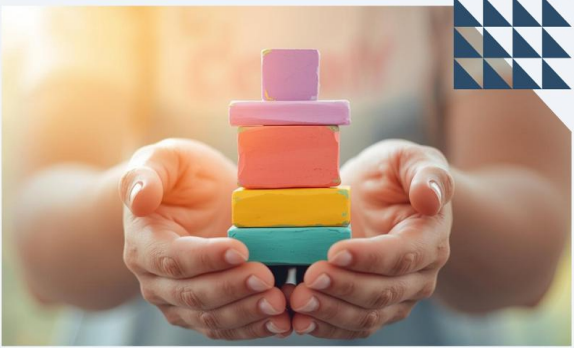
Building a **solid foundation** for pediatric eye care



Pediatric Ocular Diseases Guidelines

APOP & OSP Lahore

GUIDELINES

Congenital Cataract
Retinoblastoma
Refractive Errors
Retinopathy of Prematurity
Congenital Glaucoma

Mission Statement

To unify pediatric ophthalmology practice in Pakistan through shared protocols, collaborative learning, and strong professional networks.

To elevate clinical standards so that every child benefits from dependable and high-quality care.

Vision

To build a nationally aligned pediatric eye-care framework that sets benchmarks for excellence.

To inspire a generation of clinicians dedicated to protecting and advancing children's vision.

Foreword

These National Pediatric Ophthalmology Guidelines reflect a unified national effort to standardize the diagnosis, management, and long-term care of children with Retinoblastoma, Congenital Glaucoma, Retinopathy of Prematurity, Congenital Cataract, and Childhood Refractive Errors in Pakistan. Developed through the collective expertise of dedicated clinicians, they aim to strengthen evidenee based practice and reduce preventable childhood blindness across the country.

We gratefully acknowledge the Editorial Board—**Prof Seema Qayyum, Prof M. Ali Ayaz Sadiq, and Prof Sorath Noorani—for leading this initiative.**

I thank the contributors:

Dr Insha e Qudrat, Dr Jawaria Tariq, Dr Muhammad Shahzeb Aslam, Dr Irfan Ali for **PEDIATRIC CATARACT GUIDELINES.**

Dr Saima Amin, Dr Sidra Jabeen, Dr Nazli Gul, Dr Asma Mushtaq, Dr Samreen Jamal, Dr Khawaja M Baqir Hassan for **RETINOBLASTOMA GUIDELINES**.

Dr. Maria Memon, Dr. Aisha Azam, and Dr. Asma Mushtaq for **REFRACTIVE ERRORS GUIDELINES**.

Dr Fiza Azhar, Dr Noor ul Ain, Dr Laiba Asif, Dr Rehma Shanze, Dr Amna Mehmood, Dr Hira Awais, Dr Sarfaraz Hussain Syed, Dr Andleeb and Dr Munir Shakir for **CONGENITAL GLAUCOMA GUIDELINES**.

Dr Khurram Mirza, Dr Sidra Jabeen, Dr Nayab Batin Farooqi for **GUIDELINES ON RETINOPATHY OF PREMATURIY.**

Their efforts embody our shared vision of reducing preventable blindness and saving young lives through knowledge, cooperation, and compassion.

It is my hope that this document will serve as a living resource—guiding clinicians, informing policy, and inspiring continued research and advocacy for the wellbeing of children across Pakistan.

Prof Dr Ch Javed Iqbal
President, Ophthalmological Society of Pakistan

Editorial Note

The National Guidelines for the Management of Pediatric Eye Diseases in Pakistan provide evidence-based, standardized protocols for diagnosing, treating, and monitoring common and critical pediatric ocular conditions. Developed through collaboration among ophthalmologists, pediatricians, and pediatric oncologists, the guidelines aim to reduce preventable childhood blindness and ensure equitable access to quality eye care across the country.

Key conditions addressed include:

- Pediatric Cataract: Standardized evaluation, surgical management, and postoperative follow-up.
- Retinoblastoma: Prompt referral pathways, multidisciplinary management strategies, and genetic counseling.
- Refractive Errors: What to prescribe
- Congenital Glaucoma: Early detection, timely surgical intervention, and lifelong follow-up.
- Retinopathy of Prematurity (ROP): Screening criteria, treatment options, and follow-up schedules adapted to local neonatal care practices.

These guidelines are intended as living documents, to be periodically updated as new evidence and local research emerge. Healthcare professionals are encouraged to adopt them in practice and contribute feedback to strengthen future editions.

Their implementation aims to reduce childhood blindness, improve clinical outcomes, and ensure that every child in Pakistan has the opportunity to see and thrive.

Prof. Dr. Seema Qayyum
Prof. Dr. Mohammad Ali A Sadiq
Prof. Dr. Sorath Noorani

Contents

PAEDIATRIC CATARACT

SCREENING AND MANAGEMENT GUIDELINES

CONTRIBUTING AUTHORS

- Dr. Insha e Qudrat
- Dr. Jawaria Tariq
- Dr. Muhammad Shahzeb Aslam
- Dr. Irfan Ali


In clearing the Cloud
of congenital cataract.
we return a child's
future to light.
Restoring clarity
renewing hope.

Chapter 1

PAEDIATRIC CATARACT

1. Background

An estimated 200,000 children worldwide are blind due to cataract. That amounts to 5-20% of Pediatric blindness worldwide. 20,000 to 40,000 children are born each year with congenital cataract. Overall prevalence of childhood and congenital cataract is inthe range of 6 per 10,000 live births. It presents an enormous problem to developing countries in terms of human morbidity, economic loss and social burden 3.

Although screening and management guidelines for Pediatric cataracts exist in the developing world, there are no uniform guidelines developed in Pakistan. A lot of variability is seen in the management of this entity within individuals as well as centers.

Need of pediatric cataract arises from the fact that:

- Blindness from pediatric cataract is avoidable
- Timely and early detection prevents undesirable sequale and progression
- There are presently no screening and management guidelines available in Pakistan.

Management of pediatric cataract is a classic example of a situation that requires multidisciplinary cooperation and coordination.

1. SCREENING PROTOCOLS

This will be done by Pediatrician/Neonatologist/Lady Health Workers. Distant Direct Ophthalmoscopy will be done as screening.

- **Birth**
- **1 week**
- **3 week**
- **6 week**

1.1 Pre-school vision screening will be done in the community at:

- **3 years**
- **5 years**

2.2 Comprehensive Eye Examination

Comprehensive eye exam will be done by an ophthalmologist at:

- **3 months**
- **1 year**

2. REFERRAL CRITERIA

Urgent referral to an Ophthalmologist is to be made if:

- **Presence of any opacity on Distant Direct Ophthalmoscopy**
- **Absence of Red Reflex**
- **Leukocoria**
- **Strabismus**

3. COMPREHENSIVE EYE EXAMINATION

Comprehensive eye exam will include

Detailed History

- Child's milestones
- Pedigree Tree
- Health problems in siblings and parents
- Cousin Marriage
- Family History
- Gestational Hx (infections, exposure to toxins, Radiations , Drugs)

Visual Acuity Testing

- **Pre-verbal Children**
  - Forced preferential looking techniques (Teller acuity cards, Cardiff card, Kecler acuity cards).
  - Fixation and following evaluation, and assessing objection to occlusion of each eye.

- **Verbal Children (3 years and above)**
  - HOTV matching.
  - LEA symbols.
  - Tumbling Es.
  - Matching game or identification of symbols and letters.

Intra-ocular pressure
Fundal Exam
Ocular Ultrasound (B-scan) if no fundal view

4. LABORATORY TESTING

- **: No Laboratory Testing required**
  - Unilateral Cataract.
  - Bilateral cataract with no co-morbidities.
  - Bilateral cataract with positive Family History.

Laboratory Testing required

- **Bilateral cataract**
  - Urine test for reducing sugar.
  - TORCH Testing.
  - VDRL testing.
  - Serum Calcium, Phosphorus, Glucose, Galactokinase.

5. WHAT TO OPERATE

Pre-Verbal Children

- Dense central opacity > 3 mm.
- Abnormal visual behavior.
- Aversion to covering the good eye.
- Hazy view of the macula on ophthalmoscopy.
- Hazy view of the macula on ophthalmoscopy.

Verbal Children

- Opacity causing a decrease in quality of life.
- Visual Acuity of 20/40 or worse 6/9.
- Visual acuity drops by two lines on glare testing.

6. WHEN TO OPERATE

Unilateral Cataracts

- Before 6 weeks.
- As early as possible if detected after 6 weeks.

Bilateral Cataracts

- Before 6 weeks for first eye.
  - Before 8 weeks for the other eye.

NOTE: Birch and Stager found no difference in visual outcomes of the surgery if the surgery is performed between birth and age 6 weeks).

7. WHEN TO IMPLANT THE IOL

IOL implantation should not be done before 12 months.

7.1 Biometry

Immersion biometry method.

7.2 Target Hypermetropia

Bilateral Cataract

- Target Hypermetropia = 6 - (Age of the patient)

Age at cataract surgery and residual refraction recommendations for target refraction

Age at cataract surgery	Residual refraction (Diopters)
<6 months	+6 to +10
6-12 months	+4 to +6
1 -3 years	+4
3-4	+3
4-6	+2 to +3
6-8	+1 to +2
>8	+1 to 0

Unilateral Cataract

- Target Hypermetropia = Hypermetropia in the normal eye.

7.3 Which IOL to Use

Single piece Acrylic hydrophobic in bag.

Multi piece Acrylic hydrophobic in sulcus.

PMMA can also be used.

8. SURGICAL TECHNIQUE

An ideal capsulotomy for an IOL with optic diameter 5.5-6 mm should be 4-5 mm.

Anterior capsular polishing should be done after cataract aspiration.

Primary posterior capsulotomy with anterior vitrectomy.

- age of 5 years in normal kids.
- age of 8 years for mentally challenged patients or those having nystagmus.

Wound closure

- Main port and port must be sutured with 10/0 nylon.

9 POST-OPERATIVE MANAGEMENT

9.1 Post-Operative Medication

Topical steroids one hourly for one week and then taper after documenting quiet anterior chamber on Slit lamp.

Topical antibiotics 2 hourly for one week.

Mydriatics for one week.

9.2 Post-Operative Examination Protocols

Target at each visit

Wound integrity.

Signs of inflammation.

Astigmatism.

centration of IOL if pseudophakic.

IOP.

Red reflex.

CD ratio.

Refraction.

9.3 Post-Operative Visual Rehabilitation

Amblyopia therapy and optical correction following cataract surgery bifocal segment is needed in children 4 years of age or older.

9.4 Post-Op Follow Up (Visual Rehabilitation, Glaucoma Monitoring)

6 weekly visits till visual rehabilitation is complete.

Then graduating to 3 monthly visits and then 6 monthly.

Aphakic contact lens for unilateral cataracts who have been left aphakic.

Bifocals to be incorporated for patients aged 3-4 years or older.

Glaucoma monitoring by:

IOP.

Axial length.

Increasing myopic shift in refraction.

CD ratio.

ROP.

Tele medicine.

Book for surgery based on unilateral cataracts

EQUIPMENT REQUIRED

S #	Name of Additional Equipment for Paeds OT
1	Teaching Microscope
2	Phaco machine
3	Anterior viterctomy
4	Retinoscope
5	Ophthalmoscope
6	Hand held slit lamp
7	Trial lenses
8	Tonopen
9	Indirect Ophthalmoscope
10	20/28D lens
11	Flourecein strips
12	Hand held A scan
13	General Anesthesia Machine with Adult & Pediatric Circuit and accessories
14	Auto refractor meter
15	Harms Trabeculectomy Set
16	Set of Refraction bars (Lens Bar)
17	Thorpe Surgical Ginio Lens, Model: OTSG

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RETINOBLASTOMA

SCREENING AND MANAGEMENT GUIDELINES

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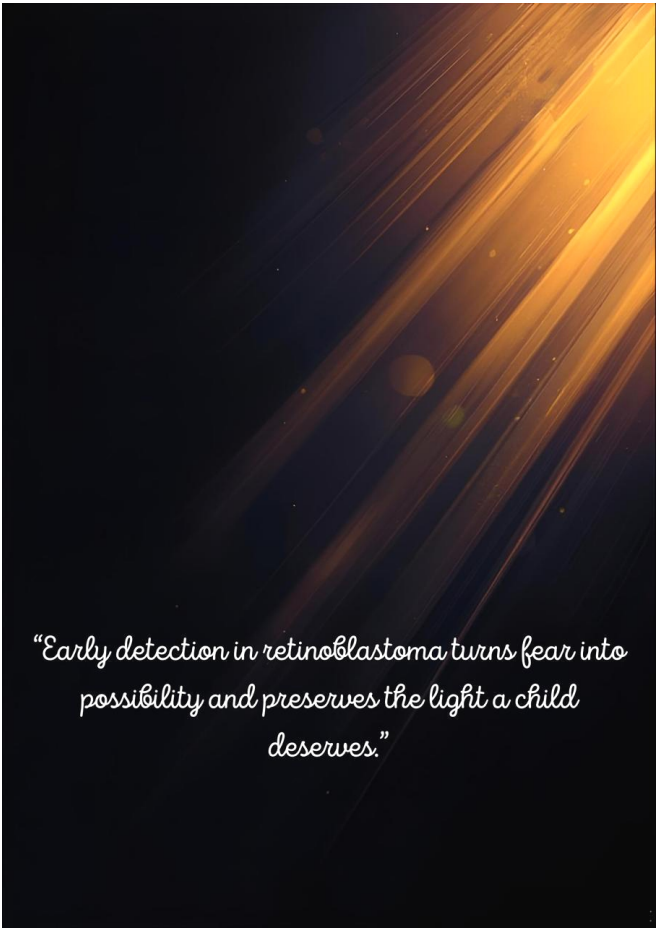

"Early detection in retinoblastoma turns fear into possibility and preserves the light a child deserves."

