Child India

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CONTENT

Editor's Note

Dear friends,

This August issue deals with various topics related to pediatric ophthalmology.

Pediatric ophthalmology is quite a fascinating branch and the remarkable advances in the last 25 years have seen it emerge as a distinct specialty. Advances in diagnostic and imaging techniques such as optical coherence tomography (OCT), widescreen digital imaging (Optos), and modern fluorescein angiography have expanded the options available for investigation of children. Application of new diagnostic techniques for deep phenotyping in combination with genomic testing has enhanced the accuracy of diagnosis and prognosis.

Childhood blindness is a terrible malady that occurs 5 times more in India than in the developed world. In certain parts of India, the causes are similar to those seen in poor countries of the world and predominantly include preventable causes (mainly corneal blindness). In urban areas, conditions such as retinopathy of prematurity, cerebral visual impairment, retinal dystrophy, etc., are seen, which are similar to those prevalent in middle-income countries. Apart from this, other treatable conditions such as pediatric cataract and glaucoma are also seen. Overall, approximately 60%–70% of blindness in children is avoidable and/or treatable. The remaining 30% of children may require low vision and rehabilitation care to improve their quality of life.

Children do not know how to show their gratitude for treatment received and may or may not be able to express themselves, irrespective of whether the treatment provided was excellent or poor. Hence, the onus of optimum and efficient care, purely lies in our hands. To make sure that each and every child receives appropriate treatment, pediatric eye care professionals need excellent training.

We thank all contributors, the OB and members of the Strabismus and Pediatric Ophthalmology Society of India and specially Dr Elizabeth Joseph for the string of articles that make this issue of Child India akin to a textbook of Pediatric Ophthalmology.

Please browse through for an overview of the subject

We celebrate World Breastfeeding Week August 1st to 7th 2021 with the theme - Protect Breastfeeding: A Shared Responsibility. The theme is aligned with thematic area 2 of the WBW-SDG 2030 campaign which highlights the links between breastfeeding and survival, health and wellbeing of women, children and nations.

Jai IAP!

Dr Jeeson C Unni
Editor-in-Chief

President's Address

Dear colleagues,

What is the status of Pediatric Ophthalmology training in our country?

It is estimated that close to 70% residency (postgraduate) programs do not have a structured rotation and surgical training in pediatric ophthalmology and this is an understatement.

As per WHO, one children's eye care center is required for every 10 million people, where at least one specialty-trained or oriented ophthalmologist should be available. This warrants immediate doubling of the number of dedicated children's eye care units across India. As on 2017-18, only 11 ophthalmic institutions in India offered fellowship training in pediatric ophthalmology and strabismus (POS). Roughly 28–30 POS fellows are trained every year, which is far below the required number.

A nationwide comprehensive survey of young ophthalmologists (trained in the 21st century) reported that while cataract and retina subspecialties were fairly well covered during residency training, the same could not be said about strabismus and pediatric visual acuity testing. The median strabismus surgeries performed independently by trainees is negligible. They are better trained in use of A-scan biometry, B-scan ultrasonography, optical coherence tomography, pachymetry, automated perimetry, fundus photography, fluorescein angiography, and use of lasers.

The lacunae are not on the ophthalmology curriculum, but failure on part of faculty in teaching and creating interest in the subject. If this happens appropriately, an opthalmologist should be able to treat the common diseases such as refractive errors, cataract and amblyopia in children and learn to refer them appropriately which will definitely increase the quality of services apart from reducing the burden on pediatric ophthalmologists.

Good fellowship programs, inspiring role models in pediatric ophthalmology, and the massive demand for children's eye care in India should motivate future ophthalmology residents/postgraduates to join this subspecialty in the years to come thereby making a positive impact on the current status of the specialty in India.

Neonatologists and pediatricians have the onus of preventing premature babies from going blind secondary to retinopathy of prematurity, blindness associated with malnutrition, diagnose and treat ophthalmia neonatorum, xerophthalmia and conjunctivitis, pick up ophthalmic manifestation of systemic disease, etc, We hope this issue will help fellow IAPians to be proactive in working in tandem with pediatric ophthalmologists to provide comprehensive eye care to children.

Jai IAP!

Piyush Gupta
National President, IAP 2021

Secretary's Message

Dear All,

Greetings!

It has been an eventful month at the IAP Child India August 2021. We had a very successful Administrative Meeting via Video Conferencing with the IAP Office Bearers on 3rd August 2021. My heartfelt thanks to everyone involved as a participant in this meeting.

We have planned to conduct IAP Executive Board Meeting on 22nd August 2021 and many other committees that met this month like IAP Election Commission and IAP Tribunal Committee, IAP Grievances Committee, IAP E-voting Committee.

This month, we have conducted 4 Zonal ToTs of our flagship program Nurturing Care – Early Childhood Development physically at Nagpur (West Zone), Delhi (North Zone) Hyderabad (Central Zone) Bhubaneshwar (East Zone) and Kochi (South Zone) is scheduled to be held on 5th September 2021.

Also we have conducted 1 East Zone ToT and total 12 workshops of Mission School Uday program in the month of August 2021. We are organizing virtual events all over India. There are many more such programs in the pipeline for upcoming days.

I also would like to mention about the Dysbiosis for PG workshop which organized excellently by Assam State, Karnataka-Bangalore Region, Andhra Pradesh State, Bihar State, Kerala State in the Month of August 2021. I am very happy to mention that, we also conducted 7 Physical and 2 virtual program of NTEP, 7 CMEs of DERMA Module, 5 workshops of CADE module in Month of August 2021. There are many more such programs in the pipeline for upcoming days.

It gives me immense pleasure to inform you all that the Bangalore Pediatric Society and IAP branch –MORENA, Madhya Pradesh Celebrated World Breast feeding week from 1st to 7th August 2021. Also we conducted National Nursing quiz on ORS and Breast feeding and IAP-UNICEF webinar on Current Trends in Pediatric AIDS has been successfully conducted on 8th August 2021.

Overall, the month of August 2021 has been very fruitful and focused on academic growth for IAP members and we look forward to having more such activities in the coming months.

Jai IAP!! Jai Hind!!

Sincere Regards,

Dr G V Basavaraja
Hon. Secretary General 2020 & 21

Infantile Esotropias, A Pediatrician's Perspective

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Introduction - Strabismus and amblyopia are one of the most prevalent health problems in children in the developing world affecting up-to 5% of their population. Infantile strabismus (strabismus starting in first year of life, not congenital) will likely affect 1% of healthy full-term newborns. Strabismus is an epiphenomenon of evolution of single binocular vision. The frontal eyed binocular single vision in primates, birds and carnivores evolved from the 2 parallel fully crossed monocular visual systems in lower lateral eyed animals with fully crossed chiasma with right eye represented in left cerebral hemisphere and vice versa. It likely happened to incorporate stereopsis in the monocular visual plant. Slowly a fully crossed chiasma proselytized to a half -crossed chiasma with overlapping of temporal and nasal visual fields of two eyes and their fusion into a single binocular image. The right side of the brain encodes for the left field and vice versa. To accomplish this, phenomenal changes in the neuronal wiring had to be executed, starting from the lateral geniculate body to the occipital cortex and associated visual areas in the temporal and parietal cortex, brainstem and the cerebellum. The retractor lentis and retractor bulbi became extinct. It transpires that the neonate is sui generis born with 2 parallel monocular visual systems, quite like lower lateral eyed animals, the binocular wiring has to take place in layers 2,3 for fusion and stereopsis and 4b for motion vision processing in the visual cortex in the 6 layered occipital lobe, calcarine cortex. Stereopsis is quintessentially detection of horizontal disparity in the Panum's fusional area where fusion can take place. If it crosses the dimensions of Panum's fusional area, the images can't be fused and will be seen as double warranting further sensory and motor adaptations. Strabismus is thus not only unique to frontal eyed animals but also a product of it and connotes a failure / derangement of binocular single vision.

Etiopathogenesis - Binocularity is thus a learned behavior, quite like language which can only take place if minimum optimal prerequisites are made available. The first requisite is that being foveate animals, we have to learn to fixate for further cascade of events and afferent input has to be similar to 2 eyes. The neonate takes about 6 weeks to develop a stable fixation reflex, any alignment before this period is totally instable and should not be commented upon. Most of infants will have an apparent exotropia with eyes teetering from eso to exo position. There is an inherent esotonus which is trying to drive the eyes into an eso- position towards an orthotropia from an built in exotropia of lateral eyed animals. If for some reason, the binocular wiring goes hey wire or is aborted, an infantile deviation is likely to emerge. Mostly these are esodeviations but exodeviations can also emerge in about 10% of cases. The other fellow travelers for failure of binocularity are smooth pursuit asymmetry, primary inferior oblique overaction, dissociated vertical deviation and latent nystagmus. While

evaluating these deviations it's imperative to look for these co- conspirators as well to nail the entity.

Infantile deviations, ipso facto are a different cup of tea as binocularity did not get a chance to develop or it was stalled midway, it's like return to lateral eyed two parallel monocular visual systems with all it's attendant phenomena and epiphenomena. It's intuitive to keep these aspects in mind as these are distinct motility problems with some kind of atavism to boot. We shall share some of the salient features of these deviations to impress upon early detection, differential diagnosis and early intervention to help retain and strengthen binocular single vision in the infant's sensitive period for visual development. It's intuitive to internalize that these deviations are not congenital but acquired in infancy by about 6 months of age and aptly called infantile deviations.

Epidemiology and risk factors - Genetic as well as environmental factors both play a role in etiopathogenesis. Large scale studies have shown that 20 to 30 % of children born to a strabismic parent will have strabismus. The prevalence of strabismus in low birth weight and premature infants rises exponentially as the birthweight plummets. The occipital cortex in newborns are particularly susceptible to hypoxia. Infants born 1500 g or less have 7 times higher prevalence of amblyopia and strabismus. Infants weighing less than 2500 g have a prevalence of strabismus 4 times higher than normal weight infants. Conditions that predispose to infantile esotropias are occipito-parietal hemorrhage, periventricular leukomalacia, intraventricular hemorrhage, hydrocephalus, Down's syndrome and many others. The role of pediatricians is very vital in diagnosing and treating systemic associations of infantile esotropia including low birth weight, prematurity, periventricular leukomalacia, cerebral palsy, autism spectral disorders, Down's syndrome and many other associated conditions.

Salient clinical features - Infantile esotropia, as stressed earlier is not present at birth, if it's present at birth, one has to think of Duane's syndrome, 6th nerve palsy, congenital fibrosis of the extraocular muscles and Moebius syndrome, just to name a few. Typically, it appears by 3 to 4 months, initially the deviation is intermittent but as time passes, it gets established. An esodeviation established by 6 months is the typical presentation and unlikely to resolve. Any deviation exceeding 40 prism diopters by 4 months is also unlikely to resolve and has to be interfered with appropriately. The deviations are constant and large angle with free alteration, amblyopia is not usual unless driven by anisometropia or other amblyogenic factors. The infant cross fixates so there could be apparent abduction deficit which may be confused with 6th nerve palsy, if not evaluated properly with Doll's eye movements or after alternate occlusion / patching. Other factors that may contribute to face turn and abduction deficit are pursuit asymmetry and latent nystagmus.

Infants who fail to develop normal binocularity exhibit smooth pursuit asymmetry. When one eye is occluded and a handheld toy is moved from temporal to nasal before the fixing eye, pursuit is smooth. Pursuit is absent or jerky or cogwheel when target moves nasal to temporal. The movement of two eyes are conjugate with direction of the asymmetry changing with change of the fixing eye. The asymmetry is seen transiently in all healthy children. When it persists beyond 6 months, it reflects delayed development of binocular connections in the visual cortex and is definitely pathological if it persists beyond one year. Another feature of inappropriate pursuit is the latent nystagmus, the deranged pursuit takes the eye off the target, visual acuity is degraded, a saccadic refixation occurs and the cycle is repeated. Accordingly, when attempting to fixate, the eyes drift nasally in respect to the fixing eye. The velocity of the slow drift and the number of corrective fast phase jerks (corrective saccades) are accentuated by covering the fellow eye hence

aptly termed manifest latent nystagmus, though the term may sound oxymoronic. Although latent nystagmus is associated with infantile esotropia, quite like smooth pursuit asymmetry, it may develop in any situation that preempts binocular wiring in the occipital cortex in first 6 months of life. Entities may encompass monocular cataracts, infantile glaucoma, corneal opacities, significant anisometropia, infantile exotropia or a vertical tropia present since birth. Motion visual evoked potential asymmetry offers additional insights that directional asymmetry of smooth pursuit and latent nystagmus is due to maldevelopment of the visual cortex and other association areas of the brain.

Infants with latent nystagmus and pursuit asymmetry prefer to view targets by positioning the eye at a more nasal position in the orbit. This is achieved by a face turn towards the fixing eye. The reduced intensity of the nystagmus improves visual acuity in the fixing eye and it alternates with change in the fixing eye. A consistent face turn may denote amblyopia in an eye that is in a more temporal position. The abduction deficit is moderate and is usually overcome by Doll's eye movements or by alternate patching for a few days. If it does not improve with alternate patching, conditions like Duane's retraction syndrome should be considered.

Additional factors that may contribute to incomitance may be observed, the most common is V pattern with (out) primary inferior oblique overaction, an A pattern can also be seen with (out) primary superior oblique overaction. An accommodative component may be riding piggyback, it's imperative to carry out refraction under full cycloplegia using 1% Atropine eye ointment or drops and have a detailed fundus evaluation as fundus pathologies giving rise to poor vision and strabismus, could coexist and are frequently classed as sensory esotropias. Accommodative esotropia due to high accommodative convergence / accommodation ratio may also add to eso deviation and has to be detected and addressed. The deviation has to be assessed after accommodative component has been taken care of.

Dissociated vertical deviations (DVDs) and Primary inferior oblique overaction (PIOOA)

Dissociated vertical deviation may be an atavistic response to dorsal light reflex seen in lower lateral eyed animals as and when establishment or sustenance of binocularity is preempted. The eye receiving less illumination rises and the one with more falls, DVD violates Hering's law. DVD may not be present in the initial phases of infantile ET, it may surface after a year or so, despite esotropia having been satisfactorily treated surgically, implying that it reflects weak binocular connections in the cerebral cortex. DVD is invariably bilateral and asymmetric. A head tilt to the side of the less affected eye is frequent, creating diagnostic conundrum with congenital superior oblique palsies.

