

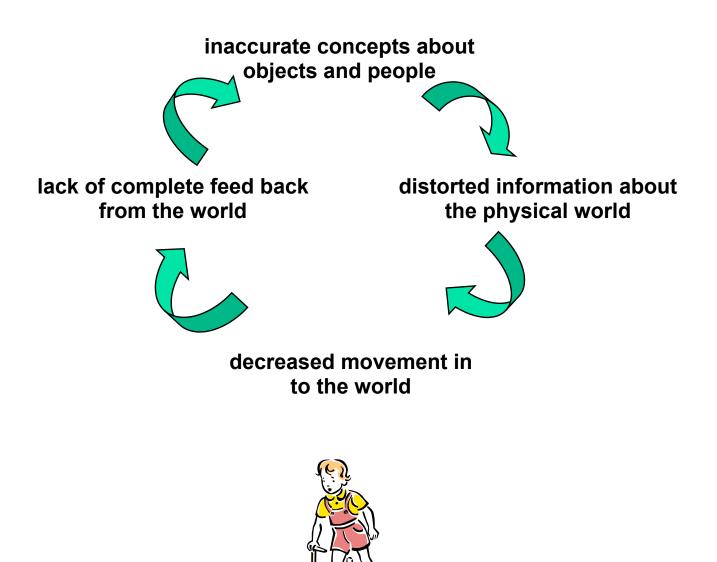
# CHILDHOOD BLINDNESS



# ARAVIND EYE CARE SYSTEM

# **CHILDHOOD BLINDNESS**

# Impact of Vision loss on Development



#### Definitions

UNICEF defines an individual aged less than 16 years aged less than 16 years as a child. A child is considered blind, if he/ she has a corrected visual acuity < 3/60 in better eye. A child is considered to be severely visually impaired if he/ she has a corrected visual acuity between 3/60- 6/60 in the better eye.

#### Prevalence

Limited data is available from the developing countries from the whole population surveys, disease specific prevalence surveys and community- based rehabilitation programmes (CBR). Using estimated based on Under 5 years mortality rates, the prevalence of blindness ranges from 0.3/1000 to 1.5/1000 children, being approximately one tenth as frequent as blindness in whole populations.

#### **Avoidable Blindness**

Avoidable causes are those which are either entirely preventable, or where treatment can prevent blindness or restore sight. The relative importance of the different avoidable causes of blindness varies considerably from region to region. Overall, approximately 40% of the blindness in children is avoidable. However a significant proportion of the "unknown" causes certainly represent preventable conditions which cannot readily be diagnosed (for example congenital rubella syndrome). 40% therefore represents the minimum estimate.

Preventable conditions				
Corneal scarring due to		Perinatal Facto	ors	
Vitamin A deficiency		Retinopathy of Prematurity (ROP)		
Measles infection		Birth hypoxia		
Ophthalmia neonatarum		Hereditary dise	ease	
Infective corneal ulcers		Risk counseling for dominant disease		
Harmful traditional practices				
Intrauterine factors				
Rubella				
Toxoplasmosis				
	Treatable	condition		
Cataract	ROP		Corneal diseases	
Glaucoma	Uveitis		Corneal ulcer/	
			opacity	

## Table 01. Avoidable causes of blindness

	ROP	Cataract in Childhood	Glaucoma in Childhood
Amount of blindness in blind schools	20%	15%	8%
Incidence /10,000 births	4	4	1
Cases/million population	10	10	2
Diagnosis	Stage 3 disease: Raised ridge with fibrovascular proliferation and posterior vessel tortuosity	Abnormal red reflex White pupil	Large eye Hazy cornea Raise IOP Cupped disc
Treatment	Cryo. or laser to the avascular zone, 360° circumference.	ECCE surgery with IOL	Goniotomy or Trabeculotomy or Others
Problems	Awareness Screening - paediatricians - ophthalmologists	Late diagnosis. Ref error correction Amblyopia	Late surgery Long-term Control

 Table 02. Surgically Avoidable childhood blindness

#### **EPIDEMIOLOGY**

#### Major causes of blindness in children.

There is marked variation n the major causes of blindness in children in different parts of the world. In poor countries of the world, corneal scarring due to vitamin A deficiency, measles, ophthalmia neonatarum and harmful traditional practices predominates. Globally, corneal scarring is the single most commonest cause of blindness in children in schools forte blind. In middle income countries, the commonest causes are retinal conditions, mainly retinal hereditary dystrophies and retinopathy of prematurity. Central nervous disorders, due to a range of conditions and retinal conditions are the commonest causes in high-income countries.

#### Trends in major causes of blindness

The major causes of blindness in children are largely determines by levels of socioeconomic development and health care provision. Causes of childhood blindness can vary over time in response to changing economic development and the introduction of health interventions. For example, infectious causes were important in Saudi Arabia, but now hereditary diseases predominate. Retinopathy of prematurity was the single most common cause of blindness in Europe during the 1950s but now accounts for only 5-15%. Retinopathy of prematurity is becoming an important cause of blindness in middle income countries as they introduce neonatal intensive care services. A reduction in corneal blindness has been documented in Asian and African countries, in response to improved measles immunization coverage rates.