Chapter 2

RETINOBLASTOMA

1. INTRODUCTION

Background:

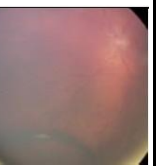
Retinoblastoma (RB) is the most common ocular malignancy in children. High cure rates of close to 100% have been reported in developed countries. However, two thirds of all cases are diagnosed in developing countries where survival rate range from 40 to 60% due to advanced stage at presentation.[1,2]

This aggressive neoplasm originates in the retina and primarily affects children younger than 5 years. With an incidence of 1 in 15,000 to 20,000 live births, retinoblastoma remains a significant contributor to global childhood cancer mortality.

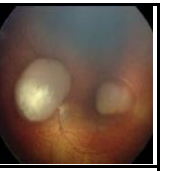
Advances in early diagnosis, imaging, and treatment have improved survival rates, particularly in high-resource settings. However, disparities persist due to delays in diagnosis, limited access to specialized care, and socioeconomic factors.[1]

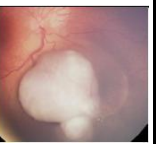
Management has evolved significantly, with a growing emphasis on organ preservation, vision salvage, and minimizing long-term treatment complications. Treatment decisions depend on the extent of the disease, the patient's age, and access to specialized resources.[3]

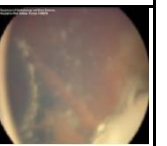
CLASSIFICATION:

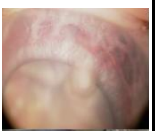
International Intraocular Rb Classification (IIRC)	
Group A: Very low risk	
Small discrete tumours not threatening vision (T1a)	
All tumours are 3mm or smaller confined to the retina	
Located at least 3mm from the foveola and 1.5mm from the optic nerve	
No vitreous or subretinal seeding	

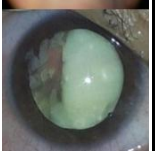
Group B: Low risk
No vitreous or subretinal seeding (T1b)
Tumours any size or location not in group A
No vitreous or subretinal seeding
Subretinal fluid no more than 5mm from tumour base
Group C: Moderate risk (T2)
Focal vitreous or subretinal seeding and discrete retinal tumours of any size and location
Local, fine and limited seeding (T3)
Discrete intraretinal tumours of any size and location (T2b)
Up to one quadrant of subretinal fluid (T2a)
Group D: High risk
Diffuse vitreous or subretinal seeding (T3b)
Diffuse intraocular disseminated disease
Extensive or “greasy” vitreous seeding
Subretinal seeding may be plaque like
More than one quadrant retinal detachment
Group E: Very high risk (T4a)
Very high risk with one or more of the following:
Irreversible neovascular glaucoma
Massive intraocular hemorrhage
Aseptic orbital cellulitis
Tumour anterior to anterior vitreous base
Tumour touching the lens
Diffuse infiltrating RB
Phthisis or pre-phthisis











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STAGING GUIDELINES

Stage 0: Patients treated conservatively.

Stage 1: Eye enucleated, completely resected histologically.

Stage II: Eye enucleated, microscopic residual tumor.

Stage III: Regional extension.

a. Overt orbital disease.

b. Preauricular or cervical lymph node extension.

Stage IV: Metastatic disease.

a. Hematogenous
Metastasis
Disease
(without central nervous system [CNS] involvement).
1. Single lesion.
2. Multiple lesions.

b. CNS extension (with or without any other site of regional or metastatic disease).
1. Pre-chiasmatic lesion.
2. CNS mass.
3. Leptomeningeal and CSF disease.

1. SCREENING PROTOCOLS:

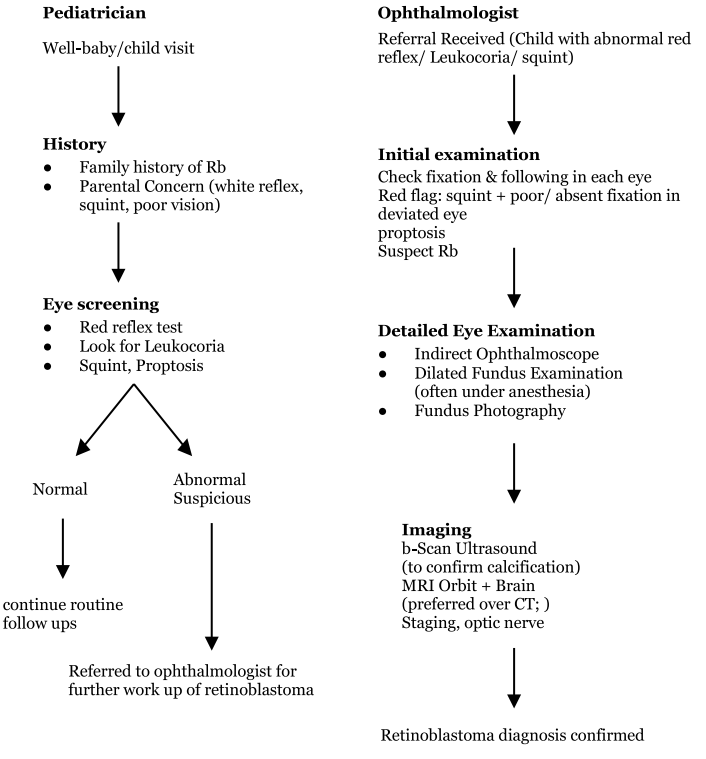
By Paediatrician

- First 2 years of life on every visit of every child by Distant Direct Ophthalmoscopy (Red Reflex)
- Corneal light reflex/ Hirscheberg.[v1]

Referral to ophthalmologist of any child with

- strabismus
- leukocoria
- abnormal red reflex
- family history of Retinoblastoma

Early detection of Retinoblastoma – flow chart for Pediatrician and Ophthalmologist



High Risk

- Parent with bilateral disease.

Intermediate Risk

- Parent with unilateral disease.
- Siblings with bilateral disease.
- Fellow eye of Child with multifocal unilateral.
- Parent with sibling with bilateral disease.

Low Risk

- Sibling with unilateral disease.
- Parent with sibling with unilateral disease.

Management Guidelines for Childhood Screening for Retinoblastoma Families

Risk Category	% risk	Eye examination schedule based upon age of unaffected child							
		Birth to 8 weeks*	> 8 to 12 weeks	> 3 to 12 months	> 12 to 24 months	> 24 to 36 months	> 36 to 48 months	> 48 to 60 months	5-7 years
High Risk	> 7.5	Every 2-4 weeks	Monthly		Every 2 months	Every 3 months	Every 4 months	Every 6 months	Every 6 months
Intermediate Risk	1 - 7.5	Monthly		Every 2 months	Every 3 months		Every 4-6 months		Every 6 months
Low Risk	< 1	Monthly		Every 3 months	Every 4 months	Every 6 months		Annually	
General Population	0.007	Screening with pediatrician							

Nonsedated office examination preferred by most centers

Examination under anesthesia preferred by most centers

Initial assessment

Suspected RB – refer to specialized regional center with referral form

How Suspected RB is confirmed by ophthalmologist

(by EUA & B.SCAN)

REFER to radiologist and paediatric oncologist for MRI, B/L Bone marrow biopsy , diagnostic CSF is indicated for group D,E or extraocular disease.

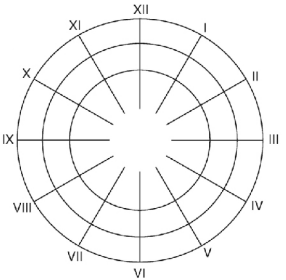
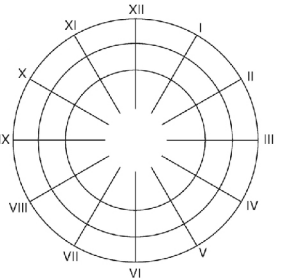
Multidisciplinary team (ophthalmologist, oncologist, radiologist) meeting to finalize stage and treatment plan

RETINOBLASTOMA REFERRAL FORM

Name		Surname			
DOB		Hospital No			
Address					
Mobile		Patient's NID			
Gender	Male	☐ Female			
Date First Seen		Date First Seen			
Presenting Complaint		HOPI:			
☐ Lecocoria/white reflex					
☐ Strabismus / squint					
☐ Orbital / prominent globe		Pedigree / Family tree			
☐ Other					
Socioeconomic History & Ethnicity					
Family H/O RB		Family H/O Cancer			
Past history					
Enucleation Right	☐	Left	☐		
Exenteration Right	☐	Left	☐		
Investigations					
B-Scan		RE		LE	
Calcifications		☐ Yes	☐ No	☐ Yes	☐ No
MRI/ CT Scan done (encircle)		☐ Yes		☐ No	
Referred To		Hospital			
		City		Contact No	
Department Refer To		Ophthalmology		Oncology	
Reference Letter given		☐ Yes		☐ No	
Contact to Rb team Done		Yes		No	
Referring Doctor		Referred from			
Contact of referred Physician (for correspondence)					

EUA REPORTING FORM

EUA Reporting Form		Date of examination
Name	DOB	
Hospital no		
Gender	☐ Male	☐ Female
B.SCAN (date and findings)		
Pre EUA assessment		
Visual Acuity	RE	LE
RAPD		
EUA	RE	LE
Orbital disease/ Conjunctiva/ Sclera		
Anterior Segment		
Horizontal Corneal Diameter		
IOP		
Anterior Chamber : Tumour seeds/Mets/Shallow/Hyphema/Hypopyon		
Iris- NVIs/Ectropion uvea		
Pupil- Irregular		
Lens- Cataract/Touch		
Vitreous- Endophytic/Exophytic/vitreous hmg		

Fundus		
		
1. No of tumours 2. Site of tumours 3. Size of tumours(dimensions) 4. Fovea spare 5. Intravitreal seeds 6. Subretinal seeds 7. Retinal fluid 8. Retinal detachment 9. Others: Diffuse infiltration of retina, orbital cellulitis like picture, phthisis		
International Intra-Ocular Classification (IIRC)	RE	LE
Comments/Plan:		

MANAGEMENT

The management of Rb depends on the group at the time of EUA and whether the disease is intraocular or extraocular.

FOCAL THERAPY GUIDELINES

1. **LASERS AND CRYOTHERAPY**

Indications

- Group A: posterior pole, away from fovea/optic disc.
- Group B: post chemoreduction
- Group C: Post chemoreduction
- Group D: post chemoreduction

Goal:

Eye salvage with minimal damage to the healthy retina and visual potential preservation.

A. Laser Photocoagulation- green laser ((Nd:YAG)

The 532nm wavelength is well absorbed by hemoglobin and melanin, making it effective for treating blood vessels within the tumor.

Indications:

- Posteriorly located tumors after chemoreduction except in GROUP A.
- Laser in normal peripheral retina posterior tumor can form a protective barrier to retinal detachment prior to cryotherapy.

Technique: Through transpupillary route, mainly treat tumor surface, 1–2 mm margins treated to prevent tumor dissemination.

Excessive power in a small spot size can lead to tumor disruption or hemorrhage, so careful titration of laser parameters is essential.

Frequency: Every 3–4 weeks; typically 2–3 sessions.

Settings:

- **Power:** Power settings usually range from 200 to 600 mW, but may be adjusted in 20mW increments during treatment.
- **Spot Size:** A spot size of 100-500 micrometer is commonly used.

- **Duration:** CONTINUOUS WAVE- time depending on the tumor size and response.
- **Endpoint**: The desired outcome is a loss of surface blood vessels, and pale appearance.