Primary inferior oblique overaction may have similar underpinnings. The primary function of the obliques in lateral eyed animals like fish is wheel rotation of the eyes in response to vestibular stimulation as the animal pitches up or down. With attainment of frontal eyed binocular vision theses vestibular reflexes are kept in abeyance and obliques have to conform to Donder's and listing's laws, constraining torsional movements to only about 7.70 of torsion so that binocular single vision can take place. The inferior obliques were depressors and superior obliques were elevators in lower lateral eyed animals, with transition to binocular single vision, the roles were reversed in the vertical plane with inferior obliques as elevators and superior obliques as depressors. It follows that oblique muscle overaction can result from abnormal central vestibular recruitment whenever binocularity is preempted, a physiological reflex gone awry? The eye elevates in adduction,

abducts and extorts, engendering clinical picture of PIOOA with V pattern. The opposite superior oblique overaction can also take place. The high prevalence of disorders of balance and motor incoordination in infantile esotropia also lends credence to the premise that early loss of binocularity may affect central vestibular tone. There is an inherent bias for upward movement due to anterior canal dominance, kept in check by cerebellar flocculus. Whenever binocularity is preempted, these inhibitory influences are tuned off ending up in PIOOA.

Functional deficits in infantile esotropia may further include absence of motor fusion, lack of sensory fusion and stereopsis, alternating monocular suppression and subnormal binocular visual evoked potential responses. Defects of motion vision processing include asymmetric monocular tracking, asymmetric monocular motion visual evoked potentials and asymmetric motion perception.

Differential diagnosis - The clinical picture is quite characteristic however certain conditions should be kept in mind as differential diagnosis. Early accommodative esotropia, Duane Retraction syndrome, congenital fibrosis of the extraocular muscles (CFEOMs), Moebius syndrome, sensory esotropias among others.

Management - Refractive errors should be corrected as a first step. Hypermetropia > +2.5 D, anisometropia >1.5 D, cylinder of > 0.5, myopia exceeding -4 D need to be corrected. Correction of refractive errors may impact the deviation and amblyopia especially if there is a component of accommodative esotropia riding piggyback on infantile esotropia. Invariably these are freely alternating esodeviations so amblyopia is not a common occurrence, if it's there, it's likely due to anisometropia or meridional. Amblyopia has to be suspected, diagnosed and treated on priority basis by occlusion therapy and other means as appropriate for optimum results. The optimum age group for treating amblyopia is up-to 7 to 8 years. Systemic associations like neuro-developmental delay, cerebral palsy, periventricular leukomalacia, autism and many other systemic associations, if present have to be diagnosed and treated appropriately. The role of pediatricians is of great import in managing these enigmatic entities.

Early surgery in the form of bilateral medial rectus recession is the norm. Since the deviation may vary in the initial period and may get stable by 6 moths only, surgical interventions are usually deferred till then. However if the deviation is fairly constant, over 40 PD by 4 months of age, that calls for surgical intervention without further delay. In order to retrieve and sustain some binocularity and stereopsis, surgery should not be delayed beyond 2 years. Children whose eyes are aligned to within 8 prism diopters by 16 months of life have restoration of random dot stereopsis and those aligned by 12 months have finer grade stereopsis. Additionally defects in motion pathways can also be repaired by early surgery. It transpires that the defects of cortical binocular wiring can be repaired to a reasonable degree by early surgery. More than one surgical procedure may be required to achieve satisfactory ocular alignment. Botulinum toxin injections to medial recti have also been used in isolation as well as as an adjunct to strabismus surgery with modest results.

The converse, Infantile Exotropia and it's associations with neuro-ophthalmic disorders - The ratio of infantile esotropia to exotropia is 10 : 1, As opposed to majority of children with esotropia who may not harbor any systemic disorder, 90% of children with exotropia have significant eye or brain abnormalities. The list is long, may include conditions like optic nerve hypoplasia, morning glory syndrome, retinoblastoma, microcephaly, craniofacial disorders like Crouzon's, albinism, congenital nystagmus, prematurity, developmental delay, cerebral palsy, hydrocephalus and Prader- Willi syndrome to name a few. The exceptions are early onset intermittent exotropia, sensory exotropia

and normal infants upto 2 to 4 months who may display a transient physiologic exotropia in early infancy before fusion lock is well established. The management is on similar lines with bilateral lateral rectus recessions and surgery on the obliques and vertical recti for oblique muscle overactions and DVD. Systemic aspects have to be screened and addressed appropriately.

Suggested readings

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Abnormal Head Posture in Children - A Short Review

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An abnormal head posture is when the head is not held in the primary straight head position.It is a common condition in children with an estimated incidence of 1.3%. Abnormal head posture has three components , namely, a face turn to right or left, chin up or chin down, and head tilt to the right or left. These may exist alone or in combination. The abnormal position of the head can be due to an ocular or a non-ocular problem. Also it can be congenital or acquired . A sudden appearance of abnormal head posture should always be managed on an emergency basis THE

OCULAR CAUSES

The ocular causes of abnormal head posture are incomittant strabismus, nystagmus , astigmatism, ptosis and gross defective vision in one eye.

1) **INCOMITTANT STRABISMUS:** In Incomittant Strabismus the amount of eye misalignment varies with different gaze positions. The person will typically place his or her head in a position where the eyes are best aligned. This will help to eliminate double vision and/or reduce eye strain.

For example, if a superior oblique muscle is weak (such as in a fourth nerve palsy) a person will tilt the head away from the affected eye because the eyes are most straight in this position. Similarly, a sixth nerve palsy results in a weakened lateral rectus muscle and a face turn toward the affected eye. Sometimes the eyes are straighter in up or down gaze and a person will tilt the head up or down depending on where the eyes are best aligned.

Pre operative photo of Child with Right 4th nerve palsy with head tilt to left

Post operative straight head posture

This child had congenital Right Superior Oblique palsy which was treated with S O Tuck and Inferior Oblique weakening which resulted in correction of abnormal head posture.

Other causes from strabismus include Duane's syndrome, Brown's syndrome, orbital wall fractures, and restricted eye movement associated with thyroid eye disease. Sometimes children with rheumatoid arthritis may develop acute diplopia , restricted eye movement and head posture due to acquired brown syndrome. They are often amenable to treatment with non steroidal anti inflammatory agents or systemic steroids or steroid injection into the affected muscle[Superior oblique].

2) NYSTAGMUS : Some patients with nystagmus will acquire a head turn or tilt if the nystagmus nullpoint is away from the primary position . Here in order to bring the null point to the centre, the child will develop a certain head position. The head position where the nystagmus is slowest, or even stopped, is called the null point. Decreased nystagmus allows for better vision. The abnormal head posture due to nystagmus is most successfully corrected by extraocular muscle surgery, aimed at bringing the null point to the primary position. Various surgical procedures are available.

3) DIFFERENCE IN VISION BETWEEN THE EYES: Sometimes a child will turn the head to place an eye with better vision closer to the target.

4) PTOSIS: A child with ptosis (or a droopy eyelid) will usually elevate the chin to help the eye or eyes see "beneath" the droopy eyelid.

5) REFRACTIVE ERRORS: At times a child may turn their head to the side if they have a significant need for glasses, particularly astigmatism. Here the head posture will disappear with the use of correct spectacles.

Children adopt a head posture to improve their vision if the head turn is from an ocular cause. Therefore, in general, it is important to not discourage the abnormal head posture in these children until the problem can be fixed .Prompt reference to an ophthalmologist to pick up and treat an ocular cause is very important.

Significant abnormal head posture could cause permanent tightening of neck muscles that can lead to chronic neck ache or headache. An abnormal head posture may also cause the facial bones to grow abnormally leading to facial asymmetry.

The photographs of patient with acute head posture and defective ocular movement due to acquired brown syndrome, treated with systemic and para trochlear steroids

NON-OCULAR CAUSES OF AN ABNORMAL HEAD POSTURE

Congenital shortening of the neck muscles (sternocleidomastoid) can cause a head tilt. This is typically called congenital torticollis.Brachial plexus injury and Kippel –Fell anomaly also may have toticolis.

Other non-ocular causes of an abnormal head position include cerebral palsy, bony abnormalities, occipitocervical synostosis , otitis media, unilateral hearing loss, absent cervical muscles, benign paroxysmal torticollis of infancy,psychomotor delay , focal dystonia etc, etc.

DIAGNOSTIC TEST TO DIFFERENTIATE OCULAR FROM NONOCULAR CAUSES

A patch test in the office can confirm this diagnosis. If the head tilt is due to tight neck muscles, then the tilt will remain the same with the patch on. If the head tilt is from an ocular cause such as strabismus, then the tilt should get better when the patch is worn.

CONCLUSION. Evaluation and treatment of a child with abnormal head posture will require a team approach involving the paediatrician , ophthalmologist, ENT surgeon, Neurologist and Orthopedician. Prompt reference is crucial , many a times.

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Through the eyes of a Paediatrician... Understanding Retinopathy of Prematurity

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Retinopathy of Prematurity (ROP) is a vasoproliferative disorder of the eye that affects infants born four or more weeks preterm and have received intensive neonatal care. [1,2] . In India the incidence of ROP is between 38 and 51.9% in low-birth-weight infants. [3,4] ROP is on a rise in India as a result of improved neonatal care and better neonatal survival rate. Being clinically silent in the neonatal period, ROP needs to be diagnosed by screening and treated promptly before progressing to a sight-threatening stage.

Normal retinal vascularization happens centrifugally from optic disc to ora. Vascularization up to nasal ora is completed by 8 months (36 weeks) and temporal ora by 10 months (39–41 weeks) as shown in figure 1.[3] Because retinas of premature infants are incompletely vascularized, ROP incidence and severity are directly proportional to the degree of prematurity. Moreover the stimulus for the abnormal growth of blood vessels comes from the peripheral immature retina.

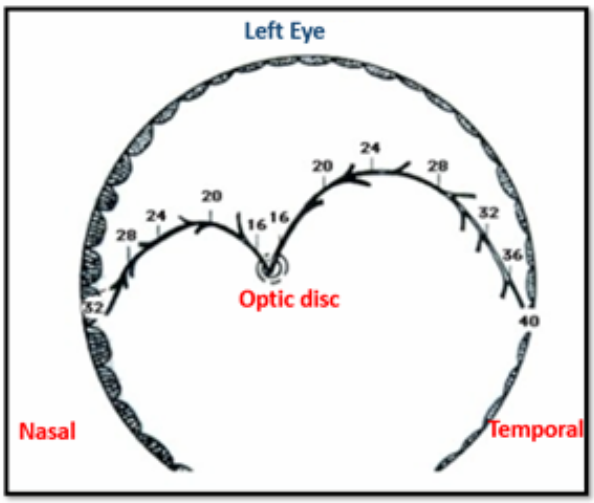


Fig 1. Normal retinal vascularization- Starts at the optic disc at approximately 16 weeks of gestation and proceeds peripherally. The vessels reach ora serrata (the periphery of the retina) on the nasal side at around 34 to 36 weeks and temporal side by 36-40 weeks. The numbers in the figure are in weeks of gestation.

Objectives of this review

To create awareness and expertise among the neonatal care providers regarding the vitality of ROP screening and timely treatment. The clinical questions for these guidelines include:

a. How does ROP occur?

b. Who should be screened for ROP?

c. When to screen for ROP?

d. How should these babies be screened for ROP?

e. How to diagnose ROP and recognise the severity of the disease?

f. What should be the frequency of Screening after the initial examination?

g. How should they be managed?

h. How to follow up these babies?

a. How does ROP occur?

Pathogenesis of ROP

The development of ROP progresses through two phases. The first phase (Vaso- obliterative phase) begins when retinal vascular growth ceases after premature birth. During this time the vessels are particularly vulnerable to injury and may be obliterated by any number of stress factors including the amount of oxygen supply. This relatively vascular-depleted retina which becomes increasingly hypoxic by an increased metabolic demand of the developing retina triggers the vasoproliferative or second phase (Vasoproliferative phase) of ROP. The formation of these new vessels is anarchic and excessive resulting in invasion of the vitreous, wherein traction on the retina and bleeding can occur. [5,6] This critical stage of ROP occurs most frequently around 33– 34 weeks post conceptionally. [7]

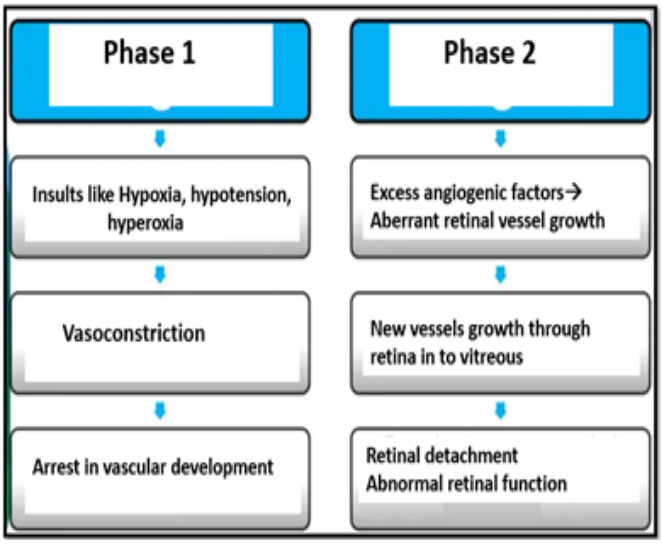


Fig 2- Flowchart depiction of pathogenesis of ROP in 2 phases

b. Who should be screened for ROP?

Whom to screen	
Screening should be carried out for the infants with either of the following:	
1. Birth weight less than 2000 gm	
2. Gestational age less than 34 weeks	
3. Gestational age between 34 to 36 weeks but with risk factors such as:	
a) Cardio-respiratory support	b) Prolonged oxygen therapy
c) Respiratory distress syndrome	d) Chronic lung disease
e) Fetal hemorrhage	f) Blood transfusion
g) Neonatal sepsis	h) Intraventricular haemorrhage
j) Poor post- natal weight gain	
4. Infants with an unstable clinical course who are at high risk (as determined by the neonatologist or paediatrician).	

Table 1- First time screening protocol for immature infants.

c. When to screen for ROP?

The criteria for ROP screening have been provided from the two large clinical trials – the multicentre trial of cryotherapy (CRYO-ROP) study and the light reduction in ROP (LIGHT-ROP) study. The scheduling of ROP screening examinations should ensure that eyes likely to need treatment are identified in a timely manner while, at the same time, minimizing the number

of examinations for infants at low risk. Thus the timing of first examination should be based on postmenstrual age (GA plus chronological age) rather than postnatal age. To simplify, ROP screening should be done at 4 weeks after birth. However, if any baby was delivered earlier than 28 weeks of gestation or birth weight less than 1200 gm should have a ROP screening at 2-3 weeks after birth.[4]

When to screen?	
All premature infants	4 weeks of birth
Gestational age < 28 weeks Birth weight< 1200 grams	2-3 weeks of birth

Table 2- Timing of screening immature infants for ROP.

d. How should these babies be screened for ROP?