## Table03. Causes of childhood blindness in different countries

## (percentage)

Causes	China	India	Czech	Chile	Jamaica	S Lanka	U.K.
Whole	25.5	25.3	10.9			35.8	
globe	4.4	26.4	1.8	6	5	2.2	0
Cornea	18.8	12.3	8.7	9	39	17.3	4
Lens	24.9	20.7	5.2	47	15	22.1	28
Retina	1.5	5.8	54.2				
Uvea	13.6	5.9	15.3	13	18	7.5	20
Optic nerve	9	2.6		8	15	5.8	1
Glaucoma	2.3	0.9	3.9	17	8	9.3	47
Other							
Hereditary	30.7	22.9	9.2	30	N/A	35.0	50
Intrauterine	0.1	1.8	0.4	8		3.5	4
Perinatal	2.3	1.4	43.7	23			33
Childhood	14.0	27.9	4.4	11		5.3	11
Unknown	52.9	46.0	42.3	28		56.2	00

N/A is not available

LEVEL	NEONATES	PRE-SCHOOL	SCHOOL	
Primary Community	Prevent ophthalmia neonatorum Examine babies eyes	Screen for amblyopia Prevent xerophthalmia	Screen visual acuity	
Secondary Mid-level	Refer cataract and glaucoma	Treat corneal disease	Provide spectacles	
Tertiary Referral	Screen & treat ROP Treat cataract and glaucoma	Specialist surgical Low vision services	Treatment of severe ocular injuries Low vision services	

 Table 04. Control of childhood blindness according to the age and health services

Anatomical Level	Number/ million population	PRIMARY (Prevent the disease)	SECONDARY (Prevent Visual Loss)	(Restore Vision)
CORNEA		Nutrition Education Measles immunisation	Early treatment of corneal Disease	Corneal grafting Low vision services
LENS		Rubella immunisation Genetic counselling	Early surgery Amblyopia treatment	Early good surgery Good follow-up Low vision services
RETINA		Avoid low birth weight Avoid hyperoxia	Screening for ROP and treatment	Low vision services
GLAUCOMA		Rubella immunisation Genetic counselling	Early, good surgery Good follow-up	Low vision services
OPTIC N /H.V.P		Good ante-natal and peri-natal care		Low vision services
WHOLE		Avoid medication in pregnancy		Low vision services
Total	200-400	/million total population	or 0.5/1000 childre	en

Table 05. Control of blindness according to the disease

#### PAEDIATRIC EYE

Of all human organs, the eye is the most fully developed at birth. Many changes occur with maturity, but the absolute dimensions of the eye are closer to adult size than nearly any other organ of the body.

In the prenatal period, the eye grows fastest between the 8th and the 14th week.

Overall growth of eyeball parallels growth of the embryo until the 30th week after which eye growth slows.

#### The Premature eye:

Premature infants (30-35 gestational weeks) have shorter ocular axial lengths, steeper corneas and higher lenticular refractive power than full term infants. The pupils are miotic and remnants of tunica vasculosa lentis are frequently present. Bilateral symmetrical lens opacities may occur in premature infants. Vascularisation of the retina is completed nasally during the eighth gestational month and temporally during the ninth month. The peripheral retina has a silver grey appearances in areas in which vascularisation is incomplete.

## The Infant eye:

By gestational weeks, the mean axial length of the eye increase, the cornea flattens and the lens power decreases. The peripheral retina is fully vascularised and the extrafoveal retina functions are nearly the adult level; however the fovea is still immature.

## Growth after infancy:

The human eye grows rapidly during the first year of the life. During the first 6 weeks of life the cornea flattens, the axial length increases, and the power of lens decreases.

#### **VISUAL ESTIMATION IN CHILDREN**

The basic function of the eye is to see and the assessment of visual acuity deals with estimating the maximum ability to see. In adults the test is simple and standardized by using the Snellen's charts at 6 meters distance. It is different for the children as there verbal responses are not even sufficiently developed and in verbal children the knowledge of letters is also needed.

#### In Infancy

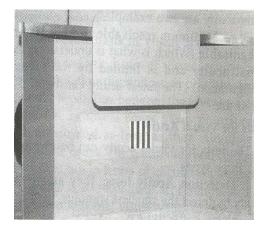
#### Catford drum.

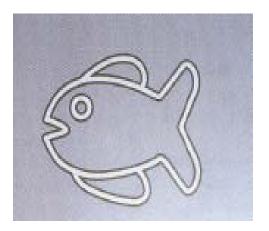
It is useful in infants and preschool children

✤ Preferential looking tests (PLT).

Based on behavioral pattern of an infant to prefer to fixate a pattern stimulus rather than a blank, both being of the same brightness: space average luminance.

Teller acuity cards (Fig.1a) Cardiff acuity cards (Fig.1b)





#### Fig.1a



#### Visually evoked responses.

VER records the change in the cortical electrical pattern detected by surface electrodes monitoring the occipital cortex following light stimulation of the retina.

Vision testing in 1-2 years Boeck candy bead. Sheridan's balls test. In 2-3 years Miniature toys test. Dot visual acuity test. Allen Preschool vision chart





#### In 3-5 years

#### Tumbling E.

It consists of different sizes of E in one of the four positions: right, left, up or down. After familiarizing the child, he indicates which direction the E is oriented. It is done at a distance of 6 meters and each eye is tested separately.

#### Landolt's C. Sheridan's letter test.

Lippman's HOTV test.

It is important to note the uniocular visual acuity which can be done in mass screening at school so that permanent visual loss due to amblyopia can be prevented.

#### INFANTILE CATARACT

#### Etiology

The formal definitions of the terms infantile, developmental and congenital cataracts differ somewhat. In contrast to congenital cataracts, infantile and developmental cataracts may not be present at birth. It is useful to think of congenital cataracts as those that occur with in the first year of life. Cataract in children comprises of 15% (Dandona et al) of the total childhood blindness i.e. nearly 30 children/ million population are blind due to cataract. At least 10 new cases/ million population/ year, nearly 1/2000 live births. In many cases, the cause of the cataract cannot be determined. Several morphological classification schemes have been suggested for congenital cataracts. A simplified version adapted from Merin is summarized in table 02

 Table 06. Morphological classification of infantile cataract

1.	Polar (Fig	Zonular		
2.	3b)	Nuclear	Spearlike	
3.	Total	Lamellar (Fig.	Coralliform, Floriform	
4.	Membranous	3a)	Capsular	
	Zonular	Sutural		

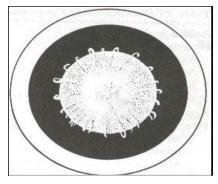


Fig .3 a Lamellar cataract

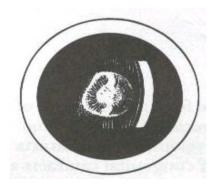


Fig .3b Anterior Polar cataract

#### Main causes of congenital cataract are the following:

- ✤ Congenital Rubella (Fig. 4b)
- ✤ Hereditary
- Traumatic (uniocular)
- Other causes like drug induced.