Group	Treatment	Follow up	Subsequent follow-up	Additional points
A	**Unilateral Disease**: Upfront laser (posterior located) (red/green) cryotherapy (anterior located tumor) **Bilateral Disease:** Upfront laser/cryo if fellow eye is undergoing upfront enucleation- if intravenous chemotherapy (IVC) being given for the fellow eye (either for extraocular disease or as intent to salvage) wait for 2 cycles of IVC then proceed with laser/cryo to minimize scarring	3 weekly follow-up – repeat cryo or laser if not completely regressed or if in doubt. Look for new lesions. Document and draw each lesion for activity and location.	Two sessions of 3 weekly follow-up recommended after the lesion has completely regressed and no new lesions seen. The duration of further follow-ups may be increased by 1 week on subsequent follow-ups	Risk of reactivation and development of new lesions are highest 6 weeks – 6months after cessation of intravenous chemotherapy. Laser treatment should include a barrage around the tumor and mainly focus on laser shots on the surface of the tumor. Tumor response includes blanching of the surface, and swelling of the surface of the tumor. Very high power can cause disruption of the tumor and subsequent tumor dissemination If mosaic or germ line disease known or suspected (bilateral disease, family history, multifocal
B	Laser/ cryo Post 02-04 cycles of IVC or 02-04 cycles of Intra-arterial chemotherapy(IAC) (depending on the tumor size and tumor regression to avoid dissemination) (diode laser is recommended for better penetration)	3 weekly follow-up – repeat cryo or laser until completely regressed or if in doubt. Look for new lesions. Document and draw each lesion for activity and location.	Two sessions of 3 weekly follow-up recommended after the lesion has completely regressed and no new lesions seen. The duration of further follow-ups may be increased by 1 week on subsequent follow-ups	

C	Laser/ cryo Post 02-04 cycles of IVC or 02-04 cycles of IAC (depending on the tumor size and tumor regression to avoid dissemination) (diode laser is recommended for better penetration) For resistant vitreous seeds (active after 4cycles of IVC) Plan intravitreal chemothrerapy) IVitC)(refer to IVitC section)	3 weekly follow-up – repeat cryo or laser until completely regressed or if in doubt. Look for new lesions. Document and draw each lesion for activity and location. For patients receiving **Intravitreal melphalan** followup 8-14 days for subsequent injection For patients receiving **Intravitreal Topotecan** subsequent followup will be after 21 days	Two sessions of 3 weekly follow-up recommended after the lesion has completely regressed and no new lesions seen. The duration of further follow-ups may be increased by 1 week on subsequent follow-ups For patients receiving IViC atleast 1 maintaince injection is required after complete regression provided the maximum number of injections have not exceeded or retinal toxicity has not developed	tumors in a unilateral disease, presenting age <1.5 years) keep child under maximum interval of 6 weeks follow-up until 3years of age. Always look out for signs of retinal toxicity in patients receiving IAC or IViC and tailor treatment accordingly. For unilateral group D disease always give patient an option of upfront enucleation if vision potential is poor and disc and macula are involved by the tumor
D	Laser/ cryo Post 02-04 cycles of IVC or 02-04 cycles of IAC (depending on the tumor size and tumor regression to avoid dissemination) (diode laser is recommended for better penetration) For resistant vitreous seeds (active after 4cycles of IVC) Plan intravitreal chemothrerapy) IVitC) (refer to IVitC section)	3 weekly follow-up – repeat cryo or laser until completely regressed or if in doubt. Look for new lesions. Document and draw each lesion for activity and location. For patients receiving **Intravitreal melphalan** followup 8-14 days for	Two sessions of 3 weekly follow-up recommended after the lesion has completely regressed and no new lesions seen. The duration of further follow-ups may be increased by 1 week on subsequent follow-ups For patients receiving IViC atleast 1 maintaince	

		subsequent injection For patients receiving **Intravitreal Topotecan** subsequent followup will be after 21 days for repeat dose	injection is required after complete regression provided the maximum number of injections have not exceeded or retinal toxicity has not developed	

B. Thermotherapy (TTT)

- **Indications**: Thicker endophytic tumors.
- **Technique**: Infrared diode laser (810 nm); use transpupillary route. Main area of treatment is tumor surface, 1-2mm margins of tumor is barraged to prevent tumor dissemination.
- **Sessions**: Multiple sessions every 3–4 weeks as required.
- **Frequency**: Every 3–4 weeks; typically 2–3 sessions.
- **Power**: Power settings usually range from 300 to 600 mW, but may be adjusted in 50mW increments during treatment.
- **Spot Size**: A spot size of 1.2 mm is commonly used.
- **Duration**: CONTINOUS WAVE-The laser is applied for a period of time, often 1-5 minutes, depending on the tumor size and response.
- **Endpoint**: The desired outcome is a gentle, light gray color change within the tumor, indicating a successful “take”.

RELATIVE CONTRAINDICATIONS OF LASER:

- Large tumor without chemoreduction particularly if located near the disc or fovea.
- Only eye tumor located around the margins of the disc or fovea →risk of vision loss.
- Tumor underlying exudative retinal detachment.

C. Cryotherapy

- **Indications**: Anterior or peripheral tumors , inferiorly settled vitreous seeding or vitreous seeding in close proximity to the periphery.

- **Technique**: Triple freeze thaw using cryoprobe to achieve an iceball covering the tumor.
- **Interval**: treatment is done at 3 weekly interval until complete regression achieved.

INTRAVITREAL CHEMOTHERAPY

NOTE: Intra vitreal chemotherapy should be administered by a trained and experienced pediatric ophthalmologist or retinal surgeon.

Indications

- Persistent vitreous seeds: When vitreous seeds remain after initial systemic or intra-arterial chemotherapy with atleast one quadrant free of vitreous seeds.
- Recurrent vitreous seeds: If vitreous seeds reappear after treatment has been completed.
- Chemo-resistant vitreous seeds: If new vitreous seeds develop that are not responding to other chemotherapy treatments.

Contraindications

- Seeds dispersed diffusely in the entire vitreous cavity.
- Anterior segment and/or ciliary body invasion.
- Secondary glaucoma.
- High bullous retinal detachment.
- Vitreous hemorrhage obscuring the fundus view.

Chemotherapy agents

- Melphalan hydrochloride.
- Topotecan.
- Combined Melphalan plus Topotecan.

Frequency:
Can be given every 2-4 week for 4-6 cycles.

6. INTRA ARTERIAL CHEMOTHERAPY

NOTE: Intra-arterial chemotherapy should be administered by a trained and experienced interventional radiologist.

There is increasing evidence that systemic chemotherapy is replaced by Intra-arterial chemotherapy.

This limits systemic side effects and allows improved eye salvage. Abramson et al, in a review of current treatment of RB recommend IAC for all unilateral group C and D eyes. They report 94% ocular salvage in this population[2]. Similarly, Sheilds et al have reported success of 100% in group C and D eyes and 33% in group E eyes.

INDICATIONS:

IAC should be offered to every child who has visual potential in intra-ocular RB irrespective of group. For vitreal seeding combine IAC with IVI Melphalan and the results are promising.[1,2,3]

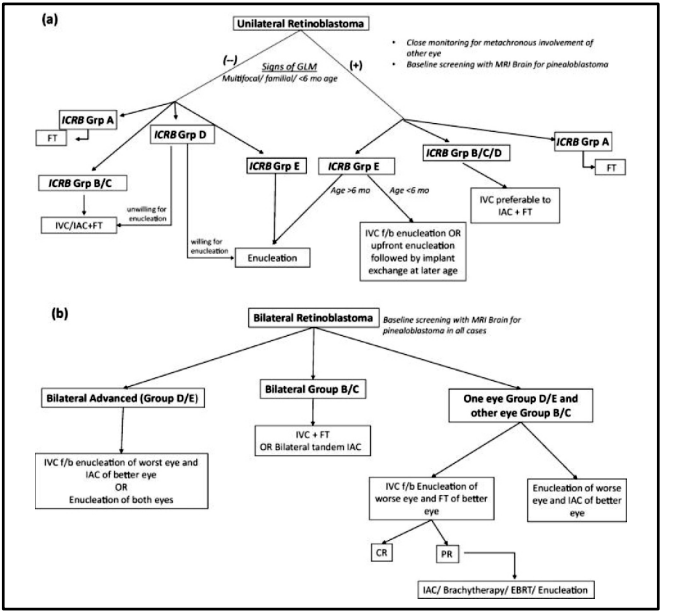
- Group B with macular involvement.
- Group C
- Group D +/- IVC
- Group E with visual potential

CONTRAINDICATIONS:

- Optic nerve involvement or intracranial extension on MRI.
- Metastatic disease

FREQUENCY:

Melphalan will be administered every 3- 4 weeks for a maximum of 4 doses. EUA with Retcam images will be performed at 3-4 weeks after each dose to assess response and therapy will only be continued if response is documented.



7. Enucleation Guidelines in Retinoblastoma

1. Purpose and Scope

These guidelines provide evidence-based, internationally informed recommendations on indications, preoperative evaluation, surgical technique, orbital implant selection and sizing, post-enucleation adjuvant therapy, and rehabilitation.

2. Indications for Enucleation

A. Primary Enucleation

- Advanced intraocular disease (**IIRC Group E, AJCC cT3/cT4**) with no potential for vision salvage and high risk of extraocular extension[1-3].

- Typical indications include: massive intraocular tumor, neovascular glaucoma, anterior segment involvement, vitreous hemorrhage, phthisis bulbi, or painful blind eye.

B. Secondary Enucleation

Performed when conservative or globe-salvaging therapies fail:

- Persistent, recurrent, or progressive tumor despite chemotherapy or focal therapy[4].
- Media opacities such as vitreous hemorrhage obscuring tumor monitoring[4].
- Phthisical or painful eyes following high-dose chemotherapy.
- Prolonged treatment (>9 cycles systemic chemotherapy, age >36 months before enucleation) increases high-risk pathology[4].
- Delays beyond 1 month after failure recognition raise metastatic risk[4].

3. Preoperative Evaluation

- **Examination under anaesthesia** anterior and posterior segment documentation.
- **MRI orbits and brain** high -resolution (1 mm slices), diffusion-weighted, volumetric T2 sequences to assess optic nerve invasion, scleral integrity, extraocular spread, and trilateral retinoblastoma[3].
- **Systemic workup**: bone marrow biopsy and CSF cytology when metastasis is suspected[3].
- **Baseline measurements**: contralateral axial length, orbital dimensions, fundus photography.
- **Clinical AJCC 8th Edition (cT)** should be documented for risk stratification, treatment planning, and research comparability[5].

4. Surgical Technique

A. Traditional Enucleation

- Involves standard “no-touch” enucleation.
- Extraocular muscles are detached and sutured directly to the implant surface.

- Provides good centration and orbital volume replacement, but motility is limited and risk of late implant exposure is higher[6].

B. Myoconjunctival Enucleation

- Rectus muscles are anchored within Tenon's capsule and conjunctiva rather than sutured to the implant.
- Enhances prosthetic motility, reduces implant exposure risk, and produces more natural cosmetic outcomes compared to the traditional method[7].

General principles:

- Preserve a long optic nerve stump (~15mm)[3].
- Ensure layered closure of Tenon's capsule and conjunctiva.
- Maintain haemostasis and asepsis to minimize hematoma and infection.

5. Orbital Implant and Prosthesis

Implant Placement

- A primary orbital implant should be placed at the time of enucleation to restore orbital volume and support growth.

Materials

- **Porous implants** (hydroxyapatite, porous polyethylene/ Medpor®, bioceramic): permit fibrovascular ingrowth, provide good motility, but are costly[8].
- **Non-porous implants** (PMMA, silicone, acrylic): cost-effective, effective, and widely used in resource-limited settings[9].

Wrapping

- Wrapping porous implants with donor sclera, mesh, or synthetic material reduces exposure risk[10].
- The myoconjunctival technique avoids direct muscle suturing onto the implant, enhancing motility[7].

Sizing by Age

- <1 year: 16–18 mm

- 1–2 years: 18–20 mm
- 3–5 years: ~20 mm[11].

6. Post-Enucleation Risk Classification (AJCC 8th Edition)

Low-Risk Pathology

- Tumor confined to retina.
- No massive choroidal invasion.
- No optic nerve invasion beyond lamina cribrosa.
- No scleral involvement.
- **Management:** Enucleation alone; no adjuvant chemotherapy[12].

Intermediate-Risk Pathology

- Focal choroidal invasion (<3 mm).
- Prelaminar or laminar optic nerve invasion.
- Anterior segment invasion. **Management:** 2–3 cycles adjuvant chemotherapy (vincristine, carboplatin, etoposide) or close observation in reliable follow-up settings[13].

High-Risk Pathology

- Massive choroidal invasion (>3 mm thickness or >4 mm^2).
- Retrolaminar optic nerve invasion.
- Tumor at optic nerve margin.
- Scleral invasion. **Management:** 4–6 cycles systemic chemotherapy; orbital radiotherapy if gross residual disease[14].

7. Complications of Enucleation

- **Immediate:** haemorrhage, infection, orbital cellulitis.
- **Early:** implant exposure, extrusion, socket contracture.
- **Late:** poor prosthetic motility, enophthalmos, socket asymmetry, psychosocial adjustment challenges.
- **Prevention:** meticulous surgical technique, appropriate implant sizing, and long-term socket care.

8. Post-Enucleation Rehabilitation

- **Prosthesis fitting:** conformer placed at surgery, definitive prosthesis at 6–8 weeks.
- **Psychological and family support:** counselling to address cosmetic concerns and emotional impact.
- **Ongoing surveillance:** monitor contralateral eye, socket growth, and secondary tumour risk in heritable retinoblastoma.
- **Multidisciplinary care:** ocularist, oncology, psychology, and genetic counselling support.

SYSTEMIC CHEMOTHERAPY:

Vincristine, Etoposide, Carboplatin (VEC protocol) 6 cycles at 28 day interval.

8.FOLLOW-UP PROTOCOL RETINOBLASTOMA

ACTIVE DISEASE:

Every three weeks after focal therapy. Once tumor is inactive keep under close surveillance and extend the follow-up period by 2 weeks to no more than 6 weeks until the disease is inactive for 3 months.

INACTIVE DISEASE

Consider as germline disease.