Place of examination- It is not wise to transport small babies to ophthalmic outpatient or ward for examination. Neonates are best examined in the neonatal unit itself under supervision of attending paediatrician/ neonatologist, monitored by pulse oximeter. The baby should be swaddled and preferably fed one hour prior to examination. Incubator dependant babies can be screened (and even treated) within the incubator itself through the slanting wall without disturbing the equilibrium of the infant. [8] If, at any time during the eye examination, the infant develops significant bradycardia or severe oxygen desaturation, the examination may need to be interrupted until the variables return to the baseline range and the baby recovers.

If babies are being screened at the Ophthalmologist's office, there should be arrangement for basic resuscitation equipment. Place should be warm enough and clean. Special attention should be given to the possibility of bradycardia, arrhythmia, asystole, hypoventilation, apnoea, or aspiration.

Regional shortages in the availability of ophthalmologists to provide ROP care are an important barrier to ensuring appropriate ROP screening. Retcam has revolutionized the technique for screening of ROP in this regard. Retinal images taken by trained nurses or technicians can be stored, transmitted to expert, reviewed, analyzed and sequentially compared over time and are useful for telemedicine purposes.

Fig 3a. Critically ill should be screened at NICU in presence of neonatologist monitored by pulse oximeter. 3b Nesting (wrapping) of infants can significantly reduce the stress during screening procedure.

Instrument checklist

All instruments used for screening should be clean and sterile. Following instruments are generally used:

- Indirect ophthalmoscope preferably wireless one
- 20, 28 or 30 D lens (28D or 30D lens are usually preferred as they allow easier viewing of the peripheral retina)
- Alfonso speculum and Infant scleral depressor
- Dilator eye drops
- Topical anaesthetic eye drop (proparacaine 0.5%)
- Topical antibiotic eye drop e.g. Tobramycin
- Sterile cotton and glove
- ROP documentation sheet
- Wide field Digital camera

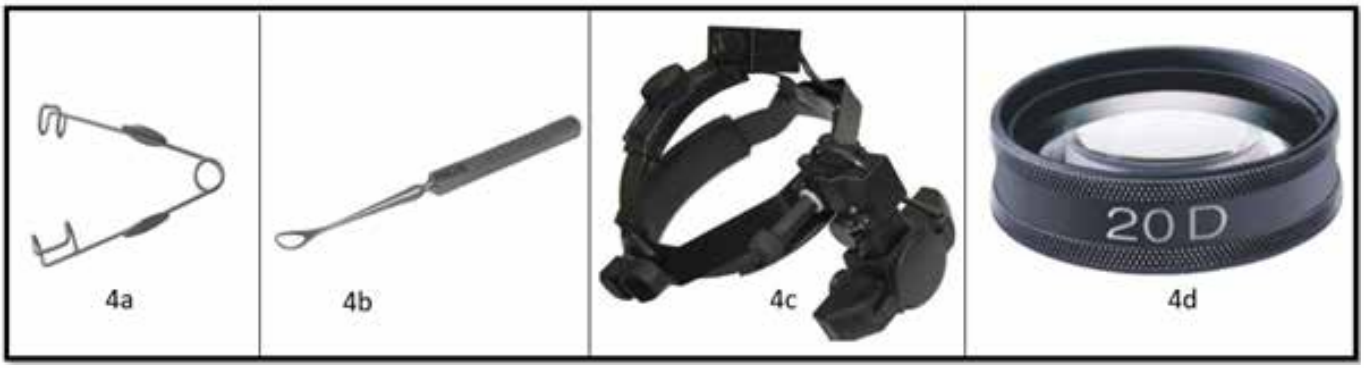


Fig 4- Instruments of ROP screening 4a. Alphonso infant speculum, 4b. Scleral depressor, 4c. Wireless indirect ophthalmoscope, 4d. 20 D Lens.

Preparation of the eye

Effective mydriasis of the pupil is essential as a well-dilated pupil enables the periphery of the retina to be examined and facilitates accurate diagnosis and staging of ROP. Mydriatic eye drops are either parasympathetic blockers which affect the pupillary sphincter muscle (e.g. tropicamide, cyclopentolate) or sympathetic stimulants which affect the pupillary dilator muscle (e.g. phenylephrine).[9]

Pupillary dilatation should be performed about an hour prior to screening. A typical mydriatic regimen consists of a combination of cyclopentolate 0.5% and phenylephrine (2.5%) drops used two to three times about 10-15 minutes apart. Tropicamide 0.5-1% is an alternative to cyclopentolate. Atropine should not be used for dilatation. Excess eye drops should be wiped off to prevent systemic absorption though the cheek skin. Over dosage carries the risk of tachycardia, hypertension and hyperthermia and must be avoided. A nondilating pupil could indicate the presence of tunica vasculosa lentis and must be confirmed by the ophthalmologist before undue excess medication for dilatation is administered. [10]

e. How to diagnose ROP and recognise the severity of the disease?

The revised 2005 International Classification of Retinopathy of Prematurity, which describes ROP according to location (zones) and severity of abnormal vascularization (stages), is used to classify and record the ophthalmologist's findings based on retinal examination.[11]

Location (Zone)

The retina is divided into three concentric circles, each centered on the optic disc. Since the retinal vessels grow out from the optic disc to the periphery and the designation of zones corresponds to the vascular developmental pattern. Disease involving Zone I is the most dangerous.

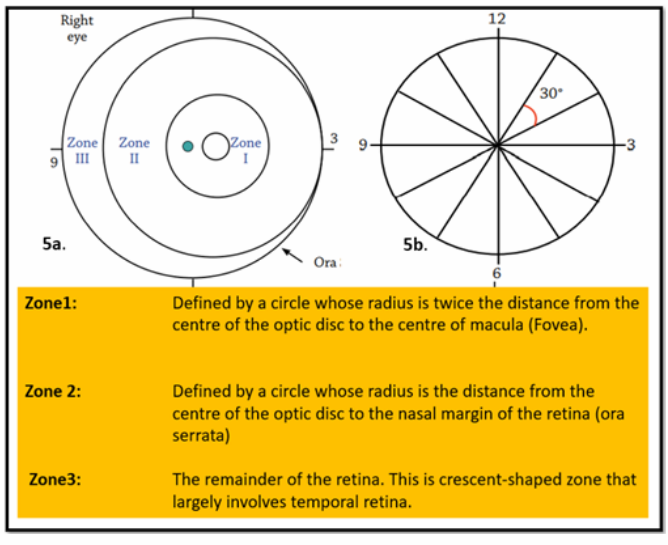
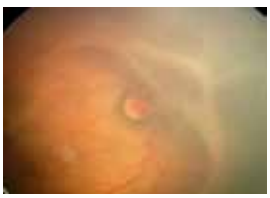
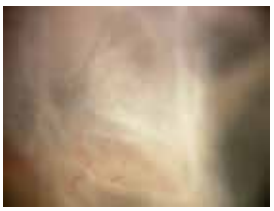


Fig 5. Diagrammatic representation of retina of the right eye showing borders of the three zones (5a) and the extent of involvement in clock hours or 30 degrees (5b), used to describe the location and severity of retinopathy of prematurity.

Extent

Disease extent is recorded as clock hours 1-12 hours or as twelve 30° sectors or 360°. The clock hours recorded is the total clock hours involved, not just the contiguous sectors a shown in fig 5b.

Stages of ROP

Stage 1 *Demarcation line*	Thin structure separating the avascular retina anteriorly from the posteriorly vascularized retina	
Stage 2 *Ridge*	Ridge increases in dimensions (height and width) and extends above the retina.	
Stage 3 *Extraretinal fibrovascular proliferation*	Neovascularization extends into the vitreous from the ridge	
Stage 4 *Partial retinal detachment*	Depending on the extent Stage 4a- extrafoveal retinal detachment Stage 4b- involving fovea	
Stage 5 *Total retinal detachment*	Generally tractional but may occasionally be exudative Usually funnel-shaped	

Additional features indicating severity of ROP

Plus disease

Plus disease is an ominous sign indicating severity of the disease. It can be associated with any stage or zone of ROP and indicates the necessity towards treatment. Plus disease is characterized by arteriolar tortuosity and venous engorgement of the posterior pole, iris vascular engorgement, pupillary rigidity, and vitreous haze.

Aggressive Posterior ROP (APROP)

Previously called Rush disease, is a very rapidly progressing severe form of ROP. If not treated timely, it will rush to stage 5 skipping the various classical stages described earlier and hence the name. The characteristic features of this type of ROP are its posterior location, prominence of plus disease, and the ill-defined nature of the retinopathy.

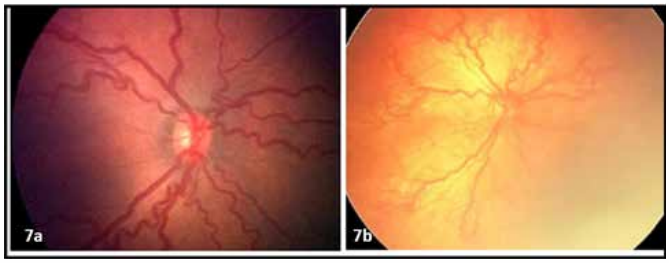


Fig. 7a - Image of the posterior pole with plus disease showing tortuosity and dilatation of the vessels. 7b- Showing features of aggressive posterior ROP.

f. What should be the frequency of Screening after the initial examination?

According to the Guidelines for Universal Eye Screening Including Retinopathy Of Prematurity In Newborns [11], the revised followup protocol according to the various stages of ROP is summarized in the table below.

1. No signs of ROP:		Follow up examination for infants at risk should be done 2-3 week intervals until the retina is fully vascularized
2. If ROP is present:		
	Zone 1:	Stage 1, 2 or 3 ROP without plus – Weekly follow-up Immature vasculature –once in 1-2 weeks
	Zone 2	Stage 1 ROP - once in 2 weeks Stage 2 ROP without - once in 1-2 weeks. Stage 3 ROP without plus should be screened at least weekly Immature vasculature- 2-3 weeks
	Zone 3	Stage 1 or 2- once in 2-3 weeks

Table 3.- Follow up schedule of preterm infants after initial screening.

g. How should they be managed?

Laser treatment

Laser is done using indirect laser ophthalmoscope under topical anaesthesia after pupillary

dilatation under care of an anaesthetist in the operation theatre.[13] The entire avascular retina up to the ora serrata should be ablated. Heart rate and apnoea spells should be monitored throughout the laser.[14] Follow-up visits after laser treatment are usually weekly till the ROP regresses and involution of all tractional elements is seen and vascularization reaches the temporal ora.

Anti-Vascular Endothelial Growth Factor (Anti-VEGF)

Anti- VEGF has evolved as an effective option in ROP especially those with severe disease or not responsive cases to convention laser treatment. BEAT ROP study which compared bevacizumab monotherapy with conventional laser therapy showed promising results for stage 3 plus ROP in zone 1 but not in zone 2. [15] In this study, peripheral retinal vessels continued in normal fashion after treatment with intravitreal bevacizumab. The current Indications for anti-VEGF treatment in ROP include.

1. Primary therapy for aggressive posterior zone 1 disease
2. Media haze due to aggressive posterior disease to improve visualization for laser treatment.
3. Failed laser treatment leading to persistent neovascularization, tractional elements or tractional retinal detachment prior to surgery

Vitreo-Retinal surgery

Surgery is done for tractional retinal detachment repair as seen in Stages 4 and 5 ROP. The aim for surgical intervention in Stage 4 ROP is to prevent progression of retinal detachment.

h. How to follow up these babies?

ROP babies are at higher risk for developing macular heterotopia, high myopia, amblyopia, strabismus, anisometropia, glaucoma, and phthisis bulbi. Assessment of vision should be carried out in all pre-term infants throughout the first year of life to detect associated disorders. Periodic monitoring of visual acuity is also carried out since severe ROP may be associated with impaired visual development.

ROP- It's a shared responsibility

- All neonatal care units must have criteria and procedures to ensure appropriate ROP screening
- Screening results should be well documented and communicated to parents
- Counsel parents regarding the chance of poor visual outcome even with therapy
- Arrangements must be made by the neonatal team for appropriate ophthalmological follow-up even after discharge/ transfer to another unit
- Follow-up programs should include visual examination for all preterm infants who have been screened for ROP
- Ensure periodic follow up for preterm infants even without ROP as they are more likely to experience visual problems than term infants

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Congenital Nasolacrimal Duct Obstruction (CNLDO)

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Congenital nasolacrimal duct obstruction is a common cause of watering in newborn children. Studies show that the prevalence is about 20% 1. In almost 90%, symptoms appear during first month of life 2 .Clinical presentation is characterized by excessive watering from eyes, constant or intermittent. Other manifestations are matting of lashes, muco purulent discharge and regurgitation of pus on applying pressure over lacrimal sac area (ROPLAS+ve). In 20% of cases the condition is bilateral3.

Pathogenesis

Delayed canalization of the nasolacrimal duct at the distal end, at the level of valve of Hasner, (fig:1) is the cause CNLDO. Other causes described are bony abnormalities or stenosis of inferior meatus where nasolacrimal duct opens into the nose4.

Medical Management

Spontaneous resolution of the symptoms happens in 95% of cases by 12 months of age. Digital massage over the lacrimal sac (Crigler maneuver) is recommended (fig 2) to increase the hydrostatic pressure which may rupture the membranous obstruction. Studies show significant improvement in watering after massage when compared to infants without massage5.Topical antibiotics are used in case of bacterial super infection. Complications like preseptal cellulitis and orbital cellulitis warrants hospital admission and intravenous antibiotics. It is important to rule out other causes of watering like congenital glaucoma and corneal anomalies.

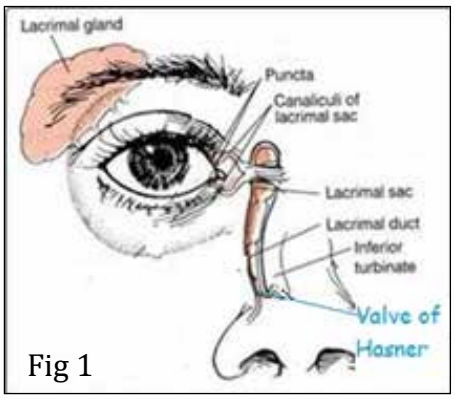

Fig 1

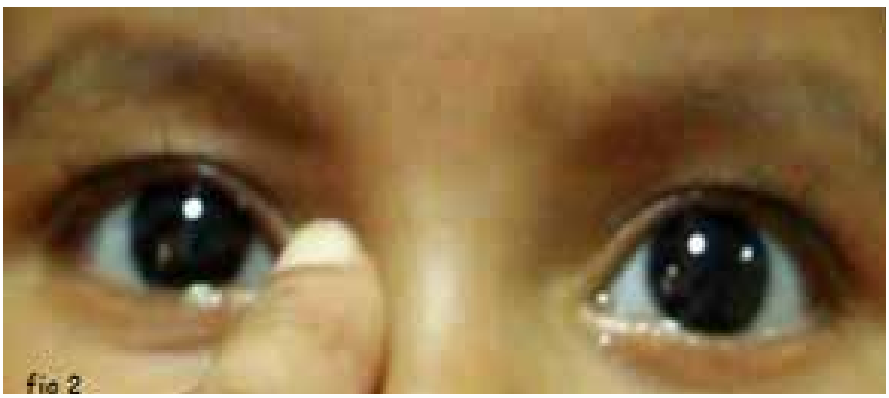
fig 2

Probing

Passing a metal probe through the lacrimal punctum down to the valve of Hasner to mechanically overcome the obstruction is a procedure with high (90%)success rate. Timing of probing is controversial. Generally it is done around one year of age under general anesthesia. Success rate increases (94% to97%) when it is combined with nasal endoscopy guidance, especially in complex NLDO, failed probing, and associated nasal pathology6. Early probing is recommended in conditions like acute dacryocystitis, profuse discharge from eye, large dacryocystocele and in need of intra ocular surgery

Nasolacrimal intubation, balloon catheter dilatation and dacryocystorhinostomy (DCR) are reserved for refractory conditions.

