Table of Contents

add abbreviations

Glaucoma in India

Introduction

Glaucoma is the second leading cause of irreversible blindness in the world,[1] and accounts for almost 8% of global blindness.[2] A meta-analysis of 50 published population-based studies has shown that the global prevalence of glaucoma in people in the age group of 40–80 is 3.54%.[3] This report also states that the 64.3 million people with glaucoma detected in 2013 is likely to increase to 76.0 million in the year 2020 and to 111.8 million people in the year 2040. Out of total 39.36 million blind people globally,[2] more than 3 million are blind due to glaucoma.[4] India contributes highest regional burden of blindness which is 23.5% of global blindness.[5] Glaucoma contributes 5.8% and is the third leading cause of blindness in India after cataract (62.6%) and refractive error (19.7%).[6] The Indian report had estimated that 11.2 million people lived with glaucoma in the year 2009, including 6.48 million people with primary open-angle glaucoma (POAG) and 2.54 million people with primary angle-closure glaucoma (PACG).[7] In India the prevalence of blindness is 8.9 million,[5] of which Glaucoma contributes 12.8% of blindness.[8]

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To meet the challenge of preventing the irreversible blindness seen in Glaucoma, it is important to utilise the expertise of all tiers of health care in India that can cover rural and urban population. This combined with screening for other "silent blinding diseases" like Diabetic retinopathy would help in early detection.[9,10] Early diagnosis and appropriate management of glaucoma can stabilize the neuropathy and slow its progression in most patients reducing the risk of blindness. A long-term real-world study conducted in Sweden over more than 20 years demonstrated that population screening for open angle glaucoma reduced the prevalence of bilateral blindness by 50% [11] Screening the entire population is neither practical nor cost effective. However targetted screening of high risk. groups such as individuals >40 years of age, high myopes, diabetics, family history of glaucoma in first degree relatives is more feasible and cost effective. Screening using an initial automated test followed by evaluation by a specialist can enhance cost effectiveness. Highly specific tests are essential to minimize false positive referals. If rate of progression and cost of visual impairment are high, then the screening becomes cost effective.. National strategies to increase attendance at eye exams such as leveraging refractive error correction and diabetic screening programmes, can support glaucoma detection. Integrating glaucoma screening with robust cataract screening programmes, or general ophthalmology clinics can further improve detection rates. Similar guidelines have been proposed for glaucoma screening in the UK as well.[12]

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Plan:

- Who should be screened? At risk groups: people >40 years of age, high myopes, diabetics, family history of glaucoma in first degree relative.
- What tests should be used for glaucoma screening? Anterior chamber depth, intraocular pressure assessment (IOP), Fundus photography and Visual field testing, including online options when appropriate.
- Where and by whom should glaucoma screening be conducted? Primary health care centers like vision centers, general ophthalmologists clinic, Opticians, diabetologists clinic
- Who will evaluate and manage patients with positive screening results ? Specilaist ophthalmologists

- How would capacity be built to manage earlydiagnosed cases of glaucoma: Developing a skilled workforce of glaucoma specialists would require targetted training programs

Glaucoma Diagnosis Gap in India: A Looming Threat to Vision Care?

Glaucoma is the leading cause for irreversible blindness in India. affecting an estimated 12 million people, with 1.2 million already blind due to the disease sccording to WHO. Glaucoma is a condition where an accumulation of fluid in the eye increases the intraocular pressure resulting in optic nerve damage. The risk of glaucoma increases with age there is a significant gap in diagnosis in India, highlighting the need for greater awareness.

There are two main types of glaucoma: primary openangle glaucoma (POAG) and angle-closure glaucoma (PACG). Primary open angle glaucoma is the more common, painless, and develops gradually , often without symptoms until significant optic nerve damage occurs. The latter could entail an acute angle closure attack requiring immediate help from an ophthalmologist for appropriate treatment including laser iridotomy.

Importance of Glaucoma Diagnosis

In India, nearly 90 percent of glaucoma cases go undiagnosed.early detection to prevent permanent blindness in patients. Regular eye exams are essential to identify optic nerve damage due to open angle glaucoma early, a timely detection is key to successful treatment. Raising awareness about the risk factors and the importance of regular eye check-ups is critical especially in rural areas where both awareness and access to qualified ophthalmologists are limited. In the preferred practice patterns of the American Academy of Ophthalmology, comprehensive eye examinations are recommended for regular patients to rule out glaucoma over the age of 40 years. However, this practice is Underscoring the need for awareness and regular eye check ups.

Diagnosis and Treatment Techniques. In India, glaucoma is diagnosed by general ophthalmologists or sub-specilaists, with treatment tailored to the type of glaucomaTo detect glaucoma, eye-specialists carry out a series of tests to check the extent of the damage. The three screening tests comprise of the intraocular pressure, gonioscopy (a test to check the fluid drainage channels in the eye), examination of the optic nerve (nerve that connects the eye to the brain), and a test to check for non-seeing areas (scotomas) in the patient's field of vision. Once diagnosed, open-angle glaucoma is treated with eye drops to alleviate the pressure in the eye. The patient must diligently take the eye drops life long as prescribed by the treating doctor. If the eye drops do not help to control glaucoma, the specialist may suggest alternatives like laser or surgery

Surveys conducted in India, both population-based and hospitlal-based (such as APEDS, CGS, VES and CIES) highlight the common types of glaucoma observed in the country

- Primary open angle glaucoma (POAG)– Ocular hypertension (OHT), Normal tension glaucoma (NTG) and Juvenile open angle glaucoma (JOAG)
- Primary angle closure disease – Primary angle closure suspect (PACS), Primary angle closure (PAC), Primary angle closure Glaucoma (PACG)
- Common secondary adult glaucoma – Traumatic glaucoma, Steroid induced glaucoma (SIG), Glaucoma in Pseudophakia and Aphakia, Inflammatory/ Uveitic and Neovascular glaucoma
- Paediatric glaucoma – Primary Congenital Glaucoma (PCG), infantile/developmental, secondary inherited and non-inherited, syndromic, traumatic, steroid induced, glaucoma after congenital cataract surgery, ROP associated and other secondary post surgical glaucoma

A comprehensive approach is essential for developing appropriate, affordable, and accessible glaucoma screening and management strategies applicable to India. This article aims to present practical guidelines derived from the consensus of various glaucoma experts to enhance screening and managemenrtstrategies for glaucoma.