Once the disease is inactive for 4 months extend the follow-up period to:

2 months for younger than 2 years.

3 months until 3 years of age.

4 months until 4 years of age (consider OPD exam under sedation).

6 months for 4 till 9 years of age.

Annual exam after 9 years. In case of relapse go back to 3 weekly protocol.

DO NOT forget to examine the enucleated socket if indicated for mass, infection, fit of prosthesis, implant exposure or extrusion at every EUA or OPD visit.

NOTE: Late-onset new tumours tend to develop in the peripheral retina, where they have lesser impact on vision, but are more difficult to visualize completely on dilated eye examinations in the eye clinic, and therefore may require EUA.

ONCOLOGY FOLLOWUP

Rb survivors treated with chemo-therapy or EBR undergo oncology clinic follow-up at 3- to 6 monthly intervals for 5 years after finishing chemotherapy, and then every 1 2 years until age 18 years, and then lifelong follow-up every 2 years.

persons carrying an RB1 germline mutation, or non-germline Rb survivors treated with chemotherapy or EBR, should be seen in an oncology clinic for counselling about risk of secondary non-Rb cancers, annually until age 18 years, then lifelong follow-up every 2 years in an adult oncology facility.

High risk RB patients (high risk pathology, extraocular RB) should be kept on close observation with the oncology team.

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REFRACTIVE ERRORS

SCREENING AND MANAGEMENT GUIDELINES

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When clarity replaces blur,
a child's horizon opens like
a flower turning to light

Chapter 3

REFRACTIVE ERRORS

I. INTRODUCTION AND RATIONALE

Refractive errors, including myopia, hyperopia, astigmatism, and anisometropia, are the most common correctable causes of visual impairment in children. When uncorrected during the sensitive years of visual development—typically birth through seven or eight years—they can lead to amblyopia, strabismus, compromised stereopsis, reduced academic performance, and long-term visual disability. Globally, nearly one in five preschool children and almost one in four school-aged children require refractive correction[1].

The burden is particularly high in South Asia. Studies from Pakistan report refractive error prevalence ranging from 8.9% in Karachi school children [2]to 24.4% among older students in Lahore, and exceeding 50% in Faisalabad and Haripur cohorts where myopia is rising dramatically[3]. Among toddlers, nearly 14% demonstrate reduced acuity due to refractive error, most often myopic astigmatism[4]. Similar trends appear across India, where pediatric ophthalmologists frequently rely on clinical experience in the absence of nationally standardized prescribing guidelines[4].

This unified guideline synthesizes global standards (AAPOS, AAO PPP, ICO) with regional epidemiology and clinical realities from Pakistan and India. It aims to provide clinicians with a comprehensive, developmentally informed, evidence based approach to pediatric refraction.

II. DEVELOPMENTAL CONTEXT OF REFRACTIVE ERRORS

The refractive state of the eye evolves significantly throughout infancy and early childhood. During the first two years of life, the eye undergoes the process of emmetropization, a visually guided mechanism that coordinates axial growth with changes in corneal and lenticular power to reduce hyperopia and astigmatism.

Most infants begin life with mild hyperopia, typically between +1.00 and +2.50 diopters, accompanied by variable astigmatism. Rapid reduction in hyperopia occurs during the first 18 to 24 months, and by age six, most children approach emmetropia[5]. Myopia generally emerges

after this age, although earlier onset is increasingly common in populations with intense near visual demand.

Table 1: Age-Expected Refractive Ranges.

Age Group	Typical Refractive State	Physiological Basis	Clinical Implications
Newborn (0–12 months)	Physiological **hyperopia** (+2.00 to +3.00 D); significant astigmatism common	Short axial length; steep cornea; thick, powerful lens	Monitor for high hyperopia, anisometropia, and high astigmatism—risk of amblyopia
Toddler/ Preschool (1–5 years)	Rapid shift toward **emmetropia**	Fast axial elongation; corneal flattening; lens thinning	Failure of emmetropization indicates risk of refractive amblyopia
School age (6–12 years)	Near-emmetropia or onset of **juvenile myopia**	Continued axial growth; reduced outdoor time; increased near work	Increasing myopia prevalence; requires regular screening and early intervention
Adolescence (13–19 years)	Progressive **myopia**, stabilizing late teens	Axial elongation slows; accommodative lag in some individuals	Myopia control strategies most effective before stabilization
Young Adult (20–40 years)	Relative stability; mild myopic drift in some	Occupational/near-work demands; stability in corneal parameters	Refractive surgery planning; watch for small myopic progression
Mid-life (40–60 years)	Stable spherical equivalent with onset of **presbyopia**; slight hyperopic shift	Loss of lens elasticity; changes in lens curvature and refractive index	Need for near correction; reassess refractive error periodically
Older Adult (60+ years)	**Myopic shift** from nuclear sclerosis; irregular astigmatism	Lens hardening, nuclear opacity; corneal flattening	“Second sight”; fluctuating refraction; cataract assessment essential

Cortical visual plasticity peaks during early childhood and remains significant until approximately seven or eight years of age. Early intervention, especially before this neural plasticity declines, is critical to ensuring the development of normal binocular vision and avoiding amblyopia[6].

III. EPIDEMIOLOGY AND RISK FACTORS

Refractive errors appear across all pediatric age groups but vary by region and environmental exposure. International preschool studies report hyperopia between 4 and 12 percent, astigmatism around 10 to 15 percent, and early childhood myopia between 1 and 4 percent. In East Asia, myopia prevalence exceeds 80 percent by adolescence[6].

Genetic predisposition is a strong determinant, particularly parental myopia[7]. Environmental factors such as limited outdoor time, increased near work, and socioeconomic constraints contribute significantly[8]. Prematurity, retinopathy of prematurity, and congenital cataract represent key perinatal and ocular risk factors. Children with neurodevelopmental disabilities have substantially higher rates of refractive error and require enhanced surveillance.

IV. CLINICAL PRESENTATION AND SCREENING

Children seldom articulate blurred vision: instead, refractive errors manifest as behavioral adaptations. These may include squinting, head tilt, holding objects very close, persistent eye rubbing, or poor attention during near activities. A decline in school performance or aversion to visually demanding tasks can also be indicative.

Vision screening is essential throughout childhood[9]. Red reflex screening is performed at birth and during infancy. Between six and twelve months, instrument-based screening—such as photo screening or handheld autorefraction—can identify amblyogenic risk factors. Between one and three years, clinicians may continue instrument-based methods or use age-appropriate acuity tools. By ages four and five, symbol-based tests such as Lea or HOTV become reliable. From age six onward, annual visual acuity testing with Snellen or Log MAR charts is recommended.

V. DIAGNOSTIC EVALUATION

Accurate diagnosis requires age appropriate visual acuity assessment, ocular health examination, and cycloplegic refraction.

In preverbal children, Teller or Cardiff acuity cards offer the best estimates of visual acuity. Verbal toddlers respond well to Lea Symbols or

HOTV presentations, while children four years and older perform best with standardized Log MAR charts. Crowding bars are essential for diagnosing amblyopia.

Cycloplegic refraction remains the gold standard for pediatric patients because active accommodation can mask hyperopia or mimic myopia. Any child under six years, as well as those with strabismus, poor cooperation, amblyopia, or unexplained low vision, should undergo cycloplegia for accurate measurement.

Table 2: Cycloplegic Agents and Recommended Use (Document 2 Table).

Agent	Concentration	Preferred Use
Cyclopentolate	1%	Standard for children >6 months
Cyclopentolate	0.5%	Infants <6 months
Tropicamide	1%	Adjunct in older, cooperative children; insufficient alone
Atropine ointment	1%	High hyperopia, accommodative esotropia, or unreliable exam

In South Asian pediatric populations, where accommodation tends to be stronger and darker irides may reduce drug penetration, atropine ointment is frequently required for complete cycloplegia, especially in esotropes younger than five years. Verification of cycloplegia through dynamic retinoscopy is advisable whenever doubt exists.

Instilling cycloplegic eye drops in a child requires careful technique to ensure safety, effectiveness, and minimal distress. Cyclopentolate is an anticholinergic agent used to dilate the pupil and paralyze accommodation (cycloplegia), commonly for pediatric eye exams. Below are the recommended steps:

Preparation

- Wash your hands thoroughly with soap and water.
- Check the label and expiration date of the cyclopentolate bottle (common concentrations: 0.5%, 1%—dose and concentration depend on child's age, weight, and indication).

- Explain the process in age-appropriate language (e.g., “We’re going to put a special drop in your eye to help the doctor see better—it might feel a little funny but won’t hurt”).
- Position the child:
- For infants/toddlers: Lay supine on exam table or in caregiver’s lap; wrap snugly in a blanket if needed to prevent grabbing.
- For older children: Have them sit upright, head tilted back or lie supine; encourage them to look up at a target (e.g., a sticker on the ceiling).
- Have tissues or gauze ready to absorb excess drops.

Administration

- Gently pull down the lower eyelid to create a small pocket (avoid touching the eye with the bottle tip).
- Instill 1 drop into the lower conjunctival sac (not directly on the cornea to reduce stinging).
- Immediately apply gentle pressure to the nasolacrimal duct (inner corner of the eye near the nose) for ~1–2 minutes:
- Why? This minimizes systemic absorption and reduces risk of side effects (e.g., tachycardia, flushing, irritability, or—rarely—CNS effects like drowsiness or agitation).
- Repeat for the other eye, using a new drop (don’t reuse the same drop/dropper tip).

Post-Instillation

- Wipe away excess with clean tissue/gauze.
- Wait the recommended time (typically–30 45 minutes for full cycloplegic effect; some protocols use a repeat dose after 5–10 minutes for deeper effect).
- Monitor for adverse reactions:
- Mild: transient stinging, light sensitivity, blurred vision.
- Systemic (more common in infants/young children or with higher concentrations): flushed face, fever, tachycardia, drowsiness, irritability, or (rarely) hallucinations/seizures.
- Seek medical help if the child becomes excessively lethargic, hyperthermic, or inconsolable.

Safety Tips

- Use lowest effective concentration (e.g., 0.5% for fair-skinned or younger children; 1% for darker irises or older children—per clinician guidance).
- Avoid in children with known hypersensitivity or conditions like Down syndrome, cerebral palsy, or seizure disorders—use with extra caution and lower doses.
- Caregivers should wear gloves if concerned about accidental exposure (cyclopentolate can cause mydriasis in adults too!).

A complete binocular vision assessment—including cover testing, stereopsis evaluation, and accommodative and convergence measurements—provides insight into the functional impact of refractive error. A comprehensive anterior and posterior segment examination ensures that decreased visual acuity is not attributable to ocular pathology.

PEDIATRIC REFRACTION PATHWAY

- -Identify indications
- -Symptoms
- -Strabismus
- -Amblyopia risk
- -Abnormal red reflex
- -High risk infants
- -Post surgery

Assess co-operation of the patient

Administer appropriate cycloplegic agent

perform objective refraction-cycloplegic retinoscopy

Interpret findings using developmental norms and amblyopia

Prescribe spectacles addressing refractive magnitude, alignment, and symptoms

Plan followup based on age, risk category and visual development

VI. INTERPRETATION AND THRESHOLDS FOR PRESCRIPTION

Prescription decisions depend on refractive magnitude, age, symptoms, and amblyogenic risk. Hyperopia, especially when associated with esotropia, must be corrected fully[10]. Myopia is corrected for functional clarity, and when progression is detected, myopia-control interventions should be initiated promptly. Astigmatism of 1.50 diopters or greater demands correction, and even lower amounts may be addressed when they impairacuity. Anisometropia exceeding one diopter in spherical equivalent or 1.50 diopters in cylinder almost always requires immediate, full correction due to the high risk of unilateral amblyopia.

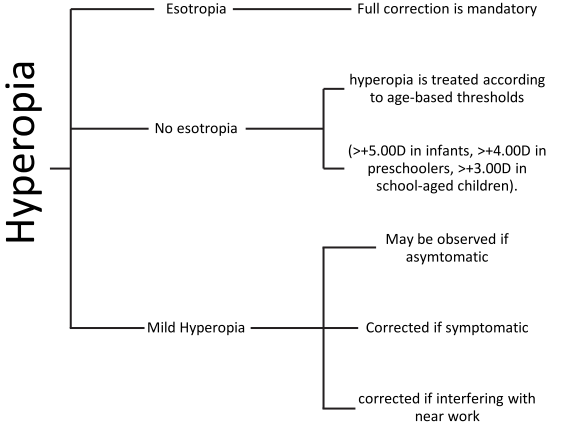
Table 3: Prescription Thresholds.

Condition	Prescribing Threshold	Notes
Hyperopia with esotropia	Full cycloplegic correction	Fundamental for alignment
Hyperopia w/out esotropia	>+5.00 D (infants); >+4.00 D (preschool); >+3.00 D (school-age)	Consider symptoms & near demand
Myopia	≥ −1.00 D	Initiate myopia control if early onset
Astigmatism	≥1.50D (infants); ≥1.00D (preschool); ≥0.75D (school-age)	Amblyogenic if uncorrected
Anisometropia	>1D hyperopic, >3D myopic, >1.50D cylinder	High amblyopia risk

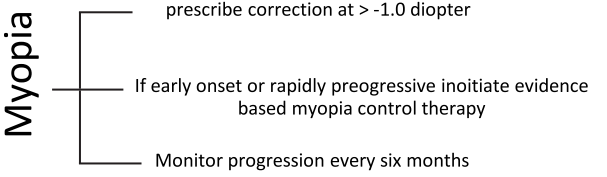
Management strategies

Full hyperopic correction is necessary when esotropia is present[11]. Older children without strabismus may tolerate slight under correction of hyperopia. Myopia should be fully corrected; under correction may accelerate progression[12].