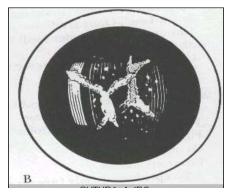


Fig .4a Sutural cataract

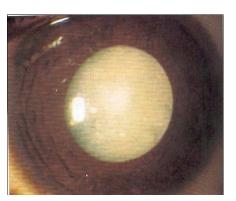


Fig.4b Rubella cataract.

## **Congenital Rubella (Fig. 5)**

An estimated 238000 children in developing countries are born with congenital rubella syndrome (CRS) every year. The number blind from CSR cataracts is unknown. In a hospital based study in southern India, 25% of infantile cataracts were associated with congenital rubella.

Rubella cataract is a part of highly destructive congenital syndrome, caused by maternal infection with rubella virus



contracted during the first or early second trimester of pregnancy. The syndrome

#### Fig.5 CRS

includes malformations of heart, deafness, mental retardation, dental defects and ocular disturbances such as cataracts (which are bilateral in 75% of the cases), congenital glaucoma, retinal disease and microophthalmos. The cataract is obvious at birth as white opacities.

#### Management

The treatment of congenital cataract depends on three major factors:

- ✤ Laterality
- The presence of associated ocular abnormalities
- Whether the cataract is total or partial?

Unilateral cataracts often carry a poor prognosis because of severe amblyopia and associated strabismus of the involved eye. Visual acuity of 20/400 or worse is common if the condition is untreated and visually significant. The prognosis for untreated partial unilateral cataract is as poor as for untreated complete unilateral cataracts. The treatment is early surgical removal of the cataract.

Bilateral complete cataracts have a more favorable prognosis. There is little disagreement about the treatment, which entails prompt surgical removal of the cataracts, operating on the second eye soon after the first. The treatment of bilateral

partial cataracts is more controversial. The difficulties encountered in measuring the visual acuity early in life interfere with assessing the visual significance of the cataracts.

#### The following are the general rules:

- Cataracts that are less than 2 mm in diameter are usually not significant.
- The density of the cataract is more important than the size and type.
- Cataracts that obstructs the examiner's view of the fundus or prevent objective refraction of the patient usually should be extracted.
- Prompt surgical removal of a cataract is indicated if the contralateral cataract has been removed.

Children with cataract do not present themselves. The cataract must first be recognized by someone else- usually a member of the family, sometimes a teacher or a trained health worker. Cataract in children is very different from cataract in adults. The surgery is more complex and the postoperative follow-up care is much more demanding. Children are not born with normal vision; they have to learn to see by using their eyes constantly during the first few months of life.

Children's eyes behave differently from adult eyes. Severe intraocular inflammation is more common. If the posterior capsule is left intact, it will invariably become opaque. The eye is growing until the child is two years old, which leads to frequent changes in he refractive state of the eye.

## Surgery

In the modern era the standard protocol is to do an extra capsular cataract extraction and implant the intraocular lens (IOL) in children above 2 years of age.

Below 2 years of age it is preferred not to put an IOL due to various reasons,

The growth of the eyeball is mainly in the first two years of life and since the eye is increasing in size the power may change after few years.

- Moreover the IOL implant will have a particular size and may be decentered due to increased available space as the eye grows
- Paediatric eyes are known to cause an inflammatory reaction to any surgery compared to adult eyes, lesser the age more the reaction.
- Though there has been lot of dispute about the material of IOL that should be put. Acrylic lens have been claimed to have lesser posterior capsular opacification.

## Correction of aphakia:

Rahi et al conducted a blind school study and found that 40% of the total cataract blindness was due to uncorrected aphakia and amblyopia. Multiple options are available for correction of aphakia. The major ones are

#### **Spectacles**

Spectacles are often used for postoperative patients. They are not very useful for monocular aphakia because of image size discrepancy between the two eyes. However, children adapt more easily to the magnification distortions caused by eyeglasses. The visual needs in infants are limited to their immediate surroundings.



Fig.6 Showing spectacles in children

## **Contact lens**

Contact lens are useful for the compliant children. The image distortions are not much and thus can be a useful tool to prevent amblyopia



Fig.7 Contact lens

#### The problems are

- Cleanliness and hygiene, as children are expected to go anywhere and everywhere and put the fingers straight to eye.
- Environment which is dry and dusty, windy are not suitable for contact lens
- Wearing and taking out can be a problem for the pediatric patients.
- Although tolerance is good with the high O<sub>2</sub> permeable lens lost lens, allergy, itching and discomfort are common in children

#### Intraocular lens

Though long term safety of IOLs is not known, they are the best options to combat aphakia. All the distortions of glasses can be taken care of. Contact lens problems can be easily tackled. Few important features however are to be considered,

- IOL surgery should be done by an experienced surgeon, as Pediatric eyes have to last longer than the adult eyes. Chances of reaction are high.
- Power of IOL is to be individualized as the eye is still in the growing phase, though lots of corrections have been suggested it is up to the ophthalmologist experience that help him to select the right power.
- The commitment is for a longer period because of the postoperative care
   Amblyopia is to be treated aggressively.
  - If IOL has not been done in primary setting, surgeon has to do it in a secondary sitting.
  - All the patients of congenital cataracts develop posterior capsular opacification within few months, so second surgery for membranectomy has to be considered.