Methods

Over 30 leading glaucoma specilaists from institutions and in private practices, across the country collabotated to create these guidelines. The framework and outline was created by IHope team with two glaucoma specialists who were the leads for the project. They designed a framework for the guidelines. The guideline was to cover primary, secondary and tertiary levels of care under the following headings

Levels of care

1. Primary ophthalmic care – opticians, Optometrists, Primary Health Centers(PHCs)/ Community Healthy centers (CHCs)
2. Secondary – Managed by general ophthalmologists at dstrict hospitals
3. Tertiary care: Providing by glaucoma Specialists at institutes and at Medical colleges

An organogram or flow chart was recommended to clearly outline the roles and capabilities at each level of eye care

Headings Practical protocols for each of the conditions

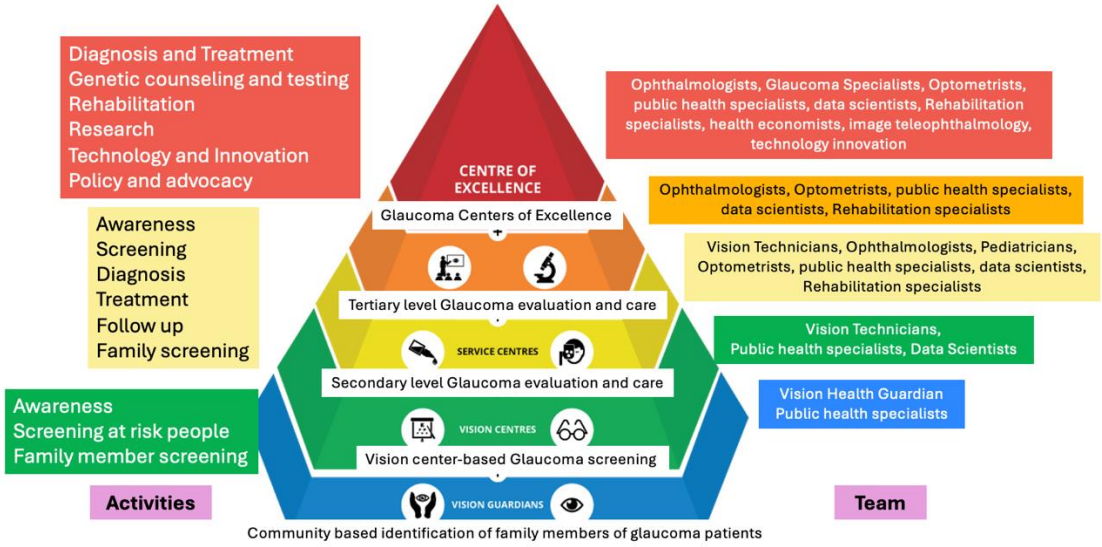
included

- Screening for glaucoma
- Evaluation protocol
- Diagnosis
- Counselling
- Therapy
- Regular review
- Re-evaluation

- Care and Support

There was a group lead, moderator and members (5-8) in each group

There were 4 groups that worked on Open angle glaucomas (POAG, JOAG, OHT, NTG), angle closure glaucoma (PAC spectrum), Secondary Glaucomas and Childhood glaucoma (PCG and others).



(i) Primary level—vision technicians/ Optical outlets--Opticians

Equipment	Evaluation	Management/Referral guidelines
• Vision chart (essential) • Torch (essential) • Slitlamp fixed or hand held (preferable) • Any tonometer (preferably Goldmann Applanation tonometer (GAT))	• History (steroids, trauma, family h/o glaucoma, past records, etc.) • Visual acuity assessment • Careful torchlight examination of	• Refer if intraocular pressure (IOP)>24 mm Hg, acuity < 6/9, family history, any anterior segment abnormality, especially altered pupillary response.

• Fundus photo (non-mydriatic camera with AI) (desirable) • Portable/ online Fields (desirable) • Teleophthalmology integration (desirable)	extraocular status and anterior segment • Pupillary shape/size /sluggish reaction • Tonometry • Slitlamp evaluation • Portable/ online field evaluation if present	• Can follow-up 3 monthly patients with IOP <24 mm Hg, and no risk factors • Refer all children with signs or symptoms of visual problems. • AI alert of glaucoma suspects confirmed by teleconsultation

(ii) <u>Secondary care level (General Ophthalmologist / Glaucoma Specialist)</u>

Equipment	Evaluation	Management/Referral guidelines
• Slit-lamp • GAT • Gonioscope – 2/4-mirror • 90D/78D/direct ophthalmoscope • Automated perimeter/ online visual field testing • Pachymeter (ultrasound or optical) • Fundus camera for disc photo with AI integration • Nd:YAG laser	Thorough history and examination. • Ask for steroid use • Repeat tonometry with properly calibrated tonometer • Gonioscopy – Indentation/manipulation • Careful clinical evaluation of optic disc and retinal nerve fiber layer (RNFL) to pick up early signs of disc damage • Visual field (VF) by online field/ automated perimetry • Record central corneal thickness (Pachymetry) • Disc diagram/photo	• Can follow-up suspects – 6 monthly -with IOP <24 mm Hg, only suspicious discs or PACS with no risk factors • When glaucoma diagnosed, start glaucoma meds singly or in combination to assess efficacy and achieve Target IOP • Laser peripheral iridotomy (LPI) for all PAC/ PACG and high risk PACS • In mild / moderate glaucoma if Target IOP achieved & status quo on perimetry, follow-up at 6 months • If severe glaucoma review every 3 months • Surgery if IOP uncontrolled on glaucoma meds or if

		patient intolerant/ allergic to meds or is progressing

(iii) Tertiary care level (Glaucoma Specialist)