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Childhood Cataracts

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Epidemiology and Relevance

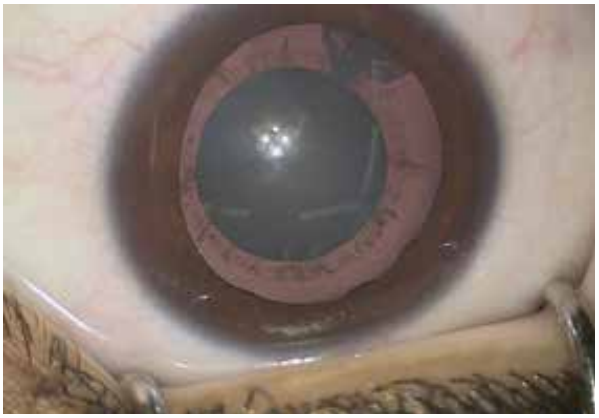
Control of childhood blindness is given priority in WHO's initiative Vision 2020-Right to sight programme , where, India has a major role through Vision 2020 India-Right to Sight initiative. Childhood cataracts account for 5- 20% of childhood blindness globally (1). In India cataract accounted for 28% of childhood blindness and is the commonest cause of treatable blindness. The overall prevalence of congenital cataract was 2.5 to 3.5 in 10000 in the west. According to WHO, it is 8 in 10000 in India.

A clear visual axis is essential for development and maturation of the visual system. If focused image is not formed by 6 to 8 weeks, permanent visual impairment due to amblyopia and nystagmus can set in. So infantile cataract which can affect vision significantly is to be evaluated immediately. There are very few conditions like galactosemia, which if detected early enough and treated conservatively can be reversible. Cataract may occur unilaterally or bilaterally of various morphology and grade of opacification. Not all cataracts need surgery. Dense cataract detected at the time of birth is an emergency. Appropriate surgical correction at the right time can help the child lead a normal life. Management involves long term follow up to look for the development of glaucoma,visual axis opacification and refractive correction and is a teamwork, involving ophthalmologists, anaestheists, paediatricians, optometrists, orthoptists and low vision rehabilitation specialists.

Congenital Cataracts

Cataract presenting at birth or soon after, often have some specific causes. The commonest cause is an infection in mother during the first trimester. Genetic or chromosomal anomalies may also cause a congenital cataract. They are detected by screening the newborn baby by the Bruckner reflex, ie by shining an ophthalmoscope light into the baby's eyes while observing through it from 1 foot distance- the red glow can be appreciated if there is no media opacity. The optical media of the eye are the cornea, the lens and the vitreous cavity. If the red glow is not seen, it is better to have a dialated examination. For the neonate, we use half strength of the adult topicamide -phenylephrine drops(0.5%+2.5%).This should be done as soon as the baby is born, before discharge from the hospital.

The **morphology of cataract** is important. It may be central foetal nuclear with calcific capsule as in Rubella infection. It may be anterior or posterior polar, all of which point to a congenital origin. The anterior polar cataract does not usually produce significant visual disability . A more posteriorly located cataract affects vision more than an anteriorly located one. Lamellar cortical cataract points to an onset much later. If cataract involves only some peripheral part of the lens, vision may not be affected early in life and though detected much later it may still have a genetic origin. Maternal malnutrition and drugs are implicated in lamellar cataracts.



Lamellar cataract

Acquired cataracts

Though occurring later in childhood, they may well be genetic in origin. Other causes are metabolic and syndromic. Trauma is a major cause of acquired cataract often due to penetrating injury and is seen more among boys and a unilateral acquired cataract should be fully evaluated for any intraocular foreign body. Steroid induced cataracts and atopic cataracts are seen later in childhood.

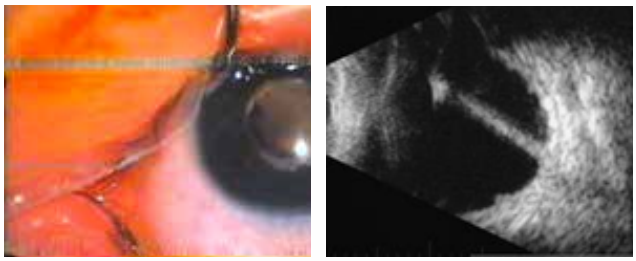
Etiology of congenital cataracts

Intrauterine infections -Rubella, CMV, Toxoplasma, Herpes simplex, Varicella zoster(TORCH) is the leading cause of congenital cataracts, accounting for 33% of congenital cataracts in India(6). Rubella vaccine though become universal in the schedule, is not being utilized to the full extent.

Bilateral Rubella Cataract

Apart from Rubella, Cytomegalovirus infection is associated in 16% of these cases. Hereditary cataracts account for about 25-30% of the nonsyndromic childhood cataracts and may be autosomal dominant, recessive or X linked, the autosomal dominant inheritance being the commonest. Genetic mutations that code for lens proteins can also occur. Parents have to be screened under slit lamp examination, since they may be unaware of it(4). About 25 % are idiopathic (5).

20% are unilateral. Microphthalmos is seen in as high as 68% of patients with congenital Rubella Syndrome(4). Unilateral cataracts may be associated with other intraocular problems .Persistent foetal vasculature(PFV) is a common cause for uniocular cataracts. Persistent foetal vasculature produces opacification by invation of lens by vessels of varying extent. The eye is microphthalmic with stretched zonules seen under dialatation. These cataracts may show a retrolenticular pink hue indicating the vascular nature.They show a higher incidence of secondary glaucoma and posteriorly located ones can develop traction retinal detachment.



PFV showing pinkish hue and B scan showing the dense shadow of retrolenticular stalk going toward the disc.

Other associated ocular abnormalities include lenticonus, anterior segment dysgenisis, Stickler syndrome etc

Posterior polar cataract may present in an older child may have a pre existing capsular dehiscence of 25% and surgery is challenging.

Infants with classical galactosemia develop oil droplet cataracts , and if left untreated, they

progress to total cataracts due to accumulation of galactitol in the lens. If detected early and galactose eliminated from diet, the cataracts may be reversible. Hence it is important to test the presence of reducing substances in the urine after a galactose containing meal [milk] in all infants. They may have symptoms vomiting, diarrhoea and failure to thrive. Enzyme assays and D N A studies can be used to confirm the diagnosis. In hypocalcemia cataract starts as punctate lens opacities and may progress to total cataract later.

The common syndromic associations are Downs, Lowe's syndrome, Nance Horan syndrome , Hallerman Streiff syndrome, Martsolfs syndrome, Marinesoco- Sjogren syndrome, incontinentia pigmenti etc. Other causes of secondary cataract are uveitis, which usually occurs later. Associated mental retardation is common. There are about 39 genetic loci to which isolated or primary cataracts have been mapped, the number is constantly increasing (8).

Investigations and Systemic evaluation

If there is a family history of cataract, an elaborate blood work up is not necessary to find the causative factor. All parents should be screened for cataract. In bilateral total cataract occurring at birth, routine blood tests with a TORCH titre, urine reducing substances, calcium ,phosphorus should be done. If associated systemic symptoms are there, red cell galactokinase and urine aminoacids(Lowe's) should be done. In unilateral cataract TORCH titre again must be done, since Rublella cataract may be unilateral too. An evaluation by the paediatrician to look for any other associations is mandatory. In Rubella cataract a paediatric cardiologist should evaluate the child and assess feasibility of general anaesthesia. Often a PDA closure is done before cataract surgery. These babies may need a day of ICU care postoperatively. Challenges of early anaesthesia -hypothermia/hyperthermia must be kept in mind.

Management

Ophthalmic evaluation

Visual assessment should be performed using fixation patterns and when possible by preferential looking charts, or pattern visual evoked potentials. Measurement of corneal diameter, I O P , pupillary reflexes, and indirect ophthalmoscopy and slit lamp examination, a hand held keratometry and axial length measurement should be done .In Rubella unilateral cataracts the other eye may show pigmentary retinopathy on fundus evaluation. . A B scan is a must in all dense cataracts where posterior segment cannot be visualised to rule out PFV which may show variable remnants of primary foetal vasculature. Sometimes a doppler B scan shows the vascularity of the same and the extent of the stalk and the approach(Limbal Vs Pars Plana) to surgery may differ in a posterior PFV. A B scan will rule out retinoblastoma as well. Sometimes an examination under anaesthesia may be necessary.

Timing of surgery

All cataracts do not need surgical intervention. **A total or dense cataract** precluding visualization of fundus needs immediate intervention. An infant presenting with dense cataracts needs emergency surgical intervention since delay can cause nystagmus and amblyopia. The visual system which is immature at birth has a latent period of approximately 6 weeks before it becomes sensitive to visual deprivation, and binocular vision first appears at approximately 3 months of age. The child loses one line of visual acuity for every three weeks that the surgery is delayed in the first 14 weeks of life. Bilateral dense cataract at birth has to be operated by 8 weeks . Rarely simultaneous surgery may be done if there is anaesthetic risk. Otherwise the second eye is operated in 7-10 days.

Unilateral dense cataract has more risk of developing amblyopia since a good (rival)eye is already there. There was a time when restoring vision ina unilateral congenital cataract was considered impossible. Unilateral dense cataract should be operated by 6weeks. Cataract surgery before 4 weeks of age increases the risk of secondary glaucoma. The pathophysiology of aphakic or pseudophakic glaucoma is poorly understood. Its aetiology has been attributed to the damage to the immature trabecular meshwork by inflammation, the loss of mechanical support of the trabecular meshwork, or a toxic substance gaining access to the trabecular meshwork from vitreous and may present as an early angle closure glaucoma or late open angle glaucoma.

Unilateral cataracts need special attention to start aggressive amblyopia management earlier with optical correction after surgery.

Dense cataract diagnosed later in childhood should be operated immediately.

In partial cataracts, evaluation is done to assess whether it is visually significant.A cataract located posteriorly, of more than 3mm size, precluding visualisation of macula and producing strabismus are all visually significant. In older children after the amblyogenic period, ie after 6 to 8 years, with manageable visual acuity, they may be followed up conservatively before contemplating surgery. During this time a dialating drop like homatropine with optical correction may be useful. We must understand that we are eliminating the accommodative function of the lens by doing a cataract surgery .So until regular visual functions are affected, we may wait.

Choice of surgery

Extracapsular cataract extraction with Primary Posterior Capsulorrhexis(PPC) and anterior vitrectomy with or without intraocular lens (IOL) is the surgical procedure in children. Paediatric eyes are not miniature adult eyes. Low scleral rigidity, elastic capsule of lens and collapsibilty of globe pose challenges during surgery. Since posterior capsule can opacify in 100% in children a PPC is mandatory upto 6 years or more if necessary. of If bilateral cataract surgery is done before 6 months, we may leave him aphakic and spectacles given for both eyes. The infant eye grows fast during the first 18months.The corneal curvature, lens power and axial length of the eyeball interplay beautifully during development to effectively produce what we call emmetropia or normal vision. So calculation of IOL power is very unpredictable during this time. He may progress with a myopic shift to land with a large myopia by adulthood. So in bilateral cataract it will be safe to implant an IOL after 2-3 years. There are various IOL formulae depending upon the age of the child to undercorrect the power , so that the residual hypermetria is corrected with glasses and slowly followed up to reduce the power.The predictability of the IOL power calculations have been extensively studied(9).

In unilateral cases, things are different. Unless you give reasonably good vision to that eye he will not use the thick aphakic glasses and the whole attempt of surgery may become futile .You have to do aggressive patching of good eye to make him use the operated eye.So in unilateral cases, if the corneal diameter and axial length are reasonably normal IOL implantation may have an advantage. The Infant Aphakia Treatment Study(IATS)looked into the feasibility of implanting IOL before 6months in unilateral cataracts. (10)A comparative study between primary IOL implanated infants and aphakia with contact lens managed children showed that the 5 year visual outcome was more or less the same. But multiple surgeries due to visual axis opacification justified avoiding IOL implantation before 6 months of age. Postoperative uveitis is a complication in children which produces membrane formation formation and visual axis opacification.

In children not implanted with IOL spectacle correction-bifocals after one year and contact lenses in unilateral, with patching if necessary(if there is a preference of fixation) should be instituted.

Follow up and Counseling

Cataract surgery is not the end of treatment. The treatment which follows surgery, ie optical correction with spectacles or contact lenses, management of visual axis opacification, management of amblyopia by patching and glaucoma evaluation and management are equally or more important. Counseling should start before surgery. The importance of follow up, patching if unilateral, optical correction all the time and looking for preferential fixation or strabismus should all be emphasized. Follow up includes Vision assessment, fundus examination, refraction, IOP measurement and axial length measurement. If there is disproportionate myopic shift during refraction which may indicate a glaucomatous change in the young eye where axial length increases , EUA must be done to look for IOP and evaluate for glaucoma. If there is visual axis opacification he may need a membranectomy. Though glaucoma medications are given to start with, often they need surgey-conventional trabeculectmy may not be enough and may need a glaucoma drainage implant. The long term development of glaucoma after surgery is found to beas high as 20 to 30% (11). Microphthalmic eyes, early surgery, PFV etc predispose for development of early glaucoma.

Conclusion

Dense cataracts at birth are an emergency-optimal time in unilateral cataract being 4-6 weeks and in bilateral, 8weeks.Surgery should be immediately be followed by optical correction and patching started immediately in unilateral cataracts. Partial cataracts may be followed up till visually significant. Follow up to look for refractive changes, IOP, visual axis opacification, amblyopia and strabismus are important and may need interventions.