#### Control

- b. Primary : Rubella immunization
- c. Secondary: treat aphakia and amblyopia
- d. Tertiary: Early, good surgery, excellent follow-up (low vision services)

#### **RETINOPATHY OF PREMATURITY (ROP)**

#### Definition

Retinopathy of Prematurity (ROP) occurs in preterm babies because the normal process of vascularisation of the peripheral retina is not complete at the time of birth. The developing vasculature is sensitive to biochemical changes (such as blood gas levels), which can lead to the formation of abnormal blood vessels in the back of the eye. These blood vessels can leak and bleed and abnormal scar tissue causes the retina to detach.



Fig.8 showing a white reflex retinopathy of prematurity

#### **Regional variation of ROP**

#### Established market economies

The "first epidemic" occurred in the West during the 1940s and 1950s and was the single commonest cause of blindness in many industrialized countries. Industrialized countries are witnessing a "second epidemic" of blindness in children due to ROP. The reemergence of the condition has come about as a result of better intensive neonatal care, with improvements in survival of extremely premature low birth babies.

#### Middle income countries

There is evidence that ROP is an increasingly important cause of blindness in middle-income countries. Birth weight of children blind from ROP in these countries shows a much wider range than in high economy countries (600-2240g)

#### Classification

ROP can be classified according to the region of the retina affected (zones 1,2 and 3), the stage of the disease (stage I- V) and other features:

"Plus" disease, which denotes breakdown of blood retinal barriers.

Threshold disease, which is defined as five continuous clock hours, or a total of eight hours, of Stage III "plus" disease.

Cicarticial disease, from scarring, which causes traction and distortion of the retina. Stage V causes blindness due to retinal detachment.

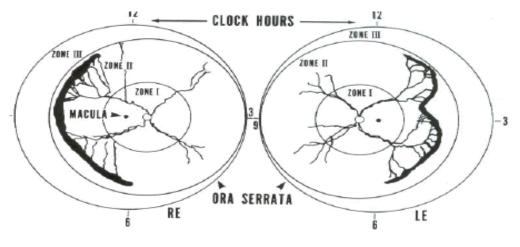


Fig .9 ROP zones

Zones I: the inner zone, extends from the optic disc to twice the disc macular distance, or 30 degrees in al directions from the optic disc

Zone II, the middle zone, extends from the outer border of zone I to the ora serrata on the nasal side and approximately the equator on the temporal side.

Zone III, the outer zone, extends from the outer edge of zone II in a crescentric fashion to the ora serrata.

#### Management:

#### Prevention of ROP

As the advanced stages of ROP are not amenable to treatment, emphasis needs to be on prevention. Primary prevention would entail preventing pre-term birth and providing excellent intensive neonatal care of premature, low birth weight babies, including blood gas monitoring and control.

Secondary prevention requires the development of screening programmes to identify babies needing peripheral retinal ablation i.e. those with stage III "plus" threshold disease. All babies included in a screening programme for ROP should also be followed up to identify and manage these other ocular problems.

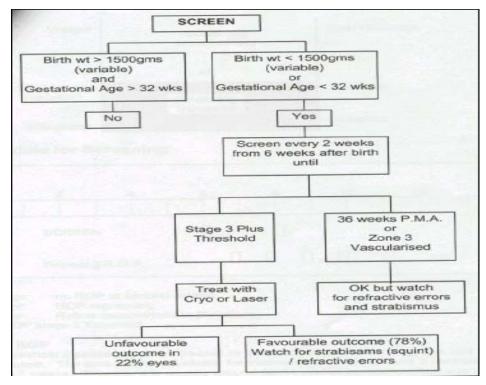


Fig :10

#### Treatment

Stage 3 plus threshold disease should be treated as soon as possible after the diagnosis and within 1 week at the latest. The time window available for treatment, and retreatment if necessary, is short- about 2-3 weeks. Treatment is usually around 36-44 weeks post-conceptual age (mean 37.7 weeks). Treatment can be performed in the neonatal unit under sedation and local anesthetic drops. It is important to have a neonatalogist present when treatment is being given.

Cryotherapy or laser is applied to the whole of the area of avascular retina. Following treatment the infant should continue to be seen at regular intervals for follow- up. The results of cryotherapy for stage 3 plus disease reduce the progression to stage 4 and 5 disease from approximately 50% to 25%. A scheme of screening and further treatment has been shown in figure 10.

#### CORNEAL BLINDNESS

#### **Causes and Management**

Corneal blindness remains the leading cause of blindness among children. In Africa and Asia, up to 70 % of childhood blindness is due to corneal disease. Corneal scarring related to vitamin A deficiency is probably the single largest cause of childhood blindness.

Vitamin A is important for maintenance of healthy epithelium of skin and mucous membrane. It is vital for mucus secreting goblet cells of conjunctiva and other mucous membranes of the body. Vitamin A requirement of growing child is much more than an adult. It is almost 5 times in infants (65mg/ kg) compared to adults (12 mg/ kg).

Night blindness is the first manifestation of vitamin A deficiency. Dryness of conjunctiva (xerophthalmia XIA) follows next. Initially there is loss of lusture of cornea and conjunctiva. Frank xerosis develops later on. Xerosis is caused by decreased mucus production by goblet cells.

Bitot spots are found in limbal region in interpappebral area of cornea and conjunctiva. Bitot's spots are triangular in shape with base toward cornea. They are usually temporal to the cornea in both eyes.