Equipment	Evaluation	Management/referral guidelines
• Slit-lamp • GAT • Gonioscope – 2/4-mirror • 90D/78D • Automated perimeter • Pachymeter (ultrasound or optical) • Fundus camera for disc photo with or without AI • Optical coherence tomogram (OCT) for evaluation of Optic nerve head (ONH), RNFL, Ganglion cell complex (GCC) • Nd:YAG laser • Green laser/SLT (Selective laser trabeculoplasty)	• Careful review of history & compliance to therapy • Comprehensive eye examination, look for any missed findings like secondary glaucoma/angle closure • Review the investigations and repeat in case of any doubt	• Monitor compliance/ diurnal variation of IOP • Recheck if Target IOP achieved • LPI for all PAC/PACG • Trabeculoplasty for open angle glaucoma • Low threshold for surgery Trabeculectomy or implants) for progression, socioeconomic reasons, availability of drugs/allergy, uncontrolled IOP • MIGS if expertise available • Cyclophotocoagulation in high-risk eyes or eyes with poor visual potential • If stable, can refer back to secondary center / primary center for regular monitoring

Childhood Glaucoma

Childhood glaucoma affects >300,000 children worldwide, often associated with significant visual loss (two-third of these children are blind).[1] There is a higher prevalence with more severe disease phenotype in the developing countries.[2,3] The prevalence of primary congenital glaucoma is 1 in 3300 as against 1 in 10,000 in the western population.[4] Glaucoma accounts for 4.2-5.0% of blindness in the population.[5,6] Key reasons for not not seeking early eye care are lack of awareness, distance from nearest hospital, low socioeconomic status and sociocultural beliefs.[7]

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Definition

Glaucoma can affect children, including newborns. Unlike adults, a childs eye responds differently to elevated intraocular pressure leading to changes in the size of the eyeball, corneal diameter and corneal clarity. For the diagnosis of a child with glaucoma atleast two or more of the following criteria are required,[8]

1. IOP > 21 mmHg on repeated testing

2. Optic nerve glaucomatous damage, such as increased cupping, focal notching, or cup-to-disc asymmetry of >0.2 between two eyes

3. Corneal changes/findings: corneal edema with, Haab striae or corneal diameter >11 mm in newborn, > 12 mm in child < 1 year of age, and > 13 mm at any age

4. A reproducible visual field defect consistent with glaucomatous optic neuropathy (in older children)

Categories : primary , secondary

Primary Congenital glaucoma (PCG) is a form of glaucoma occurring in children less than 2 years of age characterized by isolated trabeculo-dysgenesis with resultant abnormal development of trabecular meshwork. It is classified in the following categories:

1. **Primary glaucoma**

- Primary congenital glaucoma (PCG)
- Neonatal onset when present at birth to 1 month after birth
- Infantile onset when present with clinical features of PCG after 1 month up to 2 years
- Late onset: > 2 years with buphthalmos or corneal changes such as Haab striae
- Juvenile open-angle glaucoma (JOAG): >2 years of age, glaucoma in isolation from any acquired or non-acquired conditions, without ocular enlargement or any cornealchanges, and with a normal seeming angle.

2. **Secondary glaucoma**

- Glaucoma associated with non-acquired ocular anomalies (Anterior segment dysgenesis, Aniridia, etc)
- Glaucoma associated with nonacquired systemic disease or syndrome (ex: Sturge Weber Syndrome, Axenfeld Reiger syndrome etc)
- Glaucoma associated with acquired conditions (ex: Steroid induced, trauma, post corneal or vitreo retinal surgery)
- Glaucoma after congenital cataract surgery (Aphakia or pseudophakia)

In view of the ocular structural changes that can occur in a child with glaucoma, the severity of the condition is dependent on the following factors:

Table 1: **Proposed severity staging for PCG[9]**

Parameters	Non-severe PCG	Severe PCG*
Age of onset	Infantile	Neonatal
Corneal diameter (in mm)	< 13	> 13
Axial length (in mm)	< 24	> 24
Corneal haze	Clear cornea or anterior segment structures can be made out despite the haze	Hazy cornea with barely or not visible anterior segment structures

Diagnosis and Evaluation Protocol for Childhood Glaucoma

Evaluation protocol

(a) Primary care level (trained Optometrist)

Early diagnosis and timely referral for treatment are essential to prevent blindness in children.

Childhood glaucoma is relatively easy to diagnose due to its distinct clinical features such as enlarged eye, cloudy cornea, photophobia and excess tearing. . Primary eye care providers must be trained to recognize these signs, counsel the parents and family members and reassure them that early surgical intervention can significantly improve outcomes, helping the child to integrate into the society. Delayed diagnosis and treatment would lead to irreversible blindness.

In addition to screening for glaucoma, the diagnostic and evaluation protocol should include the following measures to prevent childhood blindness:

a. Retinopathy of Prematurity: Premature infants for Retinopathy of Prematurity (ROP) as per the national guidelines as it is very important to prevent secondary sequelae and loss of vision (broad eligibility criteria for screening are gestational age ≤34 weeks, birth weight ≤2000 gms)
b. Steroid induced glaucoma: To educate about avoiding topical and other forms of steroid eye drops in the eye or ointment over the skin in preventing steroid induced glaucoma and blindness.