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Typical Ophthalmic Features
Road to Diagnose Inherited Metabolic diseases

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Introduction

Inborn metabolic disorders (IMDS) are rare group of disorders of metabolic pathway with heterogeneous presentations which affect the various systems of the body. IMDS have an underlying genetic defect associated. Of all the systems involved the Ocular involvement plays a very crucial role as there are very peculiar ocular features of a particular IMD therefore it is prudent for an ophthalmologist to be familiar with the ocular features of IMDS .On the basis of these features a suspicion of a particular IMD can be made and a battery of upper level laboratory tests can be performed and a rapid definitive diagnosis can be made and the patient can be sent for further management. Early diagnosis is mandatory because of the following 3 reasons .First, IMDs are rapidly progressive and cause irreversible damage early in the course of the disease. Second, the treatment can often be effective, if commenced early and long-term outcome may be improved. Lastly, correct early diagnosis helps in genetic counselling. The Diagnosis and management is carried out with coordination of multiple group of specialist including an ophthalmologist, paediatrician, biochemist, and medical geneticist.(1)

Mechanism

Metabolic disorders generally are inherited in an autosomal recessive fashion. The IMDs most postulated mechanism is based on the Garrod's Hypothesis which says that the majority of IMDS are caused by absence or deficiency of a specific enzyme that catalyses in the biochemical pathway, with subsequent accumulation of the reaction's substrate which can be toxic directly or indirectly(conversion of the substrate via an alternative pathway to a toxic substance).(2)(3) Environmental factors may also trigger the onset and severity of disease. It also depends on degree of accumulation of toxic substances before metabolic block, for example, diet, intercurrent infection, fasting drugs, and so on.(3)(4)

The eye and IMD

The IMDS can manifests in any part of the ocular system either as a complication or a sequel or as the initial presentation where the vision may or may not be affected. The varied presentations can be cerebral visual cortex dysfunction in many IMD with progressive central nervous system degeneration, to optic neuropathy, retinopathy, lens opacification and cataracts, corneal clouding and various other keratopathies, and abnormalities of eye movements if the extraocular muscles are affected or if the central optic motor nuclei are dysfunctional. (3)

Early diagnosis and treatment may improve visual outcomes.

The following table will focus on the key IMD conditions that have distinct presentations in the eye.

Conjunctival/scleral

Acrodermatitis enteropathica -conjunctivitis

Alkaptonuria- scleral pigmentation

Cystinosis- conjunctival crystals

Fabry's syndrome-dilated conjunctival vessels

Gauchers (non cerebral juvenile) –conjunctival pigmentation

Table 1. Ocular abnormalities and metabolic disease (5)

Hypophosphatasia-calcium deposits in the conjunctiva

Tyrosinaemia type II-white conjunctiva

Corneal

Cystinosis -corneal crystals(Figure 1)

Fabry's syndrome -whorl like corneal opacities

GM1 gangliosidosis -corneal clouding

Lecthin cholesterol acyl transferase deficiency -corneal opacities

Mucopolysaccharidoses (I IV VI VII, II rarely) -corneal clouding

Mucolipidoses I II I IV -corneal clouding

Tyrosinaemia type II -dentritic ulcers, scarring

Wilson's disease -Kayser-Fleischer ring(Figure2)

Fuscosidosis-Corneal clouding

Iris

Albinism -thin and pale

Maternal phenylketonuria-coloboma

Lens:

Cataracts

Diabetes mellitus

Fabry's syndrome

Galactosaemia

Gyrate atrophy

Hypoparathyroidism

Mannosidosis

Marinesco-Sjogren syndrome

Peroxisomal disorders

Wilson's disease Lowe's syndrome

Dislocation

Homocystinuria

Hyperlysinaemia

Sulphite oxidase deficiency

Ehlers Danlos syndrome

Retinal disorders

(1) Retina

Abetalipoproteinaemia -- pigmentary degeneration

Albinism -- diminished pigmentation

Batten's disease (juvenile, late juvenile) -- pigmentary degeneration

Cystinosis -patchy depigmentation

Diabetes mellitus -- haemorrhages, exudates, neovascularization

Farber's disease - diffuse pigmentary mottling

Gyrate atrophy -patchy atrophy which fuses pigmentary degeneration

Kearns-Sayre syndrome - pigmentary degeneration

Peroxisomal disorders - pigmentary degeneration

Hyperoxaluria type 1-- white flecks

(2) Macula

Batten's disease (juvenile late infantile), - macular and retinal degeneration

Farber's disease

Gangliosidoses GM1 Type I, II

Gangliosidoses GM2 Type I, II

Metachromatic leucodystrophy

Niemann Pick type A, C

Sialidosis type I, II

Optic nerve

Adrenoleucodystrophy

Batten's (late infantile)

Leigh's disease

Mannosidosis

Menke's syndrome

Metachromatic leucodystrophy

Porphyria (acute intermittent) - optic neuritis

Disorders of ocular movement and position

Albinism -nystagmus

Abetalipoproteinaemia – Nystagmus, ophthalmoplegia

Gaucher's disease -paralytic strabismus

Hartnup disorder -nystagmus, strabismus

Kearns-Sayre syndrome - ophthalmoplegia

Leigh's disease -- ophthalmoplegia

Niemann Pick D - supranuclear ophthalmoplegia

Porphyria -partial 3rd nerve paralysis

Hypothyroidism – Nystagmus

Figurre-2 Cystinosis; crystalline deposits in the cornea

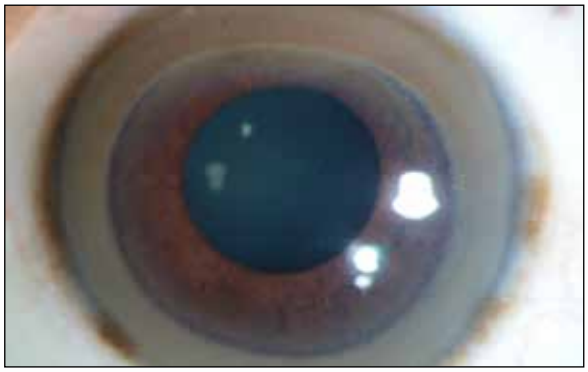

Figure 1 Kayser Fleischer ring in Wilson disease; deposits of brown-green copper containing substance in the peripheral cornea

Newborn Screening

Newborn screening is important for the early detection of inherited genetic and metabolic disorders, allowing doctors to preemptively treat or manage affected babies to reduce illness, disability, or death. The screening is performed soon after birth and involves a simple blood test alongside a non-invasive hearing test. The process of newborn screening is relatively quick and easy. Between 24 hours to seven days after birth, a few drops of blood are taken from an infant's heel and placed on a special card. The paper is sent to a specialized laboratory for testing. The results of the blood tests are sent to the infant's pediatrician within two to seven days. If any of the tests come back positive, further testing will be done to confirm the diagnosis. Parents do not have to request the tests; they should be automatically performed. Simple screening tests may aid in diagnosis and evaluation of a suspected case of IMD and provide direction for more comprehensive laboratory analysis. In most cases, diagnosis can be established without any biopsy through biochemical analysis of blood and/or urine for specific metabolites. On special occasions, cerebrospinal and amniotic fluids (for estimation of amino acids, carnitine, organic acids, mucopolysaccharides, enzyme(s), and so on) and chorionic villi are also used for diagnosis based on biochemical investigations Symptomatic management is the mainstay for many of the disorders.

Diagnosis:

Inherited metabolic disease have several forms that vary in age of onset, clinical severity, and often, mode of inheritance. IMDS can be divided in to various types based on level of pathway of synthesis or catabolism of metabolic products (proteins, carbohydrates, or fats) affected as the Small molecule disorders and large organelle disorders.(3)

Identification of an IMD requires a low threshold of clinical suspicion. Within the clinical history, evidence of extra-ocular features should be sought to determine if the presenting eye signs or symptoms are in the context of a multisystem disease. The age of the child is important; a neonate with failure to thrive, recurrent vomiting, prolonged jaundice or other developmental concerns may have an IMD. The family history is important, especially other affected siblings or relatives, unexplained neonatal or childhood deaths, recurrent miscarriages. Enquiring about parental consanguinity is important as many of the IMD are inherited in an autosomal recessive manner, although these conditions still occur in children of non-consanguineous parents. Subsequent dietary preferences, abnormalities of normal childhood developmental progress (especially developmental regression, i.e. loss of developmental milestones previously achieved), detection of hepato/splenomegaly or dysmorphic features can also allude to an IMD. (3)

Management:

Treatment approaches to the different IMD are varied depending on the aetiology of the condition. Symptomatic management is the mainstay for many of the disorders. Some IMD have specific dietetic interventions (e.g. galactose restricted diet in the galactosaemias), while others respond to high dose vitamin or cofactor supplementation.(2)

Recently enzyme replacement therapy, enzyme enhancement therapy, and substrate deprivation therapy have been tried. The impact of these therapies on the ophthalmic manifestations of the disease is very variable, in some situations (e.g. vitamin E replacement in primary vitamin E deficiency or abetalipoproteinaemia, or biotin replacement in biotinidase deficiency) this may prevent or reverse visual impairment, but in others the eye is relatively refractory to therapy.

Specific ophthalmological interventions may be required. For those with refractive error or visual impairment, appropriate provision of glasses or other visual aids will be required. Standard interventions for correction of strabismus may be undertaken, although understanding the natural history of each disorder is important in determining whether a therapy is appropriate. Management of cataract may require lensectomy, although in some situations instigation of appropriate dietetic management may facilitate resolution of the cataract if not established. For some conditions other surgical interventions including corneal grafting to correct corneal clouding has been beneficial, but appropriate investigation to determine retinal function should be undertaken in the surgical planning process as there may be concurrent optic atrophy or visual loss if there has been chronic uncorrected raised intraocular pressure. Certain disorders may lead to specific topical treatments such as cysteamine in cystinosis.

Conclusion

IMDS are challenging because of their rarity. Patients with inborn errors of metabolism may have characteristic ocular changes which are in some cases diagnostic, but for others detailed investigation may be necessary to establish the diagnosis. This is essential for the appropriate treatment to prevent progressive ocular disease and to enable the family to be counselled accurately about the genetic risk and the consideration of prenatal diagnosis. Therefore a combined approach by the various specialists is required in majority of the cases. Traditional therapies for IMDs include dietary therapy, such as protein restriction, cofactor supplements, and so on. Recent advances in diagnosis and treatment have significantly improved the prognosis for many infants with inborn errors of metabolism.

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The eye as a reflection of childhood illness (other than IEM)

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The eyes, including the ocular adnexae and visual pathways, are often involved in diseases of other systems, either directly or indirectly.

The eye is a complex organ. 40% of all afferent fibres to the CNS are related to vision. Eyes and the pathways taken by the visual pathways provide ample opportunity for its affection.

General conditions other than IEM with ophthalmological associations

Raised intracranial pressure and papilloedema

The ophthalmoscopic signs of papilloedema are blurring of the edge of the optic disc, hyperemia, venous dilatation in the presence of splinter haemorrhages, and the absence of disappearance of venous pulsation. When the oedema is marked, the papillary surface may be raised above that of the retina; a phenomenon which is expressed in diopters, measured as the difference between the point of focus on the papilla and that on the retina; 3 diopters are equivalent to 1 mm.

The mechanism of development of papilloedema is primarily due to a rise of CSF pressure in the optic nerve sheath, which produces axoplasmic flow stasis in the optic nerve fibers in the surface nerve fiber layer and prelaminar region of the optic nerve head. Axoplasmic flow stasis then results in swelling of the nerve fibers, and consequently of the optic disc. Swelling of the nerve fibers and of the optic disc secondarily compresses the fine, low-pressure venules in that region, resulting in venous stasis and fluid leakage; that leads to the accumulation of extracellular fluid. Contrary to the previous theories, the various vascular changes seen in optic disc edema are secondary and not primary. Thus, optic disc edema in raised CSF pressure is due to a combination of swollen nerve fibers and the accumulation of extracellular fluid. Papilloedema does not occur immediately following a rise in intracranial pressure. There is usually an interval of 24 hours before signs appear on examination of the fundus.

Raised intracranial pressure makes one think of supratentorial space-occupying lesions, viral and bacterial infections such as meningitis and endocarditis, and neoplasms.

The difference between papilloedema and papillitis depends on the degree of functional disturbance: practically none in papilloedema, very marked in papillitis.

Sometimes the appearances of pseudo-papilloedema are difficult to distinguish from true oedema, especially in young patients in whom there may be drusen of the papilla, hypermetropia or embryonic rests. Signs such as the presence or absence of spontaneous venous pulsation, transillumination of the drusen, or the presence or absence of hemorrhages may be helpful

Diabetic retinopathy (DR)

Diabetes in the young child and adolescent is characterized by two ocular complications: cataract and diabetic retinopathy, which may become proliferative.

In insulin-dependent diabetes the prevalence of diabetic retinopathy is less than 10%, where the disease has been present for less than 5 years. Prevalence of DR has been shown to be rare in young people <10 years but increases with increasing age. DR in those aged 10-14 years is 2% and in those aged 15-19 years is 10%. The incidence of DR is higher among patients with T2D and occurred with a shorter duration of disease, as compared with T1D.

The leakage of contrast medium in fluoresceine angiography is the first sign of diabetic involvement of the retina. Angiographic diagnosis is possible in 50 % of patients when ophthalmologic diagnosis was only made in 16 % from the presence of microaneurysms. These latter are therefore not the first lesion to appear or to be detectable.

If not properly treated, diabetes can also be complicated in the child by vascular proliferation and vitreous hemorrhages, sometimes requiring active treatment, such as laser therapy and vitrectomy.

The prevalence of early diabetic cataract in the population varies between 0.7 and 3.4% of children and adolescents with T1DM. The occurrence of diabetic cataract in most pediatric patients is the first sign of T1DM or occurs within 6 months of diagnosis of T1DM. The pathophysiological mechanism of early diabetic cataract has not been fully understood; however, there are many theories about the possible etiology including osmotic damage, polyol pathway, and oxidative stress.

Vitamin deficiency

The different eye signs of vitamin A deficiency (VAD) in children, as graded by the WHO, are: night blindness (XN), conjunctival xerosis (X1A), Bitot's spots (X1B), corneal xerosis (X2), corneal ulcer covering less than 1/3 of the cornea (X3A), corneal ulcer covering at least 1/3 of the cornea, defined as keratomalacia (X3B) and corneal scarring (XS).

Other vitamin deficiencies: B2 – optic neuropathy and angular blepharo-conjunctivitis; B12 - 'flame shaped' retinal hemorrhages; K – ocular hemorrhage.