## Table07. Classification of xerophthalmia by WHO

Primary signs	Secondary signs	
X1A Conjunctival xerosis	XN Night blindness	
X1B Bitot's spot's with xerosis	XF Xerophthalmia fundus	
X2 Corneal xerosis	XS Xerophthalmia scars	
X3A Corneal ulceration with		
xerosis		
X3B Keratomalacia		

Other causes of corneal blindness include

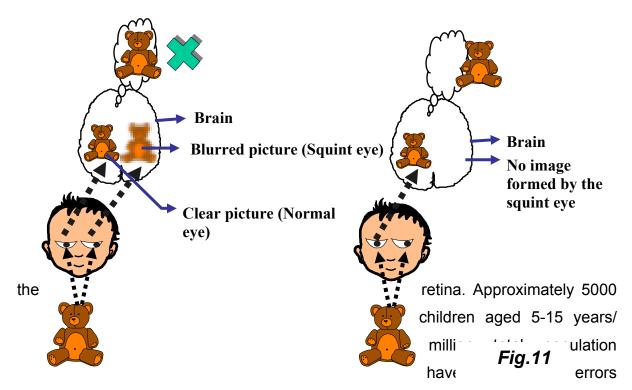
- Measles infection
- Application of traditional eye medicines
- Ophthalmia neonatarum
- Herpes simplex keratitis
- Other corneal infections

## Management

Treatment includes 200,000 IU of vitamin A orally on diagnosis followed by the same dose on next day and 2 weeks later. Aqueous vitamin A may be given intramuscular injection in a dose of 100,000 IU. Since vitamin A deficiency is common in developing countries vitamin A supplement program is designed. In this 200,000 IUI of vitamin A are given every 6 months to children in India.

## **REFRACTIVE ERRORS**

For clear sight, the cornea (the clear front window of the eye) and lens (behind the pupil) must properly focus or refract the light rays on retina, which lines the back wall of the eye. Conditions in which the shape of the eye affects the way the eye refracts or bends light and focuses on



greater than -1.00 dioptre sphere in both eyes. (variable prevalence).

#### Types:

#### It can be broadly divided into

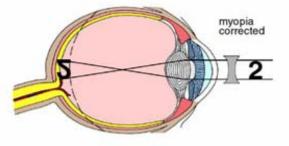
I. Myopia or short sightedness, a condition in which the eyes can see close objects but are unable to see distant objects clearly. The word myopia comes from the Greek word for *closed eyes*. Myopia when inherited is often detected in children eight to twelve years old. The eye grows along with the growth of the body but the growth in the eye is so much that it lengthens the eye to cause difficulty in focusing of rays.

- II. Hypermetropia or long sightedness, a condition in which the eye is usually shorter than normal (opposite of myopia). This shortness make it difficult to focus the light from close up objects clearly on the retina and hence light focuses somewhere behind the retina.
- III. Astigmatism. For normal undistorted vision, the cornea should be smooth and equally curved in all directions. When an individual has astigmatism, the cornea is "warped" or unevenly curved with distortions which means it curves more in one direction than the other.

#### **Treatment Options:**

Many treatment options are available depending upon the type of the refractive

error. There are two main options available, glasses and contact lenses. Both can be availed, both have their advantages and disadvantages. In children it is better to use spectacles because contact lenses, though safe, need a lot of care and can result in disastrous results like keratitis and vision threatening complications if care is not taken properly.





As mentioned above, vision acuity assessment is essential for detection of such uniocular refractive errors and goes a long way to prevent amblyopia.

#### Figure uniocular blindness its importance

Those children who have refractive errors should have 6 monthly vision tests particularly during the growth years. Complete fundus examination with peripheral Ophthalmoscopy should be done to rule out any retinal degenerations at an early stage in high myopes and moderate myopes.

## Surgery

Refractive procedures such as laser surgeries are not indicated in children since the pediatric eye is still in a growing phase. As such no surgery is recommended for correcting the refractive errors in children.

#### STRABISMUS AND AMBLYOPIA

This is commonly called asquint or crossed eyes. It is a condition in which the eyes are misaligned and point in different directions. This misalignment may always be noticeable, or it may come or go. Strabismus is a common condition among children affecting about 4% but can occur later in life.

Children do not outgrow strabismus. Treatment for strabismus is most effective when the child is young. Straightening of the eyes remains possible at any age and may result in improved side vision.

#### a. Types

The exact cause of the eye misalignment that leads to strabismus is not fully understood. Six eye muscle controlling eye movements are attached to the outside of each eye.

The two most common types of strabismus are esotropia and exotropia.

## Esotropia (Fig. 13)

Esotropia describes an inward turning of the eye and is most common type of

strabismus in infants. Young children with esotropia do not use their eyes together. Accommodative esotropia is a common form of esotropia, which occurs, in farsighted children. When child is young, they can focus their eyes to adjust for farsightedness but the focusing effort (accommodation) required to see clearly stimulates the eyes to cross because of the induced convergence



## Exotropia (Fig. 14)

Exotropia, or an outward turning eye is another common type of strabismus. This occur most often when a child is focusing on distant objects.



#### b. Amblyopia

With normal binocular vision (two eyed) vision, both eyes are aimed at the same target. The visual portion of the brain fuses the two pictures into a single 3 D image. When one eye turns, as in strabismus, two different pictures are sent to the brain. In a young child, the brain learns to ignore and the image of the misaligned eye and sees only the image from the straight or best seeing eye.

This causes loss of depth perception and binocular vision and leads to a condition called lazy eye or Amblyopia. Amblyopia can also develop because of the differences in the refractive state of the eyeball. Since the image of one eye is unclear, brain starts suppressing that eye's image and gradually it leads to even permanent uniocular loss of vision.

#### Management

#### Strabismus

Treatment goals for strabismus are to preserve the vision, to straighten the eyes and to restore binocular vision. Depending upon the cause treatment may involve repositioning of eye, removing a cataract, or correcting other conditions which are causing the eyes to turn.

In accommodative esotropia glasses reduce the focusing effort and can straighten the eyes. Sometimes bifocal are necessary for close work. Eye drops, ointment or special lenses called prisms can also be used to straighten the eyes. All the patients with strabismus should be checked for glasses and the required power should be prescribed.

Strabismus surgery involves making a small incision in the tissue covering the eye which allows the ophthalmologist access to the underlying muscles. Which eye muscles are to repositioned depends upon the direction the eye is turning.

## Amblyopia

Amblyopia can be treated by patching the preferred eye or better seeing eye to strengthen and improve vision in the weaker eye. If amblyopia is detected in the first few years of life, treatment is often successful. As a rule the earlier the Amblyopia is treated, the better the visual result.