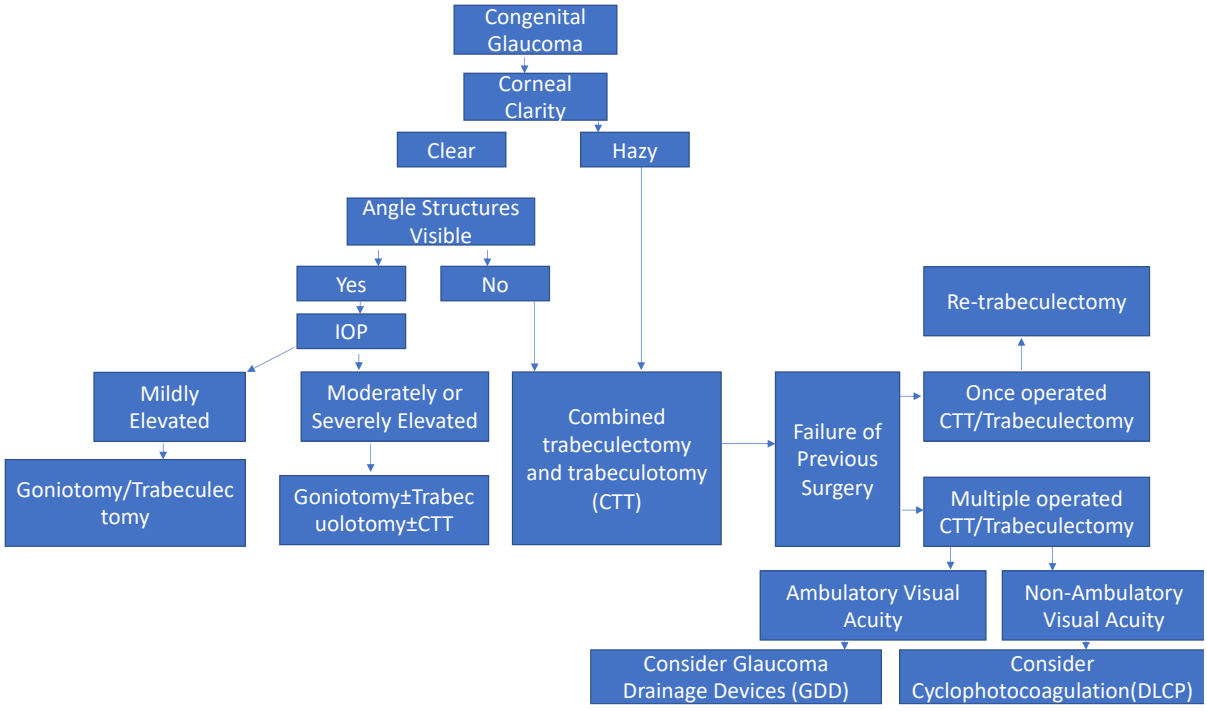
c. Leukocoria (white reflex): Although a rare, eukocoria in a child should be promptly recognized and referred for evaluation to rule out and manage sight and life threatening conditions like retinoblastoma.

d. Prevention of trauma: Families should be educated on preventing children from playing with sharp objects, as trauma is a major cause of vision loss and secondary glaucoma, particularly in India.

e. Chemical injuries: Raising awareness about accidental lime ("chuna") injuries ie essential, as they are a serious cause of bilateral blindness in children. Training primary care providers to administer first aid and emergency referrals can help mitigate this risk

f. Post-Surgical Monitoring: At the primary and secondary levels, follow up of post-surgical cases to detect any elevation in IOP is critical, especially following congenital cataract surgery, corneal or retinal surgeries.Educating caregivers and strengthening referrals can significantly reduce childhood blindness.

Figure: Management guidelines for congenital glaucoma



Recommendations for Glaucoma following congenital cataract surgery (GFCS)

Glaucoma following cataract surgery is a distinct concern due to its high incidence in children.rangeing from 3% to 65.6%. Glaucoma following cataract surgery can occur in both pseudophakic and aphakic eyes witonset often delayed by a median of 5 years . It is usually open angle type when the age of onset is delayed, whereas in early onset type it is more likely to be angle closure. Since openangle glaucoma is usually asymptomatic, life-long follow up is crucial to detect and treat the condition as early as possible.

Refer patients to Glaucoma specialist if

- There is progressive myopia or astigmatism
- IOP is > 21 mm Hg. In children in whom IOP cannot be measured look for slit lamp findings suggestive of raised IOP like hazy cornea, shallow ACD and iris bombe, etc.
- Presence of increased cup to disc ratio or characteristic glaucomatous optic nerve head changes.
- Note: It is highly recommended to include IOP measurement with Goldmann applanation Tonometry/ Perkins at the primary and secondary level eye care system.

Care and support for children with glaucoma:

Genetic testing and Counseling: Glaucoma is the second most heritable ocular condition.

Drawing a family pedigree is paramount to establishing the mode of inheritance. This mandates screening of at least 1st generation family relations. Genetic testing is not typically needed for ocular management of childhood glaucoma which is guided by phenotype. However, genetic testing and counseling will be needed to understand the genetic mutation in the child and to some extent predict prognosis and disease behaviour. Parental counseling regarding future pregnancies and possibilities of similar pathology in the siblings has to be clearly counseled. It is also possible to suggest and offer prenatal testing and diagnosis in future pregnancies.

Counseling and Education:

Method of enabling the patients or parents of a child with visual disabilities to choose the best fitting intervention for him by the interaction between trained professionals and patients or parents. Rehabilitation services provide a range of assessment and support services, including optometry, vision rehabilitation and advice for welfare benefits, in a 'one-stop shop'. The following aspects while providing counseling and education can be considered:

- Disease leading to a vision problem, prognosis, treatment, etc.
- Low vision and rehabilitation assistive devices
- Illumination amplification techniques: glare protection, lighting enhancement with adjustable gooseneck lamp, overhead lighting, wall mounted lighting, LED lamp, CFL with various brightness capacity, etc.
- Patient safety education: reducing risk of falls, or accidental mishaps related to fire, electricity, bathroom tile, sharp objects, cooking cylinder, etc.

- Environmental modification (home safety): arranging furniture, tables, desks, corridors, spacing in the central part of the house, or wall side as per needs
- Daily living activities include personal care, clothing, cleaning, bathing, brushing, eating, home management, financial management, shopping, etc.