Hematological conditions

The ocular effects of the leukemias, above all the acute forms, are extremely important. On examination of the fundus one can find hemorrhages, most often retinal, dry and cotton wool exudates, venous dilatation and sometimes perivenous sheaths. Leukemic cells very commonly invade the choroid, and although this is well recognized by pathologists, it is often missed on clinical examination. Ultrasound examination allows a better demonstration of the infiltration, due to the different ultrasonic characteristics of the infiltrate and the subjacent and overlying tissue, even if the neoplastic layer is rather thin. The eyelids and the orbit are other sites of infiltration. Eleven percent of children presenting with unilateral exophthalmia have one or other form of leukemia.

The optic nerve may be invaded in its pre1aminar part, visible by ophthalmoscopy, or in its retrolaminar part, giving an appearance of optic neuritis. The differential diagnosis is between papilloedema and an infiltration of the nerve, because of the risk of blindness. Finally, cases have been reported, in which there is involvement of the iris, with the symptomatology of iritis, sometimes with hypopyon. These ocular involvements, which are sometimes isolated, are interpreted as being due to the relatively poor penetration into the eye of systemically administered drugs.

The anemias, in general, only show retinal hemorrhages and venous dilatations, and only if the level of hemoglobin is very low. The haemoglobinopathies must be considered separately. Thus, for example, the hemoglobin S induces the formation of sickle red cells with slowing or arrest of the circulation. In the retina this leads to vascular blockage, to zones of capillary non-perfusion and to the development of neovascular tufts inside the vitreous. This process is similar to what occurs in proliferative diabetic retinopathy in which retinal ischemia causes a similar sequence of events.

Diseases of connective tissue

Juvenile rheumatoid arthritis: Ocular involvement is essentially an anterior uveitis which may be asymptomatic. Curiously, the eye condition is often more serious in forms involving fewer joints than in those involving many at the same time. A band-shaped uveitis can develop secondarily to the chronic uveitis: deposits of calcium salts occur under the corneal epithelium.

Kawasaki disease – most common is bilateral conjunctival injection. Iridocyclitis, superficial punctate keratitis, vitreous opacities, papilledema, and subconjunctival hemorrhage also may occur.

SLE – 1/3rd have ocular manifestations. The most common manifestation is keratoconjunctivitis sicca. The most vision threatening are retinal vasculitis and optic neuritis/neuropathy. Optic neuropathy has strong prediction for CNS lupus.

In ankylosing spondylitis, a condition which mainly affects older, male adolescents, one also finds an anterior uveitis. From the ophthalmological point of view the lesions are similar in both conditions.

Behcet syndrome - panuveitis was the most common, cataract and optic atrophy

Sjogren syndrome - Severe dry eyes can cause corneal ulceration, scarring, infection, and even perforation. Risk of vision-threatening corneal complications se higher in presence of scleritis.

The phakomatoses

These are congenital dysplasias affecting the skin, the mucous membranes, peripheral nerves, the central nervous system, and eventually the viscera by means of their innervation. The ones to be considered are von Recklinghausen's disease of neurofibromatosis, Sturge-Weber-Krabbe Syndrome, von Hippel-Lindau disease, and tuberous sclerosis.

A characteristic feature of neurofibromatosis is the neurofibroma of the upper eyelid with S-shaped ptosis. If the tumour is situated more deeply in the orbit, it can produce an exophthalmos. In certain cases, one can demonstrate a pulsatile exophthalmos due to the absence of a part of the sphenoid allowing transmission of the pulse, by herniation of cerebral tissue into the orbit. Glioma of the optic nerve is also a common lesion. Nodules have been described in the iris. The development of a buphthalmos by congenital glaucoma is not uncommon. Blindness can occur in neurofibromatosis, as well as malignant transformation in tumour of the skin and nervous system.

Retinal angiomatosis, or von Hippel-Lindau's disease, occurs in an older age group and will not be discussed here. Sturge-Weber-Krabbe syndrome, is characterized by facial angioma in the territory of the 5th cranial nerve, a choroidal angioma and meningeal angioma. All three occur on the same side. The neurological signs develop in the first years of life, with epileptic fits and signs of deficit in the side opposite to that of the cutaneous angioma, for example a contralateral hemianopsy. Glaucoma is common, either congenital with buphthalmos, or developing later with an anomaly of cleavage of the anterior chamber angle. Treatment is difficult, and the prognosis for sight is poor.

In tuberous sclerosis, papillary hamartomata and retinal lesions are found which in fact are hamartomata of the neurofibrillary layer.

Renal patholog y

In general, the importance of retinopathy in renal insufficiency is a function of the associated arterial hypertension. A few special syndromes deserve attention:

in Lowe's syndrome renal tubulopathy - Dense congenital cataracts are found in all affected boys and infantile glaucoma in approximately 50%. All boys have impaired vision; corrected acuity is rarely better than 20/100.

in Alport's syndrome glomerulo-nephritis with haematuria is associated with deafness and some- times cataract;

in Wilms' tumour aniridia is a common finding, which justifies a search for the tumour when the ophthalmologist finds the ocular anomaly.

Muco-cutaneous conditions

Erythema multiforme occurs after sensitization to certain drugs. Amongst those incriminated are sulphonamides, salicylates, etc. It also occurs after certain infections. It presents with a conjunctivitis which is sometimes pseudomembranous. This may proceed to symbleraphon and dry kerato-conjunctivitis.

Epidermal necrolysis is related and affects young children, often being associated with a staphylococcal infection. Conjunctival involvement is less severe, and scarring is less common.

In incontinentia pigmenti a retinal dysplasia is described with the development of a retrocrystalline mass. Pigmentary retinopathy is also a common finding.

In Ota's naevus the skin in the territory of the two superior branches of the trigeminal nerve takes on a greyish colour, as does the sclera on the same side. Retinitis pigmentosa, cataract and glaucoma are described as being associated with Duane's syndrome.

Ocular involvement in ectodermal dysplasia occurs in the eyelids: agenesis of the glands, eye-lashes, eyebrows and lacrymal papillae. As well as the systemic and cutaneous disorders of Rothmund-Thomson syndrome, there is absence of eyelashes and eyebrows, and cataract which is early.

Stevens-Johnson syndrome: Mild ocular involvement - eyelid skin desquamation and denudation, eyelid edema, mild corneal involvement, mild conjunctival injection, mucous discharge, or chemosis. Moderate - membranous conjunctivitis, epithelial defects with more than 30% healing with medical treatment, corneal ulceration, or corneal infiltrates. Severe - acquired eyelid malposition, formation of symblepharon, nonhealing corneal epithelial defects, complete or partial visual loss, or foreshortening of conjunctival fornix.

Musculo-skeletal conditions

The muscular dystrophies do not usually present any particular ocular lesion. Myotonic dystrophy is an exception, since ptosis and cataract, taking the form of multicoloured small opacities, are quite often described. Myasthenia is usually an adult condition, but there is also a rather uncommon juvenile form, in which external ophthalmoplegia and ptosis may be encountered. Ehlers-Danlos disease can be associated with an epicanthus, a keratoma, subluxation of the lens and retinal detachment.

Amongst the anomalies of the bony development of the face, Goldenhar's syndrome (oculo-auriculo-vertebral syndrome) is an example in which there are vertebral anomalies, dermoid and epibulbar tumours, and anomalies of facial development. The craniostenoses presents ocular problems secondary to the bony anomalies: exophthalmos, hypertelorism, compression of the optic nerve, etc.

Amongst the generalized anomalies of the skeleton: Marfan's is characterized by a luxation or subluxation of the lens, usually upwards. This can be complicated by acute glaucoma from closure of the angle if the crystalline luxates into the anterior chamber. In Weill-Marchesani syndrome a similar luxation or subluxation of the lens is associated with general morphological characteristics which are quite different from those of Marfan's disease: the patient is of short stature, with short, stocky limbs with well-formed muscles, short fingers and restricted hand movements. They may also have severe myopia.

38% of 5-year-old patients with untreated homocystinuria have lens subluxation, and almost all patients have it by age 25. The zonules are absent, as compared to the stretched zonules in Marfan syndrome. Anterior lens subluxation leads to elevation of intraocular pressure and to corneal decompensation from lens-endothelial contact.

Osteogenesis imperfecta - blue sclera, hyperopic or myopic eyes, retinal detachments, decreased corneal rigidity, and glaucoma.

Infective disease

Congenital rubella syndrome: congenital cataract, microphthalmia and a retinitis known as "pepper

and salt". Intra-uterine infection with cytomegalic inclusion virus can result in a localized or diffuse retinitis. Congenital toxoplasmosis, as well as its general effects, can be responsible for a macular chorioretinitis. Central vision is often lost. In addition, there can be a recurrence at the edge of the initial lesion.

Diphtheria can result in oculomotor paralyses and paralysis of accomodation due to the secreted neurotoxin. Ocular involvement presents as a membranous conjunctivitis with copious secretion.

Tuberculosis - Tuberculosis can give rise to a uveitis which can be anterior and posterior at the same time- most common ocular presentation of tuberculosis in child is posterior uveitis. Phlyctenular kerato- conjunctivitis is thought to be an allergic reaction to tuberculin, but this is not the only allergen which can trigger such a reaction.. Serpigenous-like retino-choroiditis, choroid tubercles, eyelid cold abscess, dacryoadenitis, etc. may be seen

Measles – conjunctivitis, keratitis, corneal ulcers and scars, and rarely retinitis, optic neuritis and blindness

Herpetic kerato-uveitis – dendritic or punctate epithelial keratitis, and stromal keratitis occur concurrently with epithelial keratitis. Usually have bilateral ocular involvement and are at risk for recurrent keratitis and amblyopia.

Leprosy - affects the eye in four ways: 1) by direct invasion of lepra bacilli into eye structures (keratitis, iridocyclitis, scleritis, episcleritis); 2) secondary to involvement of facial nerve (lagophthalmos) and oph-thalmic division of trigeminal nerve (corneal anaesthesia); 3) hypersensitivity reaction to the antigenic substances released in the breakdown of lepra bacilli (iridocyclitis, scleritis, episcleritis); 4) secondary to changes in the skin and support tissue of the lids, tear drainage system (madarosis, trichisis, entropion, chronic dacryocystitis).

Pertussis may present as subconjunctival hemorrhage

Fungal infections: due to penetrating trauma by objects contaminated by vegetable matter of the cornea or globe or, by extension, of infection from adjacent paranasal sinuses. Fungal endophthalmitis and cho-rioretinitis, on the other hand, are usually the result of antecedent fungemia seeding the ocular tissue. Candida spp. are the most common cause - although initial infection with dimorphic fungi may lead to scarring of the chorioretina. Contact lens wear is associated with keratitis caused by yeasts, filamentous fungi, and Acanthamoebae spp. Most parasitic infections of the eye, however, arise following blood-borne carriage of the microorganism to the eye or adjacent structures.

HIV - The overall rate of ophthalmic involvement in children is around 35%. The most common finding was a non-purulent conjunctivitis, followed by perivasculitis of the peripheral retinal vessels and molluscum contagiosum.

Ocular toxocariasis is a rare infection caused by roundworms, Toxocara canis and Toxocara cati. Most common presentation is unilateral granuloma of the posterior pole or peripheral retina.

Toxoplasma - ocular involvement is serious because, in many cases, it affects the macular region. Besides retinochoroiditis, other ocular manifestations of congenital toxoplasmosis are described, such as microphthalmia, optic nerve atrophy, and abnormalities of the iris, cataract, and strabismus.

Gastrointestinal disorders

Ulcerative colitis: acute anterior uveitis in 6% children

Crohn's disease: acute anterior uveitis in 3%, nodular scleritis, diffuse and nodular episcleritis also seen.

Alagille syndrome: most common is posterior embryotoxon - a condition marked by thickening

of the ring that normally lines the cornea in the eye (usually benign). Rarely, Axenfeld anomaly, a condition in which strands of the iris are abnormally attached to the cornea, or pigmentary retinopathy.

Orbital disease

Metastases of neuroblastoma occur preferentially in the orbit, and can be bilateral in half the cases, producing exophthalmos with palpebral ecchymoses. Sometimes a mass can be felt at the level of the zygoma.

Ewing's sarcoma is also responsible for an ap preciable number of metastases in the orbit. The clinical picture is similar to that of neuroblastoma.

Hyperthyroidism, or Grave's disease can cause of exophthalmos

Non-accidental injury

Child abuse - The "shaken-baby syndrome" brings together intra-ocular and intra-cranial haemorrhages with raised intracranial pressure. There is no external sign of injury.

Birth injury – subconjunctival hemorrhage and sometimes multiple retinal hemorrhages. Severe ocular accidents by forceps delivery in the form of hyphema, corneal edema, facial palsy, and corneal abscess.

Chromosome anomalies

Downs syndrome - Cataract is common, and significant disorders or refraction are described, keratoconus which often terminates by corneal rupture, convergent strabismus, etc. Brushfield's spots, greyish- silver spots at the periphery of the iris, are found in 85 % of cases.

Other genetic anomalies are less common. Ocular lesions are seen in Turner's syndrome (refractive errors, amblyopia and strabismus), trisomy 13 (rarest and most severe of the three viable autosomal trisomies is associated severe ocular defects, including microphthalmos, iris coloboma, and retinal dysplasia), trisomy 18 (though various eye findings are reported, the more common ones include narrow palpebral fissures, ptosis, epicanthal folds, hypoplastic supraorbital ridges, punctal agenesis, discontinuous eyebrows, long eyelashes with distichiasis, hyper- or hypotelorism, and blepharophimosis), and cri du chat syndrome (hypertelorism, strabismus, downwardly slanting palpebral fissures, epicanthic fold).

Conclusion

A pediatrician should be aware of the clues to disease that the eye could provide and referral to a pediatric ophthalmologist is in order when the numerous conditions mentioned in this newsletter are being considered. Post graduates and we pediatricians must make it a point to see for self the findings that our pediatric ophthalmology colleagues pick up in our patients. The ophthalmologists in turn must be aware that their own findings may not be limited only to the eye. The skills of both are complimentary, and they will benefit from collaboration in the interests of the well-being of children.

The newsletter is not intended to be a complete, encyclopedic account of all those complex diseases giving rise to multiple clinical changes in the eye and elsewhere.

Cortical Visual Impairment

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Cortical Visual Impairment (CVI) is one of the most common causes of visual impairment in the developed countries[1] and is fast becoming so in developing countries too[2]. It is also referred to as Cerebral Visual Impairment or Neurologic Visual Impairment. Strictly speaking, the term cerebral visual impairment is better than cortical visual impairment because many causes of CVI involve subcortical structures in addition to or instead of the cerebral cortex. The definition of CVI keeps evolving, but it is characterized by impairment in visual acuity or visual field or the tasks guided by vision, in which there is disorder in the post geniculate visual pathways of the brain, and no ocular pathology.[3]

Causes and Pathophysiology

The most common cause of CVI is hypoxic ischemic damage in premature children. Other causes include neonatal hypoglycemia, traumatic brain injury, hydrocephalus, metabolic disease, structural brain anomalies and seizures.