#### CONGENITAL ANOMALIES OF THE GLOBE

Congenital anomalies include all forms of developmental defects present at birth, whether caused by genetic chromosomal or environmental etiologies. The etiologies include monogenic causes, chromosomal anomalies, multifactorial disorders, environmental agents and unidentified causes. Unfortunately, the latter category, unknown etiology, accounts for 50% or more of these malformations. Regardless of etiology, from a developmental point of view congenital anomalies may be organized into the following categories.

## Types:

- Agenesis: developmental failure (anophthalmos)
- *Hypoplasia:* developmental arrest (eyelid cooboma)
- Hyperplasia: developmental excess (distichiasis)
- Abnormal development (cryptohpthalmos)
- Failure to divide or canalize (congenital naso lacrimal duct obstruction)
- **Dysraphia** (choroidal coloboma)
- **Persistence of vestigial structures.** (Persistent pupillary membrane, remnant hyaloid artery)

A malformation implies a morphological defect present from the onset of development or every early stage.

A disturbance to a group of cells in single developmental field may cause multiple malformations. Multiple causations may result in similar field defects and patterns of malformation.



Fig.15.Showing ongenitally small eyes

#### Genetic counseling:

Genetic counseling is meant to tell the patient about the risk of successive sibling being affected by the disorder by which either one of the parent or the first sibling is affected.

First of all let us see what is are the different modes of inheritance of diseases. It could be either sex linked or somatic chromosomes. If it is y chromosomes it will be present only in male child. If it is X linked dominant it will be there in all the children, but if recessive then only can be there in females.

There are two alleles for any gene. Dominant indicates that even if only one of the two is present, it will be expressed. Recessive indicates that until two of them are present it cannot express itself. Therefore, it requires both the alleles to express genotype.

It is here that the problem of consanguineous marriage comes into play. Since similar mutations are there in the family members, even the recessive disorders may get expressed. These autosomal recessive disorders can be brought under control if one can educate the public about the potential dangers of the consanguineous marriages.

#### Tumors

#### Retinoblastoma

It is the most common primary malignant intraocular childhood tumor. It arises from the primitive retinal cells. Its incidence is 1 in 20,000 births. There I no sexual no racial variation.

<u>Genetics.</u> Retinoblastoma can be divided into hereditary and non hereditary. The gene responsible is a tumor suppressor gene and is located on chromosome 13. <u>Genetic counseling.</u> Risk of retinoblastoma depends on presence or absence of family history and whether the tumor is unilateral or bilateral.

Table08. Showing chance of following people to have a baby with	۱
retinoblastoma	

History	Parent	Affected child	Normal sibling
Family history	40%	40%	7%
No family			
history	6%	40%	1%
Bilateral	1%	8%	1%
Unilateral			

## Management.

#### Aims

1<sup>st:</sup> to save life.

2<sup>nd</sup> to save eye

3<sup>rd</sup> to maximize the vision

#### Treatment methods

Enucleation: removal of the eyeball as a whole with the tumor.

External beam radio therapy: using radioactive isotopes to shrink the sizeof the tumor

Chemotherapy: using anti cancer drugs to shrink the size of the tumor Focal therapy: implanting radioactive isotopes near the tumor. Orbital exentration: removing whole of the eye and part orbit for tumors extended out of the eye

#### Follow-up:

Patients with retinoblastoma and sibling s at risk need to be followed indefinitely Second cancers are the leading cause of death in patients with the hereditary type of retinoblastoma. Osteogenic sarcoma is the most common cancer. Others can be bony and soft tissue sarcomas and skin tumors.

Other tumors Benign Dermoids Hemangiomas and lymphangiomas Malignant Lymphomas

#### PAEDIATRIC GLAUCOMA

Glaucoma is a condition where optic nerve is progressive damaged due to various causes; increased intraocular pressure is one of the major reasons for that. It is mainly a disease of aged but can occur in pediatric patients. 1-10% of childhood blindness i.e. is nearly 2-20 children/ million population are blind due to developmental glaucoma. Its incidence is 1/ 10,000 live births. It has been classified on the basis of age of occurrence into

- Congenital glaucoma (up to 1 year of age)
- Infantile glaucoma (between 1-3 years of age)
- Juvenile glaucoma (between 3- 16 years of age)

#### Main features:

In *congenital glaucoma* due to raised intraocular pressure, in addition to optic nerve damage, the globe enlarges because to sclera in the eye of the baby is distensible. The corneas and sclera of these babies are, therefore, large. The layers of the cornea which are not elastic gets torn and may result in opacification.

Diagnostic features are tearing, photophobia (avoidance of light) and enlarged eyes.

The classical features of congenital glaucoma are not present in *infantile glaucoma and juvenile glaucomas*. The eyes are not enlarged and symptoms like watering ad corneal enlargement are lacking. This type of glaucoma is occasionally diagnosed during routine screening or when the child is examined because of a family history of glaucoma.

#### Management:

The treatment is mainly surgical. Till the surgery is done intraocular pressure reducing drops can be given. Surgery done can be either trabeculotomy or trabeculectomy. Trabeculotomy consists of opening up the trabecular meshwork (the space meant for aqueous humor) and in trabeculectomy, an artificial opening is created in the eye from where aqueous can filter out.

The surgery is done to reduce the intraocular pressure and to prevent the further visual loss. In early cases the visual loss may be reversible but in late cases the loss is irreversible. Nevertheless, amblyopia treatment and low vision rehabilitation must be tried in all the cases

#### LOW VISION SERVICES

A person with low vision is one who has impairment of visual functioning even after treatment and /or standard refractive correction, and has a visual acuity of less than 6/18 to light perception, or visual field of less than 10<sup>0</sup> from the point of fixation, but who uses, or is potentially able to use, vision for the planning an/ or execution of a task. The clinical evaluation for pediatric patients for rehabilitation purposes differs markedly from the routine eye examination. Ophthalmologists are accustomed to measure how

much vision is lost. In rehabilitation, the goal is to measure the child's abilities and needs. Residual vision is used to advantage, permitting the children to develop optimally.