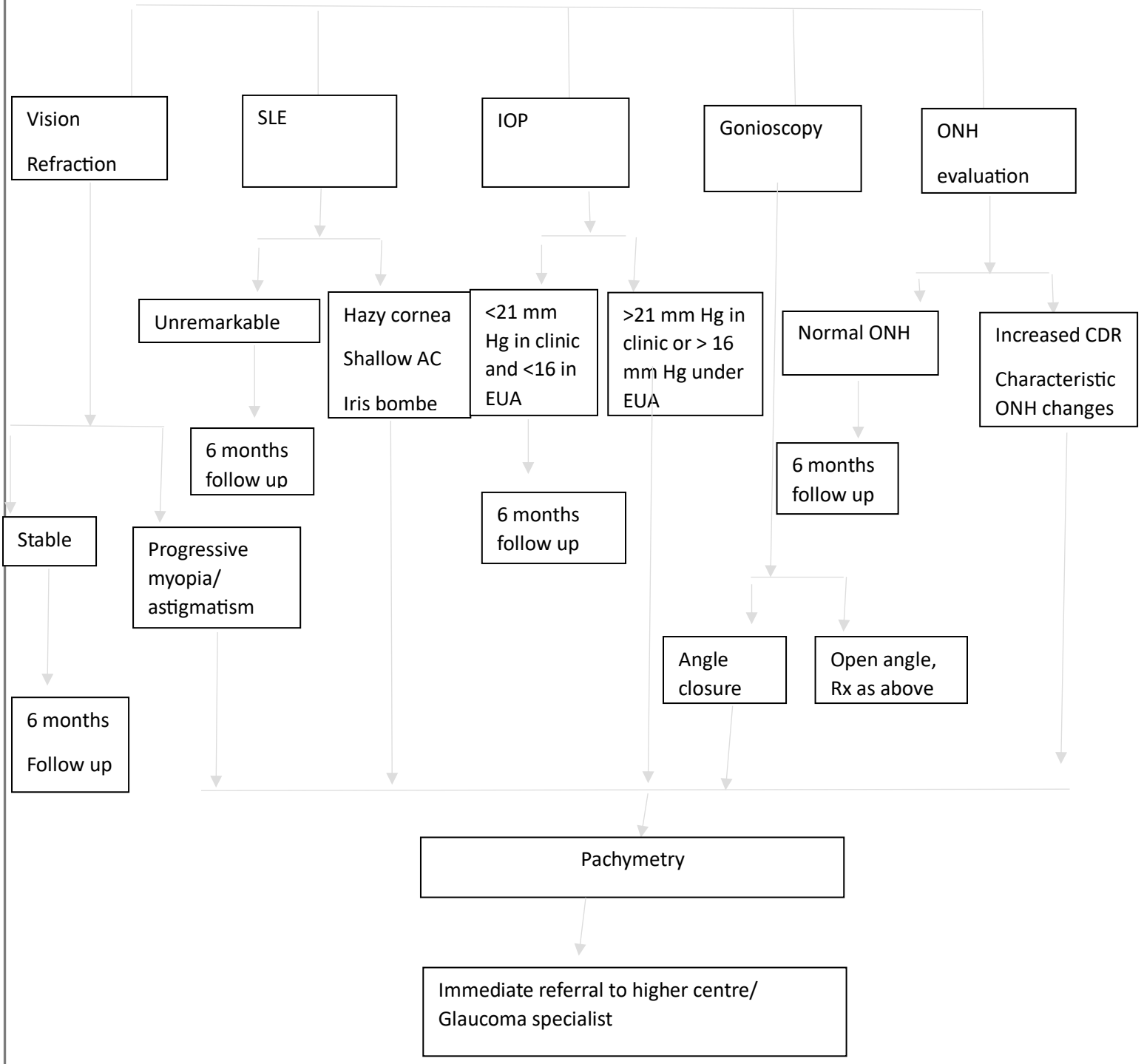
Evaluation of caregiver burden and appropriate counseling

Glaucoma following cataract surgery (GFCS)

AT THE PRIMARY AND SECONDARY OPHTHALMIC CARE CENTRES

For Glaucoma following cataract surgery (GFCS) or after any intraocular surgery

6 Monthly Check up



Primary Open Angle Glaucoma

Primary open angle glaucoma is a spectrum of conditions It can begin as raised intraocular pressure called ocular hypertension, which may progress in certain individuals with risk factors to a glaucomatous optic neuropathy. There is also a possibility that the optic neuropathy may develop with IOP <21mmHg, and this is termed Normal tension glaucoma (NTG) or low-tension glaucoma.

Introduction

The prevalence of primary open angle glaucoma (POAG) worldwide is 2.2%[1]. India accounts for 12.9% cases of POAG-induced blindness, with a prevalence of 2.7%-4.3%[2,3] cases of open angle glaucoma. Community and hospital data indicate that 75-90% of POAG remain undiagnosed.[4]

Need for screening

The asymptomatic nature of POAG leads to a delay in diagnosis and treatment. All adults coming for a presbyopia correction, those with family history of glaucoma, high myopia should preferably get a check-up including tonometry, Van Hericks test, slit lamp exam to rule out pseudo exfoliation/ pigmentary disturbance, iris clues to rule out angle closure element and an un-dilated disc evaluation with slit lamp biomicroscopy, with documentation with nonmydriatic/ mydriatic fundus photography andif possible integrated with AI.

People with systemic risk factors like diabetes, high systolic or diastolic BP,[5] dip in nocturnal blood pressure, family history of glaucoma, history of trauma, steroid use should be evaluated for glaucoma.

Mass screening is not possible in resource-constrained settings like India,[6] hence strategies and guidelines must be devised for early detection and timely management.

