy may be started.

In mild amblyopia, 2 hours occlusion may be advised as a part of occlusion therapy.

In cases of bilateral refractive amblyopia, appropriate refractive correction should be prescribed and there is no role for occlusion.

Children less than 1 year of age should be followed up every two weeks, for older children on part-time occlusion the follow-up is every 6-8 weeks.

Visual deprivation amblyopia

As this is the most severe form of amblyopia, the cause of significant visual deprivation like congenital cataract, corneal opacity, non-resolving vitreous haemorrhage needs to be first treated surgically.

In case of unilateral disease, full-time occlusion of the normal eye is to be started immediately following the surgery. Contact lens rehabilitation is preferred over spectacle rehabilitation in case of unilateral aphakia.

If the disease is bilateral and symmetrical, the need for occlusion should be assessed based on other factors like presence of constant strabismus, significantly smaller size of the eye, unilateral significant astigmatism, etc. In cases where there is no significant difference in both the eyes, proper optical correction is only necessary; however, occlusion can be started when one of the eyes has greater risk factors for amblyopia.

Young children less than 1 year of age should be followed every 2 weeks and older children every 4-6 weeks.

At every visit, the clarity of visual axes, proper optical correction, status of spectacles or contact lenses, anterior segment examination including intraocular pressure assessment has to be done. Refraction, axial length estimation and fundus evaluation is to be done on six monthly visits.

The cases where there is no improvement after institution of amblyopia therapy with adequate compliance for 2-3 consecutive visits, the diagnosis of amblyopia needs to be reconsidered. A re-look into the refractive status or revision of diagnosis may be needed and appropriate investigations as per the perceived requirement may be done.

End-point of amblyopia therapy

The desired goal of amblyopia therapy is BCVA of 20/20 vision in the amblyopic eye, however 20/30 is also considered as full visual recovery.

Amblyopia therapy is to be weaned once the BCVA of 20/20 or 20/30 is achieved and stable for 3 consecutive visits. In case of suboptimal visual gain, the best corrected stationary visual acuity for 3 consecutive visits is considered as the best achieved goal. The occlusion therapy can be tapered, the full-time occlusion either may be reduced in the duration of days of occlusion of the better eye e.g. 6:1 occlusion pattern is reduced to 5:1, then 4:1 and so forth, on each visit 4-6 weeks apart or full-time occlusion may be shifted to a part-time occlusion of 6 hours daily and then tapered off further.

The part-time occlusion can be tapered from 6 hours to 4 hours to 2 hours daily on consecutive visits 8-10 weeks apart.

As the child maintains the vision through the tapering schedule, once 1:1 occlusion pattern is reached in full-time and 1 hour daily occlusion in part-time, the patch can be stopped at the following visit. Bangerter's filter or transparent tape on spectacle of the better/ normal eye can be used just before fully stopping the occlusion. During follow-up, it must be ensured that the best visual acuity achieved does not fall.

After completion of successful amblyopia therapy, the child needs to be followed up every 6 months for the next 3 years and then yearly till at least 9-10 years of age. A comprehensive eye examination including cycloplegic refraction should be done on every visit.

Those who fail to improve on amblyopia therapy, Levodopa or citicholine may be used as an adjuvant to therapy along with occlusion¹³.

Failed amblyopes should be advised refractive spectacles and the children with no refractive error should be advised to use zero power spectacles as amblyopes have higher risk of injury to the better/normal eye. They should be regarded as one-eyed individuals and a yearly comprehensive eye exam should be done.

Conclusion

The purpose of these guidelines is to establish uniformity in the practice of paediatric vision screening, ocular examination and refraction at primary, secondary and tertiary eye care levels and provide clinical indicators for referral to a higher centre. In the presence of good synergy at all eye care levels and well-developed referral pathways, these practice patterns will enable the ophthalmologist to provide efficient paediatric eye care services and timely intervention for avoidable or preventable causes of blindness.

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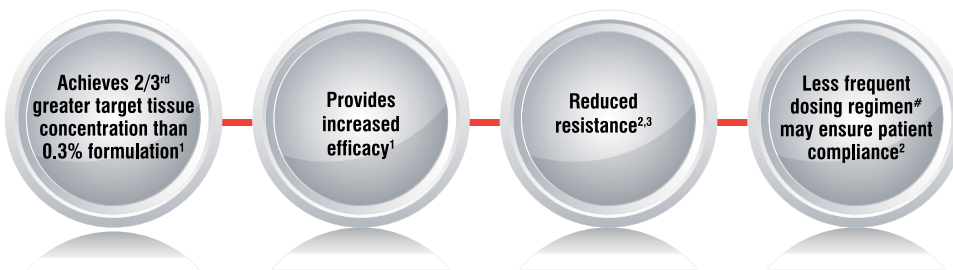
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