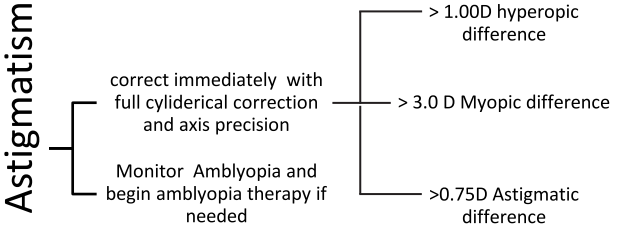
Hyperopia Management



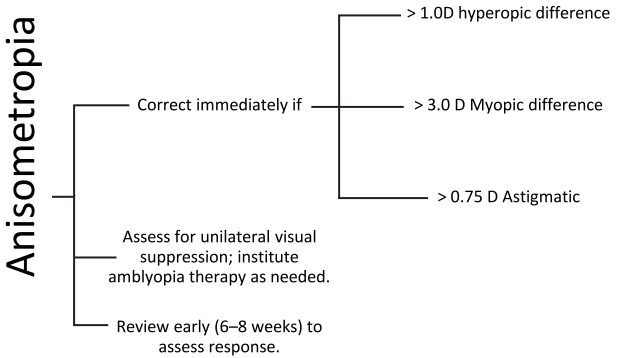
Myopia Management



Astigmatism Management



Anisometropia Management



VII. Management strategies

Spectacles remain the primary modality for refractive correction. Polycarbonate lenses are recommended due to impact resistance and ultraviolet protection. Proper frame fitting enhances compliance and ensures appropriate optical alignment. Frame should be Break-resistant, light, colorful, preferably with nose pads, elastic head bands for infants Materials- titanium (ideal), plastic materials (cellulose acetate, polyamide), silicon-based rubber frames (light and pliable) Soft ear support, elastic band to prevent peeping over the frame. Other metal frames for e.g., those with nickel can cause allergic reactions. Lens should be Impact-resistant, thin, light, scratch free, UV protection, high index (preferably).

Type of Lenses and their characteristics				
Materials	**CR39**	**Trivax**	**Polycarbonates**	**High index plastic**
Impact resistance	Good	High	High	Very high
Refractive index	1.498)	1.586	1.586	1.6-1.74

- Contact lenses provide excellent optical outcomes for high anisometropia, irregular astigmatism, and selected adolescents. Myopia-control therapies, including low-dose atropine, orthokeratology, dual-focus or multifocal contact lenses, and DIMS/ HALT spectacle lens designs, are now well supported by clinical evidence and reduce myopia progression by 40 to 65 percent.
- Amblyopia management begins with optical correction alone for several weeks. Many children show substantial improvement without further therapy. Persistent amblyopia requires occlusion therapy or pharmacologic penalization. Treatment typically continues until vision stabilizes, with careful monitoring for recurrence.
- Environmental and behavioral interventions, particularly outdoor exposure for a minimum of two hours daily, have demonstrated protective effects against myopia onset. Regular breaks during near tasks and attention to posture and lighting further support visual comfort.

Prescription in special conditions:

- Prescription in pseudophakia and aphakia: Refractive error as assessed by retinoscopy should be prescribed immediately irrespective of age in pseudophakia and aphakes[13]. Only near glasses are sufficient in children up to 2 years and thereafter with the added demand for distance vision bifocals are to be prescribed. Occlusion/ patching is needed in unilateral pseudophakic/aphakes or bilateral cases with unequal vision.
- Gross developmental delay, Down's syndrome or mental retardation: Refraction and prescription according to retinoscopy can be given as early as 6 months of age[14].
- Retinopathy of prematurity (ROP): Myopia tends to progress in cases of ROP and prescription can be given according to retinoscopy as early as 6 months[15].
- Esotropia- Hyperopia of ≥+1.5D must be prescribed in children with esotropia. Overcorrection can be done provided vision does not fall below 6/12. Bifocals are needed in cases with high AC/A ratio. Weaning of glasses should start at the age of 7 years depending on the retinoscopy but a close watch on esotropia is needed, and weaning is stopped or reversed if esotropia recurs with glasses on weaning.

VIII. FOLLOW-UP AND MONITORING

Follow-up schedules must reflect the period of greatest visual plasticity and risk.

Table 4: Follow-Up Intervals

Age/Condition	Follow-up Interval
Infants	Every 3–6 months
Preschool children	Every 6–12 months
School-aged children	Yearly
Amblyopia treatment	6–8 weeks
Progressive myopia	Every 6 months
Post-operative children	Individualized, often more frequent

Cycloplegic refraction should be repeated whenever visual acuity worsens, compliance is doubtful, or before concluding amblyopia therapy.

IX. DOCUMENTATION STANDARDS

Thorough documentation ensures continuity of care. The medical record should capture the cycloplegic agent, dosage, timing, retinoscopy values with working distance correction, cooperation level, binocular findings, prescription rationale, and follow-up plans. Such documentation becomes especially critical during the amblyogenic years when small changes in refractive status may have significant developmental impact.

X. FUTURE DIRECTIONS

Rapid advances in refractive research continue to transform clinical practice. Artificial intelligence may support early detection and risk stratification. Genetic testing offers promise for predicting refractive trajectories. Studies are underway to determine the impact of digital screen exposure on emmetropization. As these innovations develop, revisions to pediatric refraction guidelines will be needed to incorporate new evidence.

XI. CONCLUSION

Pediatric refraction is a critical component of child eye health and requires an understanding of visual development, accurate diagnostic techniques, rigorous prescribing standards, and consistent follow-up. Early and precise correction not only improves visual acuity but also supports neurodevelopment, academic readiness, and long-term ocular health. In high-burden regions such as Pakistan and India, standardized, evidence-based approaches such as those outlined in this unified guideline are essential to reducing preventable visual impairment and ensuring every child achieves their full visual potential.

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RETINOPATHY OF PREMATURITY

SCREENING AND MANAGEMENT GUIDELINES

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"Against ROP, timely intervention becomes the gentle hand that protects a newborn's dawn."

Chapter 4

RETINOPATHY OF PREMATURITY

1. BACKGROUND

Retinopathy of Prematurity (ROP) is a progressive vaso-proliferative retinal disorder affecting premature and low-birth-weight infants and is a major cause of preventable childhood blindness, particularly in low- and middle-income countries. With improving neonatal care in Pakistan, more preterm infants survive, resulting in a higher incidence of ROP. Timely screening, accurate staging, and prompt treatment are therefore essential.

This guideline adapts international evidence-based recommendations (ICROP-3, FRC Ophth, AAO/AAP) to Pakistan's healthcare context. Persistent challenges—including high prematurity rates, variable NICU standards, and limited trained personnel—continue to hinder consistent screening, and the absence of a national protocol leads to heterogeneous practices.

The Ophthalmological Society of Pakistan's 2019 recommendations highlighted the need for structured collaboration between neonatology and ophthalmology and standardized screening pathways. Local data support the use of international criteria: G-ROP has been validated in Pakistani infants, and treatment patterns mirror global trends, with laser preferred for zone II disease and anti-VEGF used for zone I and aggressive ROP.

Technologies such as wide-field imaging, teleophthalmology, and artificial intelligence offer opportunities to expand screening and improve diagnostic consistency, especially in underserved regions.

These guidelines aim to establish standardized national practices for ROP screening, treatment, and follow-up by integrating international evidence with local realities.

3. ROP SCREENING GUIDELINES:

3.1. Eligibility for Screening

ROP screening should be performed for all infants meeting one or more of the following:

At Risk Infants	Eligibility Criteria
Gestational Age	All infants born ≤ 36 weeks should undergo screening.
Birth Weight	Infants ≤ 2000 g at birth should undergo screening.
Clinical Course	Larger infants with unstable clinical courses (oxygen therapy, sepsis, poor weight gain, ventilation, transfusions) may be screened at neonatologist discretion.

3.2. Timing of First Screening

This schedule ensures early identification and timely management of ROP[5].

1. All premature infants should have 1st ROP screening at 4 weeks chronological age irrespective of gestational age at birth.
2. For high risk infants screening can be done earlier at 3 weeks; at discretion of neonatologist.
3. If oxygen given for 3 to 4 weeks then aggression of ROP may show itself after another 2 weeks.

Rationale:

The **4-week screening timeline** captures the critical window when ROP changes typically begin to appear, ensuring timely detection without overburdening neonatal teams. Establishing a **standard 4-week rule** offers a **clear, consistent, and safer protocol,** minimizing confusion and variability in practice and promoting timely identification of disease.

3.3. Examination Considerations

ROP screening should be performed in an environment conducive to the safe handling of preterm infants. Prior to examination:

- The infant should be clinically stable and monitored appropriately.
- Adequate mydriasis should be achieved using standard dilating protocols.
- The examination should be conducted using indirect ophthalmoscopy or wide-field digital imaging where available.

- Trained personnel should assist in positioning and monitoring the infant during the procedure.
- Examination should be done at NICU if the staff is well trained or a Minor Op type setting where many babies can be seen one after the other.

3.4. Screening Protocol:

- **Indirect ophthalmoscopy** (20D/28D/30 or 40D lens; with scleral indentation should be done to view the periphery) performed by a trained ophthalmologist is the **gold standard** for ROP screening.
- **Wide-field digital imaging** (RetCam or similar) – captured by trained personnel and reviewed by ROP-trained ophthalmologists, useful for documentation, telemedicine, or when ophthalmologist availability is limited.
- **Dilatation protocol:** pupils are dilated by using2.5% phenylephrine and 1% tropicamide eye drops, one drop each instilled twice 1 hour before examination.

4. ICROP-3 Classification (2021–22 Update) [14]

4.1: Zones of Retina

Zone	Definition
Zone I	Circle centered on optic disc, radius = 2 × (disc → fovea)
Zone II	From Zone I to nasal ora serrata
Posterior Zone II *(new)*	First 2 disc diameters into Zone II (higher risk)
Zone III	Temporal crescent outside Zone II
Notch *(new)*	Extension of ROP from one zone into another

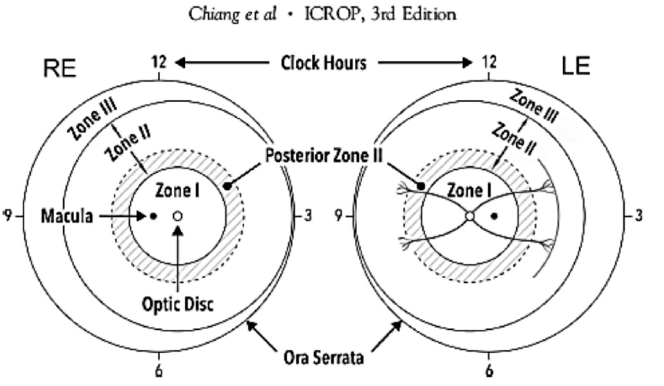

Figure 1: *Schema of right eye (RE) and left eye (LE) showing zone borders and clock hour sectors used to describe the location of vascularization and extent of retinopathy. Solid circles represent borders of zones I through III, and dotted circles represent borders of posterior zone II (2 disc diameters beyond zone I). Ahypothetical example of examination findings is shown in LE, representing approximately 3 clock hours of stage I disease in zone ii (note single line on drawing to document presence of stage I disease),*

4.2: Staging of ROP

Stage	**Description**
Stage 1	Demarcation line
Stage 2	Elevated Ridge
Stage 3	Ridge + extraretinal fibrovascular proliferation
6tage 4A	Partial detachment (extramacular)
Stage 4B	Partial detachment (macula involved)
Stage 5a *(new)*	Total detachment (open funnel)
Stage 5b *(new)*	Closed funnel, optic nerve obscured
Stage 5c *(new)*	With anterior segment abnormalities