Evaluation protocol

Primary care level-

Can be done by Optometrist/ vision technicians/ Opticians trained in slit lamp examination, applanation tonometry and to acquire a good fundus photo– with facilities for consulting ophthalmologist directly or through tele-ophthalmology Secondary care level

Secondarylevel facilitiesallow the ophthalmologist to identify risk factors for glaucoma, and to confirm if it is POAG rather than trauma or steroid induced.

They also have the opportunity to determine diurnal variation of IOP and the extent of glaucomatous neuropathy. They can establish a target IOP and initiate treatment as per the flow chart below.

Optic nerve head evaluation preferably using a 90/78 D lens or by fundus or optic disc photography

Look for

a. Definite signs of Glaucomatous optic neuropathy (Hard Signs)

i. Excavation or Notching, a localized loss of the neuro retinal rim (NRR).
ii. Optic disc haemorrhage

iii. Retinal nerve fibre layer (RNFL) defect

b. Corroboratory signs (Soft signs)

i. Peripapillary atrophy in the region of NRR loss
ii. Nasalization of vessels
iii. Baring of circumlinear vessels (BCLV)
iv. Asymmetry in the cup disc ratio between the two eyes of > 0.2
v. Thinning or loss of the inferior NRR, against the ISNT rule

1. Perimetry

Reproducible defects corresponding to the optic disc changes, on more than one automated white on white perimetry having any of the following features, between 10 – 20 degrees from fixation, would be a definitive glaucomatous visual field defect.

- A cluster of 3 contiguous non-edge points in 30-2 (or even edged points in 24-2) significantly depressed, likely to be seen in < 5% of the normal age matched population.
- Glaucoma hemifield test 'Outside normal limits"
- PSD with a p<0.5%

- Adherence to be encouraged and follow up visits scheduled with local area ophthalmologist (wherever possible) or at the tertiary centre.
- **Education** regarding disease with emphasis on irreversibility of vision loss despite use of medication.

C : Tertiary care level

A glaucoma specialist at this level should review available history, investigations, and repeat or add examinations or investigations as deemed appropriate.Target IOP may need to be reassessed and changes in medications or a surgery if IOP is uncontrolled on maximal tolerable medications, or for socioeconomic reasons.

Follow up

- First follow up should be after 4-6 weeks to check response to treatment or occurrence of side effects. Repeat examination every 3-6 months if deemed as stable on treatment.
- At each follow up IOP and fundus evaluation to be done. Perimetry to be repeated after 6 months and then after 1 year in established glaucomatous damage.

- Gonioscopy will need to be repeated 1 yearly or when IOP is elevated to rule out a new cause or additional mechanism for IOP elevation
- If IOP control is inadequate, drug therapy to be substituted only after verifying compliance and systemic conditions.
- Any patient uncontrolled on maximal tolerated topical drugs or progression of disc damage with medication or unacceptable side effects to drugs/ non-affordability of medications, surgical option should be given
- Stable disease requires perimetry or OCT imaging every > 6 (8-9)months for first 2 years and then annually. Gonioscopy to be repeated annually.
- Advanced glaucoma / progression would need 3-6 monthly checks.

Management

Medical: Prostaglandins (PG) in most cases. Review efficacy and response after 4-6 weeks. If < 20% IOP drop, substitute with another PG. Review again after 6 weeks. If no response or if target IOP is not achieved- drugs need to be added one at a time, ensuring further reduction in IOP –B blocker/ ROCK inhibitors/ Alpha agonist/ Carbonic anhydrase inhibitors

If target IOP still not achieved -

- ✓ SLT can be done if poor compliance/ side effect/ allergy
- ✓ MIGS (angle based surgeries like ab interno-goniotomy GATT, KDB goniotomy/BANG, IStent etc) is a safer surgical option for early glaucoma and/or poor drug compliance/tolerance
- ✓ Surgical option - Trabeculectomy augmented with antifibrotics for poor IOP control, progression and non-compliance in moderate to advanced disease.

If trabeculectomy fails → encysted bleb may need early needling with 5FU/ MMC → wait for 1 month for response → if partial / no response → repeat 2nd time

If the bleb is flat/vascularized→ add AGM (Beta blocker/ Alpha agonist/ Carbonic anhydrase inhibitors), Rho kinase inhibitors maybe first drug in failing trab due to effect on bleb fibrosis (hyperemia side effect explained to patient) else 5FU injections/ bleb needling with MMC / laser suturelysis can be considered

Management Algorithm for POAG

Record/Note Highest baseline < 30/> 30mmHg

Document baseline IOP preferably with GAT, gonioscopic findings, pachymetry, define Target IOP, and baseline visual fields. Additional documentation of Diurnal variation, any associated dip in BP, systemic risk factors,

Start medical management Prostaglandins (PG) as first line therapy

Add drugs one at a time after ensuring further fall in IOP

If < 20% IOP drop, substitute with another PG. Review again after 6

If < 20% IOP drop, substitute with another PG. Review again after 6

If target IOP not achieved- 2nd drug add

- SLT can be done if poor compliance/ side effect/ allergy
- Surgical option - Trabeculectomy augmented with antifibrotics for younger than 50 y

If trabeculectomy fails → do early needling with 5FU/ MMC → wait for 1 month for response → if partial / no response → repeat 2nd time

If IOP still uncontrolled → add AGM (Beta blocker/ Alpha agonist/ Carbonic anhydrase inhibitors), Rho kinase inhibitors maybe first drug in failing trab due to effect on bleb fibrosis (hyperemia side effect explained to patient)