## Objectives of low vision care.

- To minimize the extent to which low vision restricts children's participation in activities normal for children of that age in the community in which they live.
- Tests appropriate to the child's age and culture should be available in order to assess vision.
- Distance, near and functional vision should be assessed in addition to assessing the impact of visual impairment on the daily activities of the child.





Figures .16 Showing a child using low vision aid

## Clinical assessment:

Does the child has low vision?

When vision is assessed, children are presented with different stimuli and typical behavioral responses are anticipated. If apparently normal children do not repond in an age appropriate manner, abnormal visual function is suspected.

Testing material should be age and ability appropriate. Vision should be checked according to the mental IQ in mentally retarded child and not corresponding to his age.

What is the child's functional vision?

Functional vision is usable vision. Peripheral vision enables children to navigate through a classroom or playground.

#### Various low vision devices available are,

- Hand and stand magnifiers.
- High plus spectacles
- Telescopes
- Non optical devices
  - Reading stands
  - Special lights
  - Large lined papers
  - o Broad tip pens

In children the low vision aids of any kind has to reach them at an earlier age before amblyopia sets in. once the child gets adapted to Braille system and non-visual senses to help him in day to day activities, even a significant improvement of vision by using LVA may fail to motivate the child to use and benefit from the devices.

#### FREQUENTLY ASKED QUESTIONS

#### When should a child have his/her first eye exam?

Your child's future success can depend on being able to see well. While most children have good vision, any vision problem needs to be caught early. Rarely will a child complain about his/her vision. Young children think that everyone sees the way they do. We recommend seeing children as early as 6 months, then again at 3 years and 5 years. With early testing,

we can check for "lazy eye", focusing or depth perception problems. Here are some early warning signs of possible vision problems:

- Squinting, blinking, and rubbing the eyes.
- Sitting too close to the television or wanting to sit close to the "big screen" in the movie theater.
- Covering one eye or tilting the head sideways.

Poor eye-hand coordination and clumsiness

# **Refractive Error**

# 1. What is the power of glasses? Is it high or is it normal to have such a power?

We can tell the power of the glasses directly and tell them something related to that. We can easily pacify the patient, if the power is not very high, that power is normal for his age and many people have a small number but that the regular check up should be done and the glasses should be worn regularly (if required)

If at all the power is high, one can tell about the high power and the regular wear of glasses required with that and the routine check up required and the other retinal complications of high myopia, importance of regular check up

## 2. Can the power of glasses wear off on is on?

One has to be practical and tell the parents the power may have fluctuations and may reduced, very occasionally it may wear off on its

own, if it at all it wears off. The power may change and so regular check up

## 3. Can any drug help in removing he power of glasses?

Strictly he should be told that there is no drug available. Also that he should not waste money on the quacks that say that they can reduce the power of the glasses with the drugs. He should be told that the drugs may harm the child and may affect his psyche adversely.

## 4. Is surgery an option at this stage at this age?

The surgery at that age that is below years is not a good option rather not a at all because the child and eye are in the growing phase and that the power of eye may fluctuate during this period and depending on the power it may further increase or decrease or may change the type, if it is cylindrical.

## 5. How long will it take to get the power of the glasses to be stable?

Normally by the age of 18-21 years the power becomes stable but in few exceptional cases the power increase may b there for a longer time that is even after 21 years of age. Nothing actually can be done for that except that patient should be on the regular followup and should wear the appropriate correction.

## Squint

## 1. What Causes Strabismus?

Strabismus may be caused by unequal pulling of muscles on one side of the eye or a paralysis of the ocular muscles.Occasionally, when a farsighted child tries to focus to compensate for the farsightedness, he or she will develop accommodative strabismus. This condition usually appears before two years of age, and can occur as late as six.

## 2. What is myopia?

Myopia is our fancy, scientific name for nearsightedness. Nearsightedness is when a person sees better at near than at far — in other words, faraway objects are blurry. This can easily be accomplished with glasses or contact lenses.

## 3. Which eye is having squint? Are both the eyes affected?

Squint is basically the misalignment of the two eyes so it is actually the relative concept that is when one eye is straight the other is squinting. Squint eye if unilateral should be considered as lazy or weak in comparison to the other eye and so various exercise are given to increase the power of the weak eye. Also if during the treatment both the eyes start squinting then it is a good sign as both eyes have equal power and so if required surgery may be tried.

## 4. What is the need of patching?

Patching is needed basically to keep the power away from suppressing the weaker one that is the weak eye. If patient is having high powered glasses in one eye or squint of a particularly eye only it is suggestive that that particular eye is amblyopic or weak in comparison to the other eye. Therefore, we patch the worse eye to give an equally good chance of gaining the same vision or the power.

## 5. Can squint be corrected with patching?

In few cases, it can be t most of the cases will require surgery if there is squint. Patching is the processes to prepare the patient for surgery. It is basically necessary if there is amblyopia or weak eye.

## 6. When will be surgery done?

Surgery depends on many factors, the amount of deviation, the vision in the two eyes, the frequency of deviation during the day and control of deviation that is stability of the angle. Surgery will be done as soon as possible but the most important aspect is the visual aspect and not the deviation as that can be corrected at any time but the vision if not stimulated at an early age then at a later age it is difficult to regain.

## 7. Will the eyes remain stable after that? If yes how many years?

Stability of eyes depends on several factors. First of all the power of glasses, again the preoperative deviation the amount of deviation whether it can be corrected in a single surgery or not. Also postoperatively exercises may be given to maintain the alignment. It is possible that patching may also be started or rather continued postoperatively also depending upon the findings postoperatively. 100% is not possible but the alignment done will be cosmetically acceptable that can be promised depending upon the angle of deviation preoperatively.