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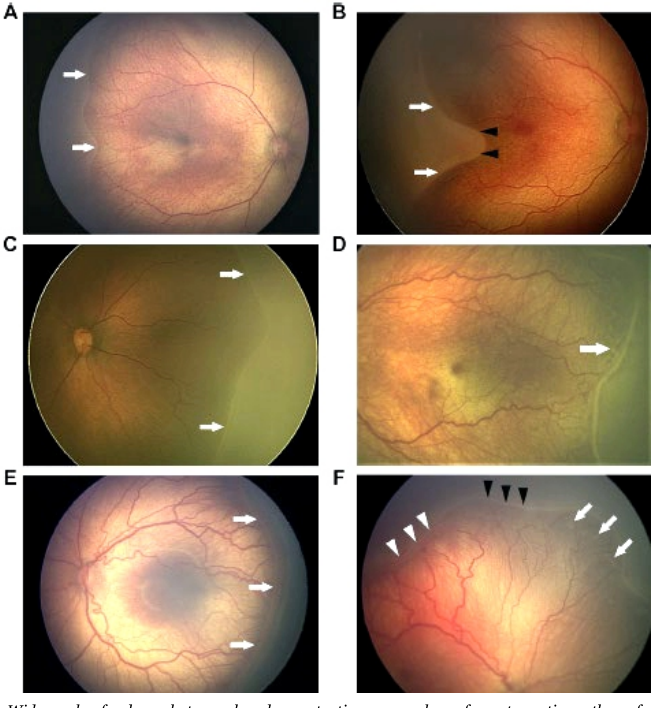
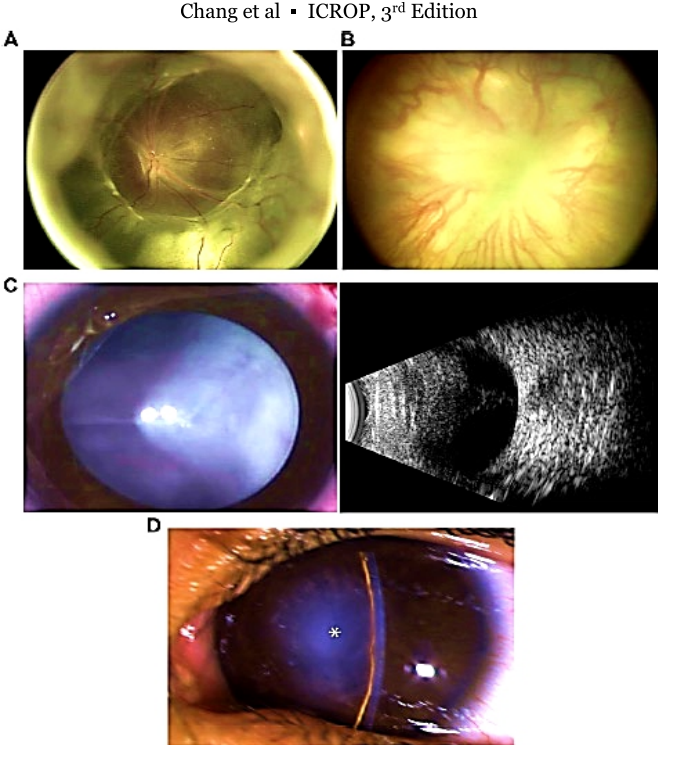


Wide-angle fundus photographs demonstrating examples of acute retinopathy of prematurity (ROP) stages I through 3. A, Stage I demarcation line at the border between vascular and avascular retina (white arrows). ***B:*** *Stage I demarcation line(white arrows) and associated notch (black arrow heads) between vascular arcades that would be considered zone I secondary to notch. Note pre-plus disease with mild retinal vascular tortuosity and dilation.* ***C:*** *Stage 2 ridge, which is raised (white arrows) and thicker than stage I.* ***D:*** *Stage 2 ridge.Note the so-called popcorn lesions posterior to the ridge (arrow) and pre-plus disease with mild vascular tortuosity and dilation.* ***E:*** *Stage 3 disease with extraretinal neovascularization (white arrows). Note plus disease with vascular tortuosity and dilation.* ***F:*** *Eye with both stage 2 (black arrow heads) and stage 3 (white arrow heads) disease and associated popcorn (white arrows). Note plus disease with vascular tortuosity and dilation.* ***Figs 5E and 5F:*** *Permission to reproduce previously published images from Arch Ophthalmol. 2005; 123:991-999.*



Images demonstrating examples of stage 5 retinopathy of prematurity (ROP). ***A:*** *Wide angle funds photograph showing stage 5aROP , characterised by a total retinal detachment with visible optic disc. Note the open-funnel configuration.* ***B:*** *Wide-angle fundus photograph showing stage 5B ROP, with no view of the optic disc or retina secondary to retrolental fibrovascular tissue. B-scan ultrasonography (right side) reveals total retinal detachment with a posteriorly closed funnel configuration.* ***D:*** *External photograph showing anterior segment characteristic of stage 5C ROP with anterior lens displacement, marked anterior chamber shallowing, central iridocapsular endothelial adhesion, and central corneal opacification (asterisk) that prevent view of closed-funnel retinal detachment.* ***Fig. 10B:*** *Permission to reproduce previously published images from Arch Ophthalmol. 2005;123:991-999.*

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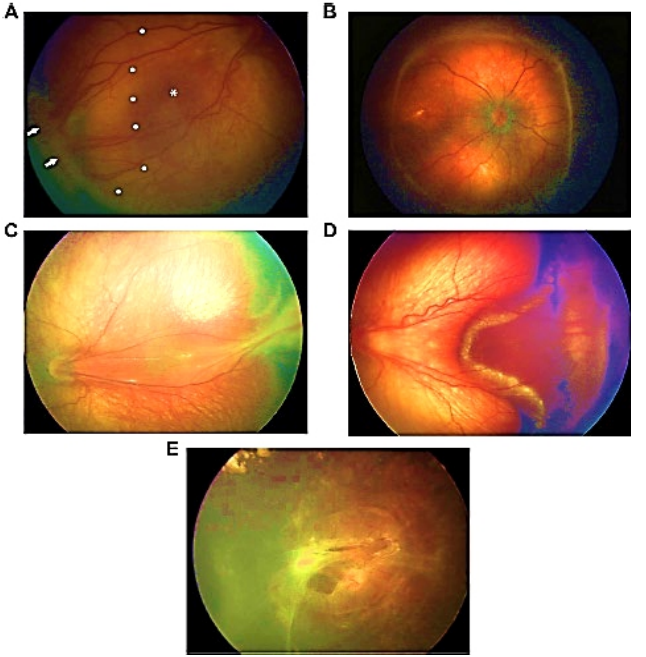

Wide-angle fundus photographs demonstrating examples of retinopathy of prematurity (ROP) stage 4. ***A:*** *Stage 4A ROP in the temporal retina. Traction on extraretinal neovascularization leads to retinal elevation (white dots), which may be recognized during ophthalmoscopy by subtle change in brightness and loss of visible retinal pigment epithelium granularity and choriocapillaris detail. Note that the approximate foveal center (asterisk) is not elevated and the extraretinal neovascularization (white arrows) may be significantly more peripheral than the posterior extent of the detachment.* ***B:*** *Stage 4A ROP with 360° tractional retinal detachment in the area of the peripheral ridge.* ***C:*** *Stage 4B detachment involving the macula. Note straightening of the arcuate vessels and dragged of the optic disc appearance.* ***D:*** *Stage 4B detachment with associated subretinal hemorrhage and lipid exudation into the macula.* ***E:*** *Volcano-shaped stage 4B ROP. In eyes with posterior ROP, contraction of pathologic neovascularization can result in detachment of vascularized retina into a volcano-shaped configuration.* ***Fig. 8C:*** *Permission to reproduce previously published images from Arch Ophthalmol. 2005;123;991-999.*

4.3: Disease Severity Terms

Definition	Description
Pre-plus	Vessel changes not sufficient for “plus”
Plus Disease	Dilated, tortuous vessels in ≥2 quadrants
Aggressive ROP (A-ROP) ***(renamed)***	Rapid progression, flat neovascularization, often Zone I/posterior Zone II, skips stages 1–2
Persistent Avascular Retina (PAR) ***(new)***	Retinal periphery fails to vascularize even after regression/treatment; risk of late complications
Reactivation ***(new)***	Recurrence of neovascularization after regression, common post anti-VEGF therapy

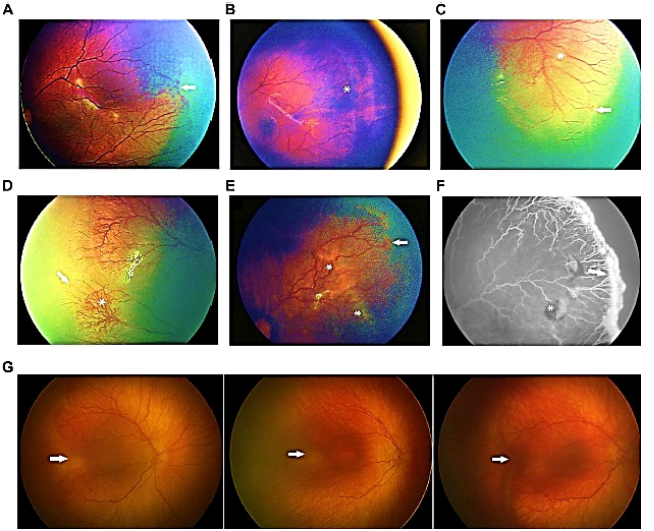

Figure 13: *Examples of retinopathy of prematurity (ROP) reactivation.**A:** Image obtained at 38 weeks' postmenstrual age (PMA) after intravitreal anti-vascular endothelial growth factor (VEGF) injection at 32 weeks' PMA with vascularization into the peripheral avascular retina. Demarcation line (arrow) at the leading edge is reactivated stage I ROP. **B:**Image showing a left eye at 100 weeks' PMA after treatment with intravitreal anti-CEGF injection at 38 weeks' FMA. Vascularization into the peripheral avascular retina is present. Often notable vascular abnormalities are present at the site of the original ridge and, in some cases, residual fibrosis (asterisk), which is not indicative of reactivation unless accompanied by increasing vascular activity. **C:**Image showing vascularization into the peripheral avascular retina with reactivated stage I disease (arrow) at 68 weeks' PMA, after treatment with intravitreal anti-VEGF injection at 37 weeks' PMA. Note multiple circumferential vascular loops at the site of the original ridge (asterisk). **D:** Image showing reactivation in a right eye at 67 weeks' PMA that had undergone intravitreal anti-VEGF injection at 33 weeks' and again at 52 weeks' PMA. Reactivated stage 3 disease (asterisk) is present posterior to the leading edge of vascularization (arrow). **E:** Image showing a left eye with reactivated stage 3 ROP at the leading edge (arrow at 50 weeks' PMA, after intravitreal anti-VEGF injection at 36 weeks' PMA. Vascularization into the peripheral avascular retina has occurred between the original ridge (asterisks) and anterior reactivation. **F:**Fluorescein angiogram obtained at 45 weeks' PMA of a left eye that had received an intravitreal anti VEGF injection at 34 weeks' PMA. Leakage is present both at sites of leading edge reactivation (arrow) and at the original border (asterisk). **G:** Image showing right eye with zone I disease treated with intravitreal anti VEGF injection at 34 weeks' PMA (left side, arrow) and that appeared regressed on clinical examination at 38 weeks' PMA*

5. Follow up protocol:

Follow-up intervals should be based on the most advanced stage of ROP observed in either eye, in accordance with international guidelines and local practice patterns [5,6].

Findings on Retinal Examination	Follow-up Interval
Zone I immature vessels / no ROP	≤1 week
Stage 1–2 ROP in Zone I	≤1 week
Stage 3 ROP in Zone II	≤1 week; consider treatment
Any suspected A-ROP	Treat Within 3-5 days (often <72 hrs)
Posterior Zone II immature vessels / Stage 2 Zone II	1–2 weeks
Stage 1 ROP Zone II / regressing Zone II	2 weeks
Zone III Stage 1–2 / regressing Zone III	2–3 weeks
Mature vasculature into Zone III	No further follow up

6. When to Stop Screening[10]

- Retina fully vascularized into Zone III, OR.
- If laser was used for treatment, Vascularization has reached ablated area.
- No progression on two consecutive visits.

7. Documentation Standards

All ROP examinations must be recorded in a structured and standardized format and include the following elements[4]:

- **Zone** of the retina involved (I, II, III).
- **Stage** of disease (0 to 5).
- **Presence or absence of Plus or Pre-Plus disease.**
- **Extent** of disease in clock hours.

- **Signs of aggressive or atypical disease.**
- **Assessment of regression or progression.**
- **Plan for follow-up or intervention.**

8. Additional Neonatal Care Consideration (for Ophthalmologists)

Oxygen Saturation Guidelines (SpO_2)

- **Key risk factor** for ROP progression = poorly controlled oxygen.
- **International recommendations**:
  - Target **90–94%** in preterm neonates.
  - Avoid prolonged SpO_2 >95%.
  - At **high altitude (>2500 m)**: threshold ≥87%.
  - During resuscitation: use **100% oxygen initially**, then titrate to **94–98%** post-stabilization.

◈ Implication for Ophthalmologists:

- Many cases of **A-ROP** correlate with inconsistent oxygen control.
- Collaboration with neonatology teams is essential.
- Reinforce that **oxygen titration is as important as timely eye exams** in preventing blindness.[14, 19, 20]
- Infants who have been on oxygen are protected for that period and can be seen 2 weeks after oxygen was stopped.

9. ROP treatment guidelines:

9.1. Indications for Treatment

Treatment is required for:

- Zone I (any stage) with plus disease.
- Zone I Stage 3 with or without plus disease.
- Zone II Stage 2 or 3 with plus disease.
- Aggressive ROP (A-ROP).

Consider treatment for:

- Zone II Stage 3 without plus disease.

Observation (with close follow-up) for:

- Zone I Stage 1–2 without plus.
- Zone II Stage 3 without plus. Treatment should be considered if high risk factors or patient is not likely to return for follow up.

9.2. Timing of Treatment[2]

- A-ROP and Zone I disease: Treat within 48 hours.
- Zone II Type 1 ROP: Treat within 72 hours.