Vision is a highly complex faculty comprising multiple components. Visual functions served by the occipital lobes include visual acuity, contrast sensitivity, color vision, visual fields, and in the adjacent middle temporal lobes, perception of movement.[4] The posterior parietal lobes serve the function of simulating an overview of the scene, identifying an object within it and guiding movement to it. The temporal lobes aid in recognizing objects, shapes and faces. Damage to these higher visual areas to any extent and in any combination affects vision or visual function. The two principal higher pathways affected by CVI are the occipitoparietal pathway (dorsal stream) and the occipitotemporal pathway (ventral stream).[5]

Characteristics of visual dysfunction in CVI

1. There is variability of visual function – worse in unfamiliar environments, patterned backgrounds (crowding phenomenon) or following a seizure.
2. Relative sparing of colour vision, impaired contrast sensitivity, looking away from the object the child is reaching out to (past pointing), increased latency of saccadic eye movements and fixation, and paradoxical light gazing.
3. Motion perception may be preferentially affected or spared. Perceiving motion in a non-seeing area has been noted in some CVI children and is termed blind sight.
4. Higher order visual perception abnormalities termed cortical visual dysfunction – impairment of recognition, depth perception, orientation, motion, and simultaneous perception. It is possible in some cases of CVI to have good visual acuity but impaired visual functioning like object recognition or motion perception.

Evaluation and Screening

Visual assessment:

is challenging because the normal methods of testing visual acuity and visual fields of pre-verbal children cannot be employed in most of these

cases. Therefore, we have to rely on questionnaires or scales administered to caregivers, in addition observing these children for their visual behavior for a longer period in a dedicated environment. The other major confounder is the neurologic and motor deficiencies which may be affecting performance.

Preferential looking tests (PLT) especially forced PLT are often used with some success for visual assessment.

Electrophysiology and Neuroimaging

Visual Evoked Potentials (VEP): On VEP it has been demonstrated that there is deficiency of global visual processing by higher cortical areas besides the primary visual cortex. However because of high variability in children and non specificity in CVI, it is currently of limited use for diagnosis of CVI. Sweep VEP shows some promise as compared to flash VEP.[6]

Periventricular leukomalacia is seen in many preterm babies with CVI. However absence of structural changes do not rule out CVI. Functional MRI (fMRI) or PET scan might provide more help in picking up these cases.

Diffusion Tensor MRI (DTI): This new technology enables visualization of axonal connections in the brain and shows potential in detailed assessment of structure-function relationship in CVI.[7]

As things stand, we need better imaging techniques to pick up the structural damage in the brain and to correlate it to functional disability. We also need better techniques to demonstrate the visual functional disabilities other than the classical visual acuity measurement. This will help identify children with disabilities and facilitate measures to help them with tasks they find difficult.

Treatment and Prevention

The best way to tackle this disease entity is to reduce its incidence it by advances in the management of premature babies and the hypoxic damage they are prone to. Meanwhile the best we can do now is to provide environmental modification and social support. Stem cell therapy and various methods of visual stimulation offer hope and need studies to assess their effectiveness. Management of comorbidities also may also play a small role in improving the visual function in the setting of CVI. Therefore, a multidisciplinary approach is needed to both detect and tackle the child's disabilities and also identify other skills that can then be employed towards habilitation.[8]

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Childhood Myopia : A Booming Epidemic

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INTRODUCTION

Myopia, or short sightedness, is one of the major causes of visual impairment in both developing and developed countries1-3. Normally, optical system of our eyes focusses light from distant objects onto retina in order to see objects clearly. However, in myopia, light from distant objects is focused in front of retina. Myopia is defined as spherical equivalence of −0.5 Dioptre(D) or greater and progresses rapidly from onset at an early age till early adulthood. Children with myopia present with several symptoms such as blurring of distant vision, headache, fatigue and poor scholastic performance. In this article, our aim is to discuss the global burden of childhood myopia, management options and preventive strategies for progressive myopia.

THE GLOBAL BURDEN OF MYOPIA AND THE INDIAN SCENARIO

Paediatric myopia needs to be considered as an emerging global pandemic in view of its rapid increase in prevalence. Prevalence of myopia varies with country, age and ethnic group and it is as high as 70 %- 90% in East Asia, particularly, in countries like Japan, South Korea, Singapore, Taiwan, Hong Kong and China though much lower rates have been reported from South Asia and India4-9. Tables1 & 2 show prevalence of myopia in India among and urban and rural paediatric population respectively10-11.

Table 1: Prevalence among urban children10-11

Age (in years)	Prevalence
5	4.7%
10	7.0%
15	10.8%

Table 2: Prevalence among rural children10-11

Age (in years)	Prevalence
7	2.8%
10	4.1%
15	6.7%

WHY DO WE NEED TO CONTROL MYOPIA ?

- Myopia progression in East Asian children is as high [−1 dioptre (D) per year]7-8. The North India Myopia study in school going children in Delhi, found an annual incidence of 3.4%, prevalence of 13.1%, progression in 49.2% children and 28.2% showing myopic progression of ≥ -0.5D12-13. It is estimated that by 2050, half of the global population would become myopic with high myopia among at least one-fifth of them. Thus, myopia is a global health problem not only due to its visual complications but also being a significant economic burden.

ETIOLOGY:

Myopia is the result of interplay of complex genetic and environmental factors. Studies

have shown that number of myopic parents and monozygotic twins have greater correlation with myopia than dizygotic twins14-15. Prolonged intense near work, reduced outdoor activities and urbanization are the major environmental factors responsible for myopia. Premature and low birth-weight infants have also been found to be more at risk of developing myopia16-17.

PREVENTION OF MYOPIA PROGRESSION

Early onset of childhood myopia is associated with high myopia in adult life. High myopia eventually can lead to several vision threatening complications such as retinal detachment, macular degeneration, early onset glaucoma and cataract. Incidence of retinal detachment increases with the degree of myopia18 (0.015% in > 4.74 D myopia, 0.07% in > 5.00 D myopia, and 3.2% in > 6.00 D myopia). Risk of developing macular choroidal neovascularization19 increases with the level of myopia (likelihood as high as 2-fold between 1.00 to 2.00 D myopia, 4-fold between 3.00 to 4.00 D myopia, and 9-fold for 5.00 to 6.00 D myopia). The ultimate goal of myopia control therapy is to slow myopic progression during the years of most active eye growth thus minimizing the sequelae.

MANAGEMENT & CONTROL

Different modalities include environmental, pharmacological, and optical options.

1. Environmental: Increasing the outdoor time is an important method in controlling the myopia. Possible theories which explain this include retardation of eye growth by the light towards the end of UV spectrum, stimulation of dopamine by high luminance which prevents axial growth, supply of Vitamin D, decreased accommodative demand and increased depth of focus and hence retinal image quality due to small pupil in light.
2. Pharmacological: Various pharmacological options include topical application of Pirenzepine, and 7- methyl xanthine are being evaluated but results with topical low dose atropine have been most promising.

- i)Atropine:.Wells in 19th century, first reported the use of atropine to slow myopia progression. It has been consistently found to regulate myopic progression with a good safety profile.

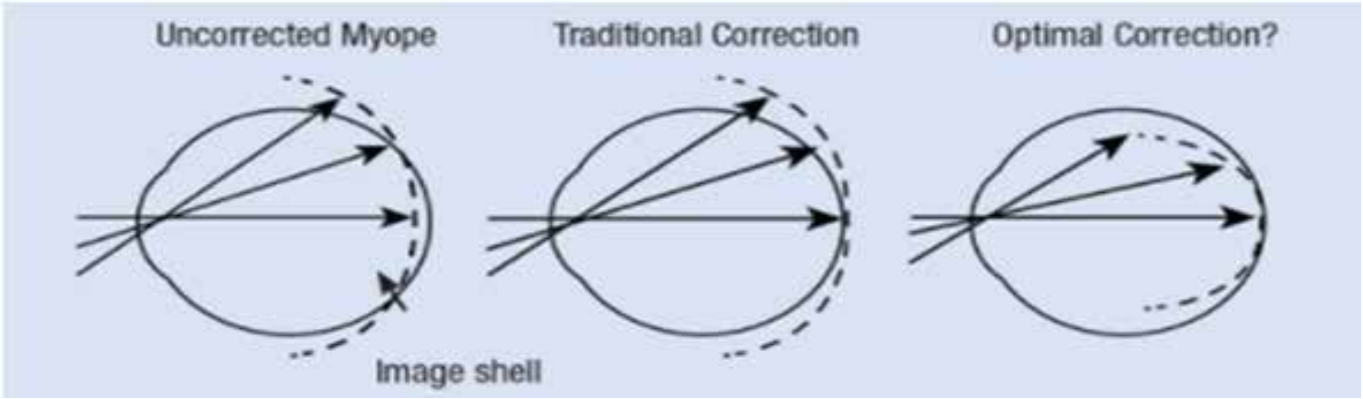
Mechanism of action

- The changes that occur in the sclera during myopia development represent a complex interaction between tissue remodeling, synthesis, and degradation which lead to axial elongation. Atropine blocks muscarinic receptors in ciliary muscle, retina and sclera thus preventing scleral remodeling. It also dilates the pupil which increases the exposure to UV rays thus hardening the collagen and thereby minimizing chances for axial elongation.
- The ideal dosage for the atropine administration remained a topic of debate. ATOM1 study20 reported that 1% atropine dose had the maximum efficiency of 77% in slowing down progression, but not without side effects. Low concentration of 0.01% atropine had a better treatment-to side effect ratio with fewer side effects and rebound after drop cessation but less effect on axial length increase21. LAMP 2 Study22 showed the 0.05% atropine dose to be optimal concentration with a 64.5 % efficiency in slowing down the myopia progression with no considerable axial length increase.

3. Optical:

Conventional strategies include single vision spectacles and soft / RGP contact lenses but they are ineffective in control of myopia progression.

Optical Options for progressive myopia: How do they work?



Pic courtesy Clin Exp Optom 2018 DOI:10.1111/cxo.1266

Most work on the principle of reducing the peripheral hyperopic defocus which acts as stimulus for further axial length elongation.

1. Bifocal/ Multifocal spectacles: are especially useful for children with an accommodative lag. With a near addition of +1.00 to +2.00 D, accommodative demand is reduced and myopic shift is induced in the peripheral retina which may remove stimulus for myopia progression.

2. Peripheral Defocus Spectacles (DIMS/ HALT)

Defocus Incorporated Multiple segments (DIMS) technology works on the concept of creating simultaneous defocus, during both distance and near viewing - one plane on the retina due to the single vision zone(s) of the lens, and one plane creating myopic defocus due to the +3.50D defocus lenslets. Highly Aspherical Lenslet Target (HALT): introduces the concept of volume of myopic defocus. Here, light rays are deviated continuously in a three-dimensional manner that creates a volume of myopic defocus.

3. Soft bifocal/ Multifocal Contact lens: Uses the technique of changing the center distance design to reduce the myopic progression
4. Extended Depth of focus Contact Lenses (EDOF) uses neuro focus optics technology where in it creates an extended depth of focus enabling around 2 times clearer vision compared to conventional glasses at all distances.
5. Orthokeratology contact lenses: The technique underlies in inducing peripheral myopic defocus in both vertical and horizontal meridians thereby enabling reduction in accommodation lag and improvement in accommodative facility. These lenses are worn overnight and the idea behind these lenses are to flatten the central zone of cornea surrounded by cornea steepening.

AVAILABILITY OF VARIOUS Rx:

Currently low dose atropine (0.01%), bifocal/ multifocal spectacles and orthokeratology contact lenses are available in India and rapid myopia progressors may be put on a combination therapy also.

CONCLUSION

- Recent findings suggest that environmental conditions, including intensive education and limited time spent outdoors, are directly related to the high prevalence of myopia in all ethnic groups23-24. With the onset Covid-19 pandemic, with more and more children transitioning in to e-learning and no outdoor play; time is now more than ever to follow up children who are at risk for myopia and who already have the diagnosis of myopia, especially progressive myopia. Future of myopia control lies

in creating myopia awareness, recognising it as a public health crisis, studies on genetic markers, bringing about environmental changes including effective regulation of near work and screen time in children.