## 8. Till how long patching is to be continued?

Patching is to give equal chance to the weak eye to have better vision so as soon as the weak eye has better vision almost equal to the better eye then patching can be reduced to maintain the vision which ahs been regained in the worse eye through patching.

## **Buphthalmos**

## 1. Will surgery improve the vision?

The surgery is basically been done to prevent the further growth of eyeball and not just for the vision. In paediatric patients there is some chance of regaining of the visual field and the vision if operated sufficiently early.

Also the surgery is being to relieve the pain and reduce the intraocular pressure, this of course cant reverse what damage has been done but can definitely prevent the further damage.

## 2. Will the size of eye decrease or not?

The size of the eyeball once it has increase never goes down. It is good that it will not increase further in surgery and also that the clarity of the cornea and the symptoms like watering and photophobia will improve.

## **General questions**

## 1. How long the surgery will take?

Depending upon the procedure it can take anywhere between 25 minutes in a patient of cataract to 45 minutes in a squint surgery with 2 muscles. This includes the time of induction and bringing out the baby.

#### 2. What are the risks of anaesthesia?

That depends from the patient to the patient a healthy person has almost negligible risk of anaesthesia but as they say that there is at least grade I anaesthesia in all the patients even when everything is fine. If there is some systemic problem that is chest problem, breathing, some infection, low Hb etc. the benefit/ risk ratio should be high.

#### 3. Can my child see television? Does television affect the vision?

Yes he can see television daily if it is not a problem for any other work that his studies etc. from the ophthalmological point of view television is as safe as anything. No study has as yet proven that television cause any adverse effect on the eyes. TV never affects the vision of the patient but yes if seen for a longer time reports of redness of eye and burning pain has been reported by the people, this is due to the dry eye which occurs as a result of decrease in the blinking rate of the patients.

# 4. Does the distance of television and patient have anything to do with the change of vision?

Distance from the TV explains the vision of the patient that is poor distant vision patient will try to have a better image and so will try to see the TV from less distance. The amount of glare increases as you go near to the Video display terminals. Patient should be checked for any refractive error if he sees TV from near. 5. What to be given preoperatively? How long he should be kept hungry preoperatively?

Preoperatively antibiotic drops only to be applied on the eye.At least 8hrs fasting before operation.

6. What is dose of vitamin A? Importance of Vitamin A? How frequently should it be given?

Dose of Vitamin 'A' – 2lac I.U from 6 months of age up to 6 years at 6 months interval

# 7. What is the chance of next child to have squint, retinoblastoma, glasses?

If one child have squint or glasses ,there is chance of squint and refractive error of subsequent child also.

Unaffected parents with one affected child have an approximately 5% risk of producing another affected child.

If two or more siblings are affected the risk of subsequent children being affected is 50%

A survivor of hereditary Retinoblastoma has a chance of almost 50% that offspring will also develop the tumour.

# Noncanalisation of Nasolacrimal duct

 When will the watering stop? How long the massage is necessary and how long the antibiotic drops will continue? With proper lacrimal sac massage and antibiotic drops watering could be stopped within 1 year of age. 2. Can the patient with congenital dacryocystitis be operated, if yes what is the minimum age for that?

Yes, Congenital dacryocystitis can be operated at 5years of age

## **Congenital Ptosis:**

1. Will the amount of ptosis improve on its own?

No amount of ptosis will not improve on its own.

## 2. What is right age for surgery?

At any age congenital ptosis can be corrected, it depends upon the amount of ptosis.

## 3. Will it affect his visual acuity?

If ptosis cover the pupillary area that only it affects vision

## Vernal conjunctivitis.

What is the cause for recurrent redness and watering?
 Due to allergic reaction recurrent redness and watering

## 2. Can it affect his vision?

If cornea involved e.g: SPK"s, Keratoconus or complication of drugs affect lens e.g: cataract than vision can be affected.

## 3. Any surgery will it be helpful for this?

No surgery will be helpful for vernal conjunctivitis

## 4. Long term side effects of the drops if any?

Specially topical steroid can cause -

- 1. secondary Glaucoma
- 2. Cataract

## 5. How long the drops will have to be continued?

- 1. Topical steroids or anti-inflammatory should be continued for few weeks
- 2. Mast cell stabiliser drops should be continued for few months

## 6. What is the treatment of childhood cataract?

Operation is the only treatment

## 7. Whether it is possible to implant IOL?

Usually children more than 2 years of age it is possible to implant IOL. Age less than 2 years it is not possible to implant IOL

## 8. Whether the baby have to wear glasses after operation?

If IOL implantation done than the baby have to use glasses for near work. If IOL implantation not done than the abby have to wear thick glasses

# 9. How long the baby have to wear glasses? Life long

## 10. How long IOL work properly?

Once IOL implanted It will work for life long of the baby.

## 11. What is microvornea?

Microcornea is a cornea with a smaller than average diameter (less than 11mm).

## 12. Is there a remedy for a stye?

Best remedy for a stye is a warm wet compress

## 13. What is nystagmus?

Nystagmus, which is typically a back and forth jerkiness of the eyes that hinders clarity of vision and one's ability to focus. Nystagmus is neurologically based, and to the best of my knowledge, there are no effective treatments for it.

## 14. What Causes Amblyopia?

Trauma to the eye at any age can cause amblyopia, as well as a strong uncorrected refractive error (near sightedness or farsightedness)or strabismus. It's important to correct amblyopia as early as possible, before the brain learns to entirely ignore vision in the affected eye.

#### 15. What exactly is glaucoma?

Glaucoma is a condition in which the eye's intraoculars(IOP) is too high. This means that your eye has too much aqueous humour either because it produced too much, or because it's not draining properly. Other symptoms are optic nerve and vision loss.

## 16. Is there any way to prevent glaucoma?

Doctors don't know of any way to prevent glaucoma.

## 17. Is glaucoma curable?

No, but it's important to treat glaucoma, because it can cause blindness.

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