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Ocular hypertension

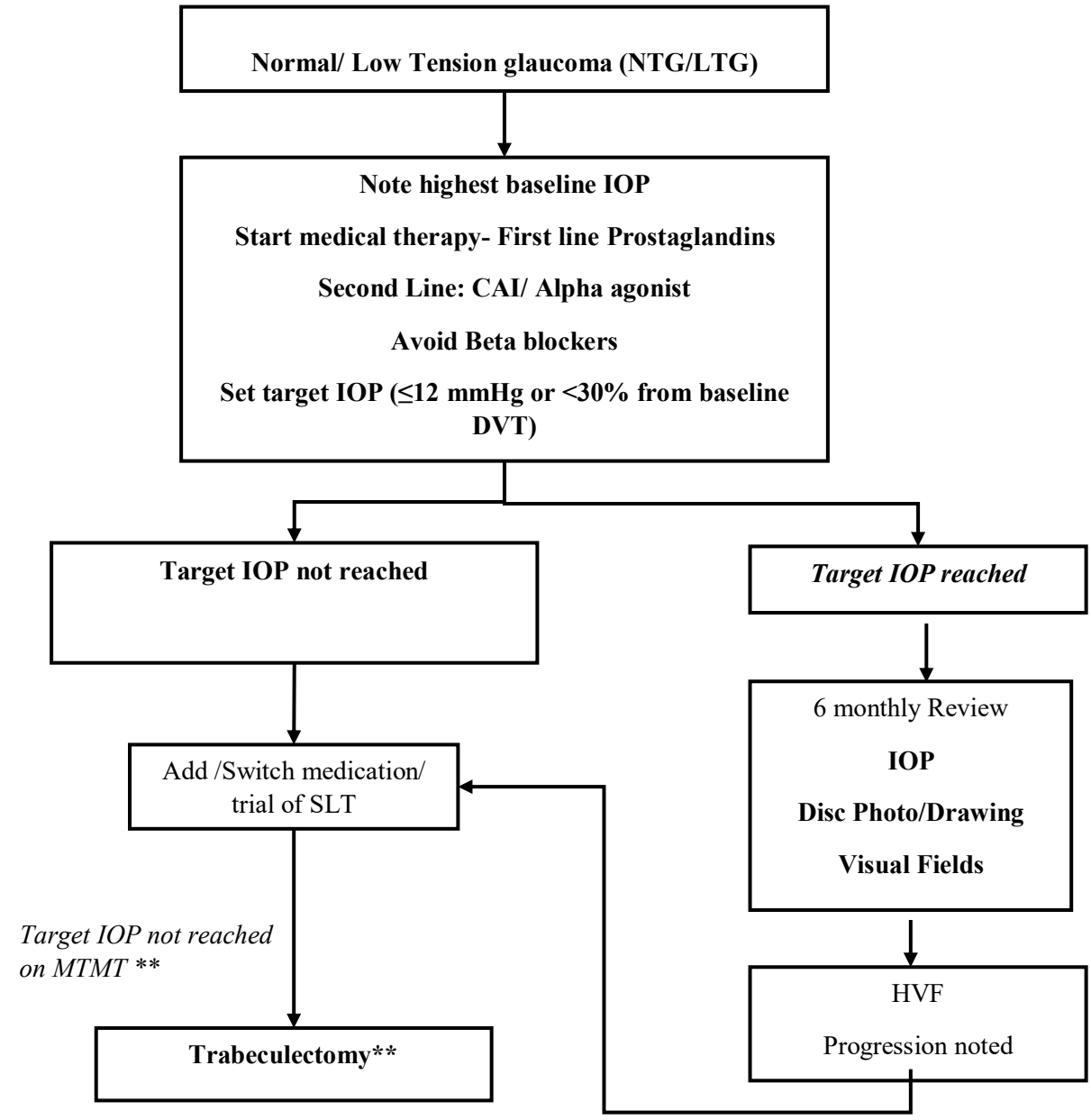
Definition:

Ocular hypertension is generally defined as: IOP>21 mm Hg in one or both eyes, open angles on gonioscopy, normal optic disc and RNFL, normal (non-glaucomatous) visual fields and no apparent systemic or local cause to explain the high IOP (like steroid use, etc.).

Screening:

Measuring IOP is not an effective method for screening populations for glaucoma. Population-based studies suggest that half of all individuals with OAG have IOP levels below 22 mmHg, the usual screening cutoff.[1] Additionally, most individuals with elevated pressures at a screening measurement do not have, and may never develop, optic nerve damage, although risk increases with higher IOP.

Evaluation protocol: Tertiary care level



***Selective laser trabeculoplasty** can be added/ done at any stage.

****Indications of Trabeculectomy**

- IOP not controlled on maximum tolerable medical therapy (MTMT)
- Progression despite maximal medication
- Poor tolerance to medications
- Not able to afford medications

 # Systemic evaluation

- Blood pressure diurnal
- Sleep Apnea
- Carotid Doppler
- Hyperlipidemia, Hyper Homocystenemia, Autonomic neuropathy (Vit B12 deficiency)

**MTMT= Maximum tolerable medical therapy

Flowchart

SCREENING

PRIMARY LEVEL:

OPTOMETRIST — OPTICIAN / PHC

SLE/ GAT-IOP/ OPTIC DISC Photo

FAMILY H/O GLAUCOMA in Parents/ Siblings

SUSPICION OF GLAUCOMA

SECONDARY CARE: OPHTHALMOLOGIST / DISTRICT HOSPITAL

DETAILED HISTORY

SLE /GAT IOP / INDENTATION GONIOSCOPY / VISUAL FIELDS / OCT (RNFL & GCA)

INITIATION OF MEDICAL THERAPY

REFERRED TO TERTIARY CENTER

TERTIARY CARE CENTER

- DETAILED HISTORY
- COMPLETE EYE EXAM
- SPECIALISED TESTS: VFA /OCT / OCTA / ASOCT / GENETIC TESTING + COUNSELING

INITIATE THERAPY: MEDICAL / LASER / SURGICAL INTERVENTION

COUNSELING AND REGULAR FOLLOWUP

SKILL DEVELOPMENT AT SECONDARY AND PRIMARY CARE LEVELS

Primary Angle Closure Disease

PACD accounts for approximately 25% of all glaucomatous optic neuropathy worldwide, but responsible for 50% of blindness caused by glaucoma.[11] The Population-based studies have shown that fewer people with PACD in a rural setting are likely to be aware of their condition in comparison with those in an urban setting.[12]

Definition:

Primary angle closure is characterized by appositional or synechial iridotrabecular contact at the anterior chamber angle caused by pupillary block1 and/or due to the crystalline lens, with no apparent contribution from any ocular or systemic diseases. The most common mechanism is "relative pupillary block", which increases resistance to aqueous flow from the posterior to the anterior chamber. This creates a pressure gradient that pushes the peripheral iris forward, leading to appositional angle closure and obstructing all or part of the filtering portion of the trabecular meshwork (TM). Prolonged or repeated contact of the peripheral iris with TM leads to peripheral anterior synechiae (PAS) anddamage to TM.2. The condition is diagnosed after ruling out other causes of angle closure such as uveitis, lens induced, trauma etc.