Delays in treatment can result in irreversible blindness.

9.3. Peri-treatment Considerations[4]

- Informed consent should explain the disease, treatment, risks, and importance of follow-up.
- Anesthesia: GA or sedation with ventilation ; Anti-VEGF usually topical anesthesia with monitoring.
- Mydriasis: Phenylephrine 2.5% + Cyclopentolate 0.5% (two doses, 5 minutes apart, one hour prior).
- Procedures should be conducted in NICU or OT settings with appropriate neonatal monitoring.

10. Treatment Modalities[2-3]

10.1. Laser Photocoagulation

Preferred treatment for most Zone II disease and where follow-up is uncertain.

Settings:

- Laser: Diode 810 nm or green 532 nm.
- Power: 200–400 mW (adjust to achieve light grey burns). Often sufficient reaction is achieved between 130 –150 mW and rarely have to go to over 180mW.
- Duration: 200–300 ms. Often 70 ms is enough.
- Pattern: Near-confluent, ≤0.5 burn-width apart, across all avascular retina, sparing fovea and optic disc.

Advantages: Definitive treatment, low reactivation risk, no systemic VEGF suppression.

Disadvantages: Requires general anesthesia and laser availability; higher risk of high myopia.

10.2. Anti-VEGF Therapy

Useful for Zone I, posterior Zone II, and A-ROP.

Doses:

- Ranibizumab: 0.2 mg / 0.02 ml intravitreal.
- Bevacizumab: 0.625 mg / 0.025 ml intravitreal.

Technique:

- Inject 1–1.5 mm from limbus, straight back to avoid lens injury.
- Maintain strict asepsis with povidone-iodine and sterile instruments.
- Use a separate syringe/vial for each eye.
- Confirm central retinal artery perfusion post-injection.

Advantages: Less destructive to peripheral retina, easier for sick neonates who are not fit for GA. Also appropriate for those with small pupils with anterior vessels around the lens which can absorb laser. Finally, if there is some vitreous hemorrhage blocking view for good laser.

Disadvantages: Risk of reactivation, unknown systemic effects.

10.3. Vitreoretinal Surgery

Indicated for Stage 4–5 ROP.

Stage 4A:

1. In stage 4A if the detachment is peripheral then one can only try to do peripheral laser to ischemic area and barrage around the detached retina. This often works well without having to do vitrectomy.
2. Lens-sparing vitrectomy.

Stage 4B

1. If freshly developing warrants vitrectomy with/ without lensectomy to relieve the vitreous pull on retina. Decision on lensectomy depends on space between detached retina and lens.
2. If long standing laser retinopexy around the horns and to non-vascularized retina to just stabilize it.

Stage 5: Lensectomy with vitrectomy.

11. Post-treatment Follow-up

11.1. Laser-treated eyes[3-4]:

Timing	Purpose of Visit	Next Steps
5–7 days after treatment	Assess laser adequacy, check for skip areas, monitor for regression of plus disease and neovascularization.	Apply additional laser if untreated areas remain
1-2 Weekly until full regression of plus disease & neovascular activity	Confirm disease regression, ensure no skip areas or progression	Add "top-up" laser if activity persists
Monthly after regression until full vascularization or vessels reach ablated area (or 60 weeks PMA)	Monitor for late recurrence (rare after laser)	Discharge to annual follow-up for refractive/visual issues
3- 6 monthly	Detect sequelae or late recurrence	Extend follow up if stable

11.2. Anti-VEGF-treated eyes:

Timing	Purpose of Visit	Next Steps
48–72 hours after injection	Check central retinal artery perfusion, confirm regression onset	
2 weeks	Confirm regression at peak effect period	If stable→ extend interval
2- 4 Weekly until at least 65 weeks PMA	Detect **reactivation** or **persistent avascular retina (PAR)**	If reactivation or PAR or revascularization stalls → apply "insurance" laser If stable→ extend follow up
Annual long-term follow-up	Detect sequelae such as high myopia, strabismus, amblyopia, late detachment	Provide refractive correction, visual rehab as needed

11.3. Long-term follow-up:

- Monitor for refractive errors (esp. high myopia), strabismus, amblyopia, and late retinal detachment.

Age/Timing	What to Check
6 months corrected age	Refraction, ocular alignment, anterior & posterior segment
Annually (childhood)	Refraction, amblyopia, strabismus, glaucoma, retinal status
Teen years	Monitor for late retinal detachment, high myopia complications

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PRIMARY CONGENITAL GLAUCOMA

SCREENING AND MANAGEMENT GUIDELINES

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"In treating congenital glaucoma, we preserve the gentle clarity with which a child meets the world."

Chapter 5

PRIMARY CONGENITAL GLAUCOMA

Primary Congenital Glaucoma Overview/Background

1. INTRODUCTION

Disease:

Primary congenital glaucoma (PCG) is the most common non-syndromic glaucoma in infants and can lead to blindness due to elevated IOP and optic nerve damage. Its incidence in Pakistani children is reported to be nearly nine times higher than in Caucasian populations, likely related to high rates of consanguinity. Genetic predisposition, including affected first-degree relatives, is a major risk factor. Limited Pakistani studies have identified CYP1B1 mutations as an important contributor.

Pathogenesis:

PCG is a severe, inherited developmental anomaly of the anterior chamber angle and trabecular meshwork. These anatomical defects impede the normal outflow of aqueous leading to increased IOP and subsequent damage to the globe and optic nerve.

Disease History:

About 65–80% of PCG cases are bilateral, with a male-to-female ratio of 3:2. The mean age at presentation is around 11 months in developed countries but earlier—3 to 4 months—in high-incidence populations such as Pakistan and India, despite delays in seeking care.

The classic symptom triad of tearing, photophobia, and blepharospasm results from rapid ocular enlargement and corneal stretching due to elevated IOP. Corneal expansion occurs until age 2–3, after which collagen matures; scleral enlargement may continue until around age 10. Parents may notice bluish eyes, enlarged globes, or corneal clouding.

Without prompt treatment, PCG can cause progressive globe enlargement, corneal scarring, optic nerve damage, amblyopia, and permanent blindness. Early diagnosis and timely surgery are critical for preserving vision.

2. CLASSIFICATION

9th Consensus Report of World Glaucoma Association 2013

- Childhood glaucoma.
    - Primary.
    - Secondary.
        - Acquired (after birth).
        - Non Acquired (present at birth).
            - Ocular.
            - Systemic.

Terms such as 'developmental', 'congenital' or 'infantile' glaucoma lack clear definition and their use is to be discouraged.[6]

Hoskin and Shaffer classification

An anatomical system used to categorize developmental glaucoma in children.[7] dividing glaucoma based on the area of dysgenesis:

Isolated trabecular dysgenesis	Iridotrabecular dysgenesis	Corneotrabecular dysgenesis
Flat iris insertion 1. Anterior iris insertion 2. Posterior insertion 3. Mixed insertion	**Anterior stromal defects** 1. Hypoplasia 2. Hyperplasia	**Peripheral** (up to 2mm into peripheral cornea) e.g. Axenfeld's anomaly
Concave (wrap around) iris insertion	**Anomalous iris vessels** 1. Persistence of tunica vasculosa lentis 2. Anomalous superficial vessels	**Mid-peripheral** e.g. Rieger's anomaly
Unclassified iridodysgenesis	**Structured anomalies** 1. Holes 2. Colobomata 3. Aniridia	**Central** e.g. Peter's Anomaly

3. SCREENING PROTOCOLS FOR PEDIATRICIANS

Routine vision screening is recommended to be done by pediatricians at all well child visits as well as Community-Level Screening involving Pediatrician/Neonatologist/Local Healthcare Providers.

Examination should therefore be:

- At Birth.
- Routine Vaccination Visits:
  - 1 Week Post-Natal Visit: (e.g., Hepatitis B vaccine visit).
  - 3 Weeks Post-Natal Visit: (e.g., early infant check-up).
  - 6 Weeks Post-Natal Visit: (e.g., DTP-HepB-Hib vaccine visit).

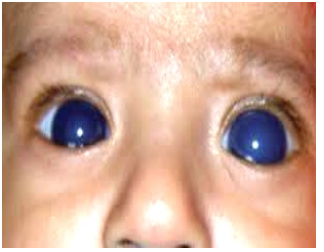
Examination should consist of:

- Gross examination.
  - Excessive lacrimation.
  - Photophobia.
  - Large cornea / haze.
  - Buphthalmos.
- Gross visual acuity examination – as per age.
- Distant Direct ophthalmoscopy.
  - Corneal haze.
  - Absent or faint red glow.

REFERRAL CRITERIA

Urgent referral to an ophthalmologist is to be made if:

- Visual acuity not corresponding with age.
- Blepharospasm.
- Photophobia.
- Lacrimation.
- Corneal Enlargement.
- Corneal haze.
- Enlargement of the globe.
- Dull red reflex.




Right: Bilateral Buphthalmos Left: Haab's striae

DIFFERENTIAL DIAGNOSIS

Common differentials include:

- Congenital nasolacrimal duct obstruction.
- Corneal abrasion/ulceration.
- Conjunctivitis.
- Megalocornea.
- Birth trauma.
- Dystrophy.
- Metabolic disorders.
- Disc coloboma, pit and/or hypoplasia.

4. MANAGEMENT PROTOCOLS

EXAMINATION IN OUTPATIENT SETTING

An outpatient department exam aims to gather as much information as possible to determine if an examination under anesthesia is warranted.

HISTORY

Following elements are important to for a good history taking in suspected cases of Primary congenital glaucoma:

- Age of onset of symptoms.
- Family history of glaucoma especially siblings.
- Parental consanguinity.
- Associated systemic illness.

- Topical and systemic medications.
- Past surgical treatments and lasers (cyclo-ablation).
- Allergy to medicines.

EXAMINATION PROTOCOLS

GROSS EXAMINATION

Look for the classic triad of epiphora, photophobia and blepharospasm.

VISUAL ACUITY TESTING

- Pre-verbal children.
  - Forced preferential looking techniques (Teller Acuity, Cardiff cards, Keeler Acuity cards).
  - Fixation and followed evaluation and assessing objection to evaluation of each eye.
- Verbal Children (3 years and above).
  - HOTV matching.
  - LEA symbols.
  - Tumbling E's.
  - Matching game or identification of symbols and letters.

SLIT LAMP EXAMINATION

- Corneal diameters
  - Normal value is 10 mm at birth. Abnormal values are ≥11 mm in a newborn, >12 mm in a child <1 year of age, and >13 mm at any age.
- Anterior chamber depth.

INTRA-OCULAR PRESSURE

Mean normal IOP in the pediatric population is 12.02± 3.74 mmHg. It can be measured by the following methods.

- I care.
- Tonopen.
- Goldman Tonometery.

CYCLOPLEGIC RETINOSCOPY

Any myopic shift from the normal age hyperopia or anisometropia should raise your suspicion for glaucoma.

FUNDUS EXAM

To document cup to disc ratio. Normal cup to disc ratio in a child is up to 0.3. However anomalies can be present also.

NOTE:

While some measurements can be unreliable in a crying child a careful clinical evaluation can provide strong clues towards diagnosis.

However, an accurate corneal diameter, intraocular pressure measurement, thorough anterior and posterior examination and gonioscopy should be performed under anesthesia.

Some ophthalmologists prefer using oral chloral hydrate in outpatient setting for a detailed exam and preoperative planning. Recommended dosage is 50-75mg per kg body weight.[8] It has the advantage of not significantly lowering IOP however requires monitoring until the child is fully awake. Common side effects include nausea, vomiting and irritability. Serious side effects like respiratory depression and bradycardia can occur.

COUNSELING

It is important to educate the parents and care-givers that PCG is a treatable disease provided it is diagnosed early and intervention is done timely. Management options should be thoroughly explained. Stress should be given to the need for follow –up visits. Parents need to be counseled that the child might need more than one procedure and that the refractive errors are prevalent in individuals with PCG, necessitating continuous use of corrective lenses. Genetic testing and counseling should be offered to high-risk families for family planning and early screening for subsequent siblings.

EXAMINATION UNDER ANESTHESIA (EUA)

EUA allows the examiner ample time for decision making and eliminates room for missing important findings.

It may be preferable that the surgeon takes a consent in advance to proceed for surgical intervention if indicated before bringing the patient in OT for EUA.

EQUIPMENT AND INSTRUMENTATION REQUIRED

- Hand held slit lamp/ Operating microscope.
- Calipers, speculum, forceps, fluorescein strips etc.
- Tonometer (e.g., Perkins, Tono-Pen, or iCare).
- Direct ophthalmoscope.
- Indirect ophthalmoscope.
- Retinoscope.
- Direct goniolens for Angle evaluation (e.g., Koeppe's or Swan-Jacob lens).
- Ultrasound pachymetry.
- Ultrasonography B-scan and A-scan.
- UBM (optional).

EXAMINATION REQUIRED

- **Gross Examination**
  - Systemic and facial examination for anomalies like mucopolysaccharidosis.