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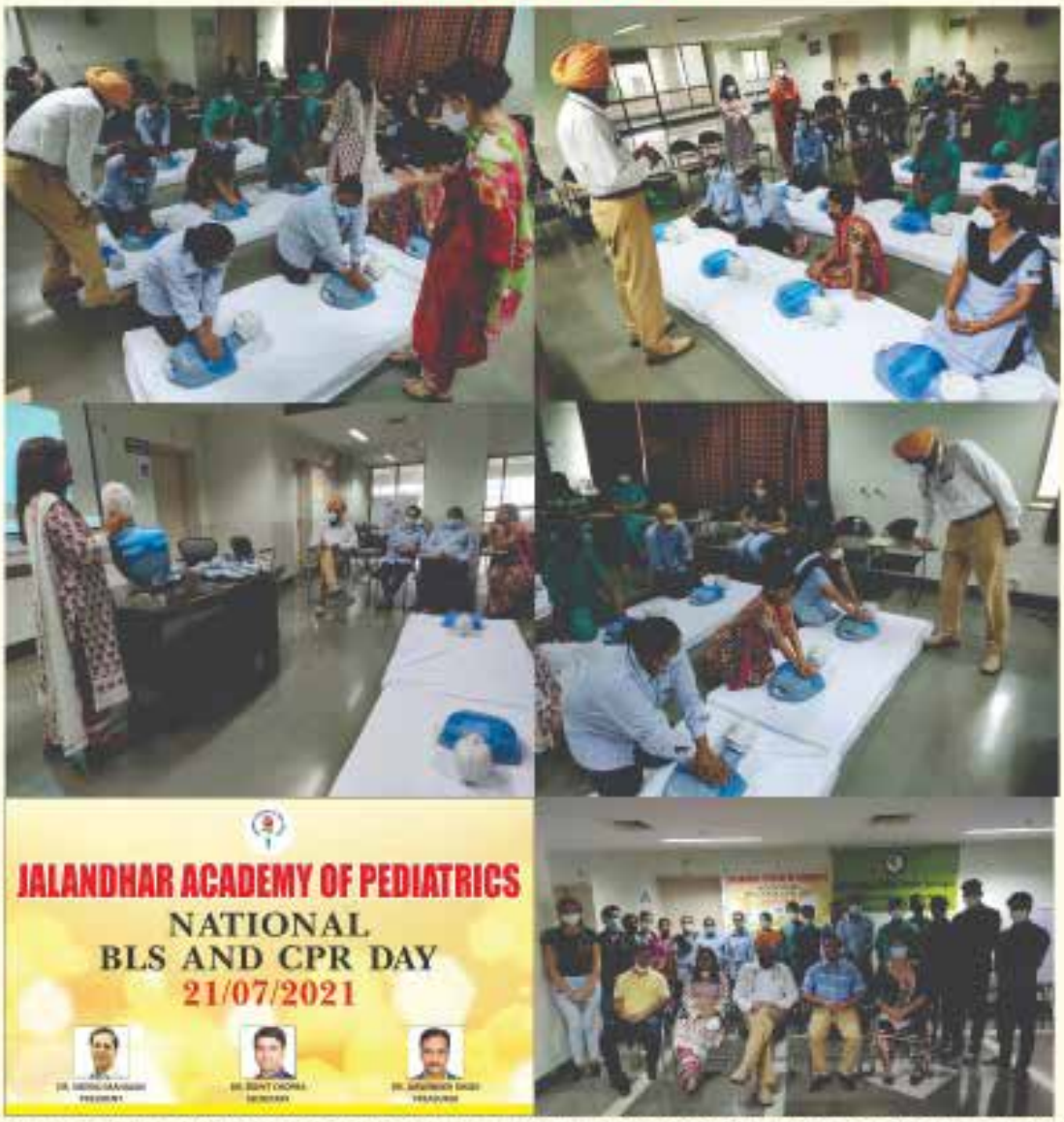
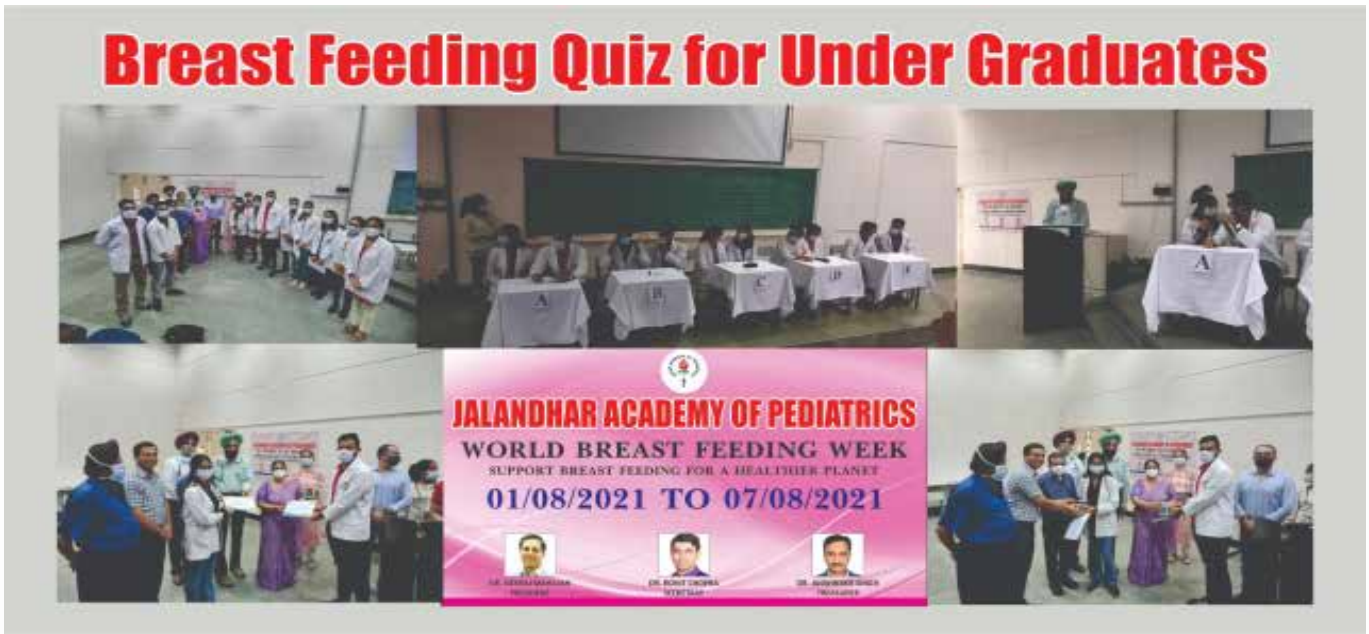
IAP Jalandhar

Jalandhar Academy of Pediatrics and Dept. of Pediatrics, PIMS Medical College, Jalandhar Celebrated National BLS and CPR Day ON 28/7/2021



JALANDHAR ACADEMY OF PEDIATRICS IN COLLABORATION WITH DEPT. OF PEDIATRICS, PIMS MEDICAL COLLEGE, JALANDHAR Conducted BLS Mass Awareness Course for house-keeping staff at IAP CPR Centre, PIMS. Course was attended by more than 20 participants who learnt the skills of CPR and choking management. Faculty includes Dr. H.S. Bains, Dr. Anuradha Bansal & Dr. Pushwinder Kaur.

IAP Jalandhar



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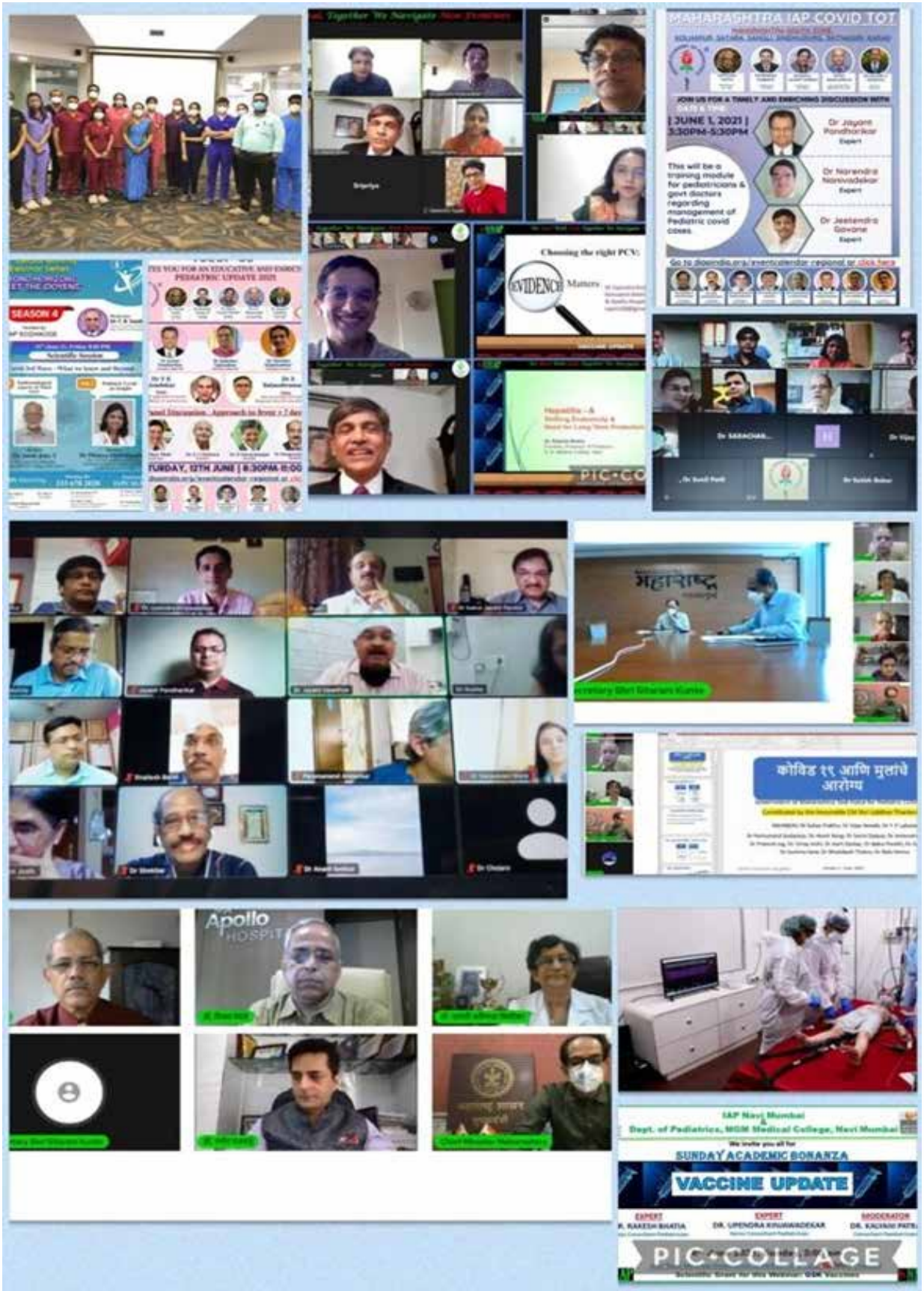
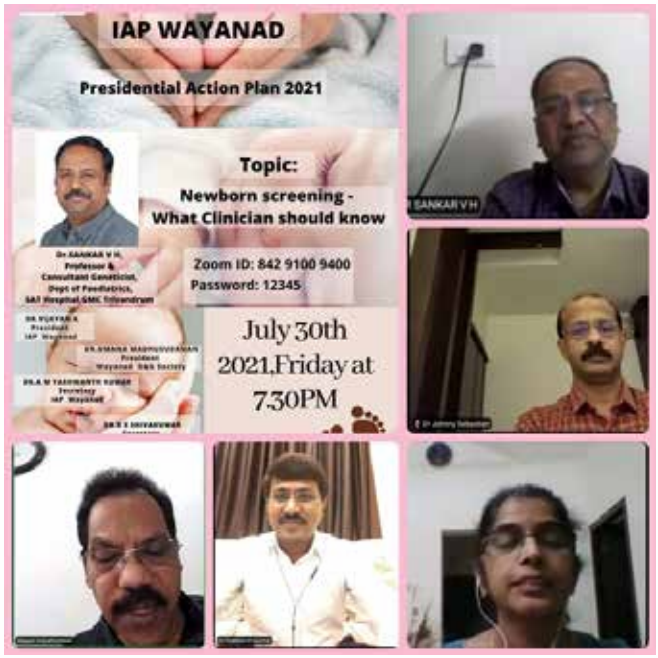
IAP Navi Mumbai



IAP Navi Mumbai

IAP Kerala


IAP WAYANAD
Presidential Action Plan 2021
Topic:
Newborn screening -
What Clinician should know
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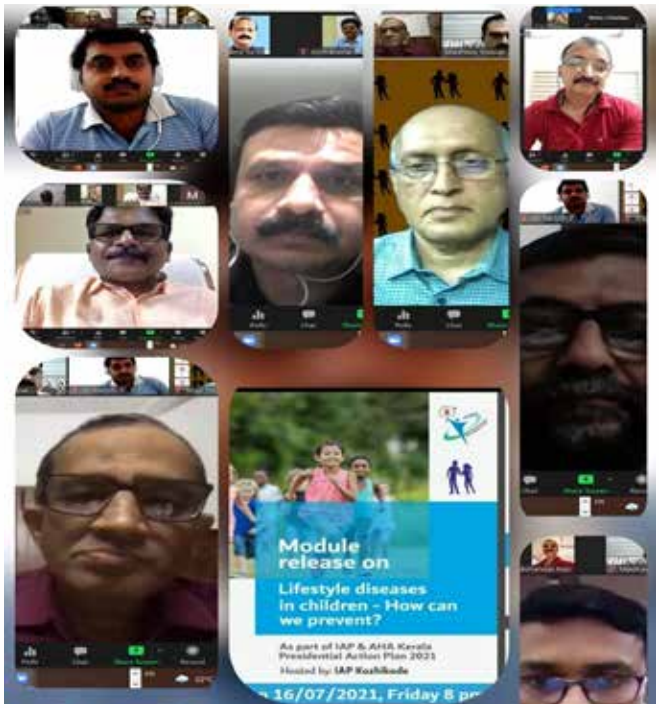
Password: 12345
July 30th
2021,Friday at
7.30PM


ORS WEEK
Celebrations 2021
(25th to 31st July)
Theme
ORAL REHYDRATION
NO MORE DEHYDRATION
ORS WEEK 2021


IAP VADAKARA
ORS Truck
RJ Jeena
CLUB FM 94.3


Module
release on
Lifestyle diseases
in children - How can
we prevent?

IAP Kerala

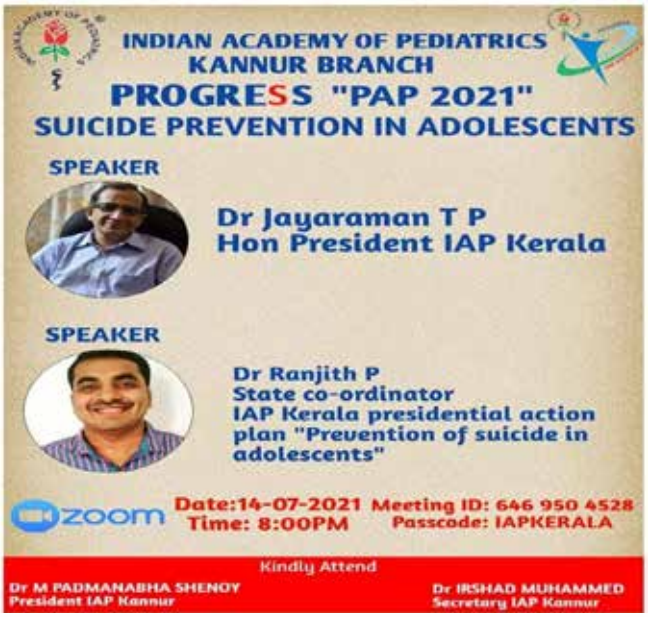

പ്രഥമ ശുശ്രൂഷയുടെ
പ്രഥമ പാഠങ്ങൾ
ആരോഗ്യ പ്രവർത്തകർക്കും
രക്ഷിതാക്കൾക്കും
ഉള്ള പരിശീലന പരിപാടി


ORS WEEK 2021 25TH TO 31ST JUL
ORAL REHYDRATION
NO DEHYDRATION
ORS


PEDIATRIC
TRAUMA MANAGEMENT


ZIKA VIRUS


Early Diagnosis of Inborn Errors of Immunity
CASE-BASED PANEL DISCUSSION ON COMMONLY ENCOUNTERED INBORN ERRORS OF IMMUNITY
Dr.Manjusha K


INDIAN ACADEMY OF PEDIATRICS
KANNUR BRANCH
PROGRESS "PAP 2021"
SUICIDE PREVENTION IN ADOLESCENTS
SPEAKER
Dr Jayaraman T P
Hon President IAP Kerala
SPEAKER
Dr Ranjith P
State co-ordinator
IAP Kerala presidential action plan "Prevention of suicide in adolescents"
zoom
Date:14-07-2021 Time: 8:00PM
Meeting ID: 646 950 4528
Passcode: IAPKERALA
Kindly Attend
Dr M PADMANABHA SHENOY
President IAP Kannur
Dr IRSHAD MUHAMMED
Secretary IAP Kannur