- **Corneal Diameter:**
  Normal value is 10 mm at birth. Abnormal values are ≥11 mm in a newborn, >12 mm in a child <1 year of age, and >13 mm at any age.
  - Calipers are the primary tool for measuring corneal diameter.
  - Both vertical and horizontal white to white diameters should be recorded in millimeters.
  - In advanced cases with buphthalmos, where limbal anatomy may be poorly defined, accurate measurement can be challenging.

- **INTRA-OCULAR PRESSURE**:
  Mean normal IOP in the pediatric population is 12.02± 3.74 mmHg.
  - IOP should be measured right after the child stops moving post inhaled anesthesia and before intubation to avoid artificially low readings.

- General anesthesia typically reduces IOP by 30%. It is advised that the anesthetist be told of the procedure in advance for the right choice of drugs given.
- Studies indicate that the use of eye speculum can elevate IOP by an average of 4 mmHg.[9]
- The time when the pressure was recorded should be noted and mentioned in documentation e.g. IOP OD 14 mmHg OS 15 mmHg 10 minutes into anesthesia.
- The same tonometer should be used for follow-up assessments to maintain consistency.
  - Handheld Goldmann applanation tonometer is the gold standard for measuring IOP.
  - The Schiotz tonometer is relatively inaccurate when used to measure IOP in infant eyes and should be avoided.[10]
  - The Tonopen device with a measuring surface of 2mm is well suited for small palpebral fissures.

- **CYCLOPLEGIC RETINOSCOPY**

  Any myopic shift from the normal age hyperopia or anisometropia should raise your suspicion for glaucoma.

- **FUNDUS EXAM**

  To document cup to disc ratio. Normal cup to disc ratio in a child is up to 0.3. However anomalies can be present also

- **A-SCAN**

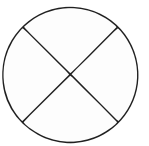
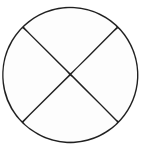
  Axial length should be measured to document any globe enlargement.

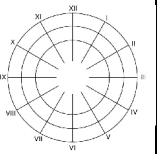
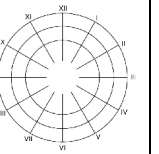
Age (Years)	Number	Axial length (mmo			
		Minimum	Maximum	Mean	SD
1	16	18.54	22.13	20.75	1.09
2	34	18.61	24.33	21.32	1.29
3	46	19.15	24.22	21.95	1.18
4	27	19.06	24.51	21.62	1.40
5	53	19.65	27.10	22.28	1.76
6	67	19.53	25.45	22.47	1.55
7	45	18.27	25.94	21.93	1.91

- Pachymetry, Ultrasound B-scan, A Scan axial length, OCT and anterior segment imaging with UBM can be done if available.

A printed pro forma should be available for writing the information collected during EUA mentioning clearly right and left eyes in two columns. The findings should be written in rows. Following information is collected and documented in systematic order to avoid missing any findings as below:

EUA REPORTING FORM

Date of Exam Name: Age: Gender MR#	Right Eye	Left Eye
IOP ➢ Time into anesthesia ➢ General anesthetic used ➢ Eye speculum (with/without)	mmHg	mmHg
Lids and adnexa (tarsal papillae)		
Anterior Segment ➢ Previous conjunctival bleb ➢ Haab striae ➢ Corneal opacifications ➢ AC depth ➢ Iris (Aniridia, atrophy, NVI) ➢ Pupil (location, irregularities, ectopia, pseudopolycoria) ➢ Lens (opacity,subluxation)		
Corneal Diameter ➢ Vertical ➢ Horizontal	mm mm	mm mm
Sclera ➢ Discoloration ➢ Thinning ➢ Staphyloma		
Gonioscopy ➢ Lens used ➢ Angle structures ➢ Iris insertion ➢ Pigmentation ➢ Vascularisation		
Vitreous ➢ Opacities ➢ Inflammation ➢ Hemorrhage		

Fundus ➢ Cup/Disc Ratio ➢ Nueroretinal rim ➢ Disc anomaly ➢ Macula ➢ Periphery ➢ Vessels		
Refraction ➢ Distance e.g. 2/3rd m ➢ Cycloplegic agent		
Axial length		
Pachymetry		

TREATMENT
MEDICAL TREATMENT

Medical treatment is only as a temporizing while awaiting surgery or given pre operatively to decrease IOP to clear the corneal haze for better visualization during examination and for surgery, and to avoid sudden decompression.

Applying pressure to the inner corner of the eye (punctal occlusion) immediately after instilling eye drops can help reduce systemic absorption and potential side effects, especially in infants. This should be performed for at least 1-2 minutes after drop instillation.

- Topical Timolol (Beta Blocker) is the first choice in pediatric glaucoma but is contraindicated in individuals with reactive airways.
- Combination of timolol and dorzolamide (Carbonic anhydrase inhibitor) twice a day in cases with insufficient reduction of the intraocular pressure (IOP) by a single topical agent brings about a good control of the IOP. Both medications are effective and well tolerated.
- The alpha2-agonists have more and potentially serious adverse effects in children and are contraindicated for children younger than 2 years of age.[11]
- Latanoprost (Prostaglandin analogue) though less effective in lowering IOP in children than in adults can be given.[12]

SURGERY SHOULD NOT BE DELAYED IN AN ATTEMPT TO ACHIEVE MEDICAL CONTROL OF IOP.

SURGICAL TREATMENT

When and what type of Surgery to be performed:

- Clear cornea.
- Age less than 3 years.
  - **GONIOTOMY**

    The success rates of goniotomy are highest in infants. After about 2-3 years of age, the trabecular meshwork becomes more collagenous and fibrous.[13]

    - **TECHNIQUE:**
      - Goniotomy ab interno requires excellent visualization of the angle and hence a clear cornea is an essential prerequisite.
      - The anesthesiologist should be informed prior to the procedure that the surgeon and the assistant will be rotating the patient's head back and forth and a secure placement of endotracheal tube should be kept in mind.[10]
      - The head of the patient is rotated nasally by 45 degrees to vertical while the surgeon sits at the temporal side.
      - Temporal paracentesis through the peripheral cornea is performed.
      - Intracameral miotic agent is injected. This opens up angle and stretches the iris to avoid engaging it with a Knife or Needle.
      - Viscoelastic is injected to deepen AC and Angle.
      - Swan-Jacob Gonioprism is placed on cornea after instilling coupling fluid on cornea,
      - The Barkan knife/23 G needle, attached to a syringe containing viscoelastic, is introduced and is passed through the limbus into the angle between iris root and schwalbe ring under gonioscopic view.

- It is then extended nasal and inferior and then nasal and superior. Nasal 120 degrees of the angle are easiest.

  A goniotomy is greatly facilitated by a surgical assistant who is familiar with the procedure and understands what the surgeon is trying to visualize and achieve.

- Difficult angle visualization
  - Age less than 3 years

    - **TRABECULOTOMY**
      - **TECHNIQUE:**
        - A conjunctival flap followed by a partial thickness scleral flap is made.
        - A radial incision at 12 o'clock is given to identify and expose the canal of schlemm.
        - Right and left Haams trabeculotome is used and the lower prong is passed along the canal on the inner side and rotated towards the anterior chamber to dissect the inner wall of the canal and trabeculum to enter the AC and then repeated on the other side of the incision. This allows opening about 180 degrees of the trabeculum communicating the canal to AC.
        - Entire 360 degree trabeculotomy can be achieved in a modified procedure using 6-0 prolene suture passed circumferentially through the canal.

- Older than 3 years
- After a failed primary procedure
- Combined with trabeculotomy

TRABECULECTOMY

Isolated trabeculectomy in PCG has very little success. It is recommended that it should be augmented by the use of antimetabolites like mitomycin-C and 5-flurouracil.[14]

- **TECHNIQUE:**
  - A conjunctival peritomy is made to expose the sclera.
  - A partial-thickness scleral flap, usually in a rectangular or triangular shape, is carefully created and hinged at the limbus.
  - A small, sterile sponge or swab, soaked in the chosen antimetabolite is placed under the scleral and conjunctival flaps. It is left in place for one to five minutes, depending on the surgeon's preference and the patient's risk factors for scarring. After the allotted time, the sponge is removed, and the area is thoroughly irrigated with a large volume of sterile saline to flush away any residual antimetabolite and prevent damage to surrounding tissues, especially the cornea.
  - A piece of the trabecular meshwork is then excised using a surgical punch to create a small hole (ostium) that connects the anterior chamber to the subconjunctival space.
  - A peripheral iridectomy is performed to prevent the iris from blocking the newly created drainage hole.
  - The scleral flap is carefully repositioned and secured with sutures. These sutures are placed to be tight enough to prevent the eye from becoming too soft but loose enough to allow fluid to seep out at a controlled rate.
  - The conjunctival flap is then closed with sutures, sealing the entire area.
  - Antimetabolites (MMC/5-FU)
    - Can be injected into the conjunctival tissue.
    - Traditionally applied using soaked sponges under the conjunctiva and scleral flap (see above).
  - Concentration of antimetabolite:
    - Mitomycin (MMC) 0.1-0.5mg/ml
    - 5-Fluorouracil (FU) 25-50mg/ml
- Duration: recommended time of application is 2 – 5 minutes.

POSTOPERATIVE CARE

- Topical steroids, either prednisolone 1% or dexamethasone 0.1% should be started along with topical tobramycin 0.3%.

- Topical medication should initially be given 2 hourly for the first one month and then tapered slowly over several weeks to months.
- The tapering schedule is highly individualized and depends upon the appearance of the bleb.

  A healthy functioning bleb will look diffuse, avascular and slightly elevated.

  - It is recommended to add oral antibiotics especially for filtration surgeries in our population to avoid risk of severe inflammation and infection after surgery.
  - Topical cycloplegic agents Atropine (1%) or cyclopentolate (1%) are given for the initial few weeks especially in first month to reduce ciliary spasm and to prevent anterior synechiae formation.

ACUTE POSTOPERATIVE COMPLICATIONS

- Hypotony (over-filtration/wound leak).
- Shallow or flat anterior chamber.
- Hyphema.
- Choroidal detachment.
- Suprachoroidal hemorrhage.
- Elevated IOP.
- Infection.

MANAGEMENT OF RAISED IOP AFTER SURGERY:

- Early Postoperative IOP Rise: Can be due to retained viscoelastic, inflammation, hyphema, or ciliary body shutdown followed by recovery. Initial management involves topical medications (beta-blockers, CAIs), massage of the bleb (if applicable and instructed).
- Late Postoperative IOP Rise: Often due to bleb fibrosis or encapsulation. Management may involve needling with anti-metabolites, topical medications, or further surgical intervention (e.g., repeat trabeculectomy, GDD).

MANAGEMENT OF POSTOPERATIVE SHALLOW AC:

The primary causes of a shallow AC after trabeculectomy can be broadly categorized into:

- **Hypotony** due to over filtration or wound/bleb leak: This is the most common cause.
- **Aqueous misdirection** (malignant glaucoma): A rare but serious complication.

Management

The management of a shallow AC is a step-wise process, moving from conservative to surgical intervention based on the severity and response to treatment.

Conservative

- **Mydriatics/Cycloplegics:** Pulls the lens-iris diaphragm posteriorly and away from the cornea. This is often the first and most effective step, particularly if there is a component of ciliary block or choroidal detachment.
- **Pressure Patching/Shielding:** this provides a gentle pressure to the eye, which can help to tamponade a wound leak and facilitate healing.
- **Bandage Contact Lens:** Covers and protects a leaking wound, providing a physical barrier that encourages spontaneous healing.[15]

Surgical and Invasive Management

If conservative measures fail or if the AC is severely shallow (e.g., Grade 3, with lens-cornea touch), surgical intervention is necessary to prevent permanent damage.

Reformation of the Anterior Chamber

Scleral Flap Sutures

Surgical Repair of Bleb Leak[16]

REFRACTORY CASES

- **Glaucoma Drainage Devices**
  Ahmed Glaucoma Valve is one of the most commonly used devices in pediatric glaucoma. The valve mechanism prevents the sudden drop in IOP in early postoperative period. Complications include tube erosion and retraction, plate migration and endothelial damage.[17]

- **Cyclodestructive Procedures**

  These include:

  Cyclocryotherapy (less common).

  Cyclophotocoagulation.

  - Transscleral cyclophotocoagulation (TS-CPC).
  - Endoscopic cyclophotocoagulation (ECP).
  - Micropulse cyclophotocoagulation (MP-CPC).

ROLE OF MINIMALLY INVASIVE GLAUCOMA SURGERY (MIGS) IN PCG

- The idea of MIGS is attractive to pediatric glaucoma specialists because most MIGS procedures are conjunctiva sparing.
- These include:
  - Kahook dual-blade.
  - Gonioscopy assisted transluminal trabeculotomy.
  - Trab 360.
  - OMNI surgical system.
  - PreserFlo ab-externo microshunt and
  - Endoscopic cyclophotocoagulation.

6. REHABILITATION AND FOLLOW UP

Visual rehabilitation is an ongoing process essential for maximizing visual potential in children with PCG.

IOP Monitoring:

- Quarterly for at least 2 years post-surgery, then
- Every 6 months or annually for life, depending on stability.

Cycloplegic Refraction: Should be performed every 6 months for life to detect and manage refractive errors (myopia, astigmatism), which are highly prevalent in PCG patients due to globe enlargement and corneal changes.

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