Standard Terminology and Classification:

Primary Angle Closure Disease (PACD) is the terminology used to cover the entire spectrum of disease, including PACS, PAC and PACG. The International Society for Geographical and Epidemiological Ophthalmology (ISGEO) classification[3] system classifies the disease based on the extent of the angle visualized on gonioscopy, presence or absence of peripheral anterior synechiae (PAS), intraocular pressure (IOP) elevation, optic nerve head (ONH) changes and visual field changes into the following three categories (Table 1). The basic clinical picture in PACD described by ISGEO being the inability to visualize the posterior TM over more than 180 degrees with the patient looking in primary position. This is considered by the WGA (World Glaucoma Association) to represent iridotrabecular contact.

- Primary Angle Closure Suspect (PACS)- Anatomically narrow angles/angles at risk
- Primary Angle Closure (PAC) - Presence of either structural damage (PAS) or functional
  damage (IOP elevation).
    - PAC can sometimes present in an acute form called acute primary angle closure (APAC) or acute congestion glaucoma, which is an ophthalmic emergency
- Primary Angle Closure Glaucoma (PACG)- structural and functional damage as in PAC+ with glaucomatous ONH changes and corresponding field changes.

Table 1: Definition of PACS, PAC and PACG (WGA)

	AC/ gonioscopy	Optic nerve head	IOP	Perimetry
Primary angle closure suspect (PACS)	Iridotrabecular apposition ≥180	Normal	Normal	Normal
Primary angle closure (PAC)	Evidence of iridoctrabecular contact such as goniosynechiae, PAS blotchy pigmentation Pupillary sphincter atrophy, Glaucomflecken	Normal	Normal to high	Normal
Primary angle closure glaucoma (PACG)	Extensive Iridotrabecular contact	C:D> 0.7/ asymmetry > 0.2/ NRR loss	Variable/ >21mmHg	Visual Field defects

Why is screening for PACD important?

Though PACD is less prevalent than primary open-angle glaucoma (POAG), affecting approximately 24 million people in India.[4] it is responsible for half of the glaucoma-related blindness in India.[5]

Risk Factors:

Anatomical Risk Factors:[6]

i. Short axial lengths
ii. Shallow anterior chamber (AC),
iii. Small corneal diameters
iv. Hyperopia
v. Thick, relatively anteriorly positioned lens
vi. Increased Lens Vault

Age, Gender and Ethnicity

i. Advancing age is a risk factor as there is thickening and anterior movement of the lens

which can result in pupillary block.[7] It is more commonly seen in 5th to 6th decade of life

ii. Prevalence of most forms of angle closure is 2-5 times higher in women than in men,

which may be part ly attributed to woman having shallow anterior chamber .[8] However Chronic PACG occurs almost equally in both females and males.

iii. More common in Asians and Eskimos

Family History

PACD can be familial.

i. A study from India has shown a 37% risk of PACG among first degree

relatives of patients with PACG.[9]

ii. The risk of developing angle closure in siblings of patients with angle closure is 1

in 3, around 35%. The risk is more in older and female siblings.[1]

Evaluation protocol

Primary care level (trained Optometrist)

Opticians / PHC providers may lack the skills needed to detectorscreen for glaucoma. However, in the presence of a family history of glaucoma in parents / siblings they should refer them to the nearest ophthalmologist for screenin

Primary Care (PHC/ Optometrists)

These clinical guidelines for primary ophthalmic care in angle closure disease describes appropriate screening, identification and timely referral to prevent visual morbidity caused by primary angle closure disease (PACD). This guideline will assist opticians, technicians, optometrists and doctors at Primary care in achieving the following goals

1. Identify people who have PACD (Primary angle closure disease) and those who are risk to develop PACD. Raise awareness in families.
2. Assess risk factors from patient history.
3. Manage a patient who has acute angle closure attack.
4. Monitor and follow up patient with PACD.
5. Educate the patient about visual complications of PACD, treatment options and the prognosis.

Screening for angle closure glaucoma

A relatively quick, non-contact screening tests can be carried out by appropriately trained healthcare professionals or technicians as a triage test to identify people at risk of angle closure and those with suspicious discs on non-mydriatic fundus camera. If the test is positive, the person should be referred to higher centre for further glaucoma specialist evaluation and management. The risk factors of angle closure that can be identified at the primary level arealready decribed above.

The symptoms that point towards risk of angle closure are

1. Frequent headaches, nausea or vomiting
2. Eye pain, photophobia, congestion in the eye
3. Coloured haloes around light

Screening tests: Two important screening tests that can be performed to identify the risk of angle closure are oblique flash light test[13,14] and Van Herick test.[15,16,17,18]

Oblique flashlight test

The flashlight test is an accessible method to detect a potentially occludable angle if no other equipment is available. The test involves shining a pen torch into the eye from the temporal limbus parallel to the iris to assess the anterior chamber depth.

Grade 1 is associated with a high risk of angle closure and should be referred to higher centre for further evaluation and laser peripheral iridotomy (LPI) if needed.

Grading uses a four-point scale, based on the proportion of the nasal iris that is in shadow.

Grade	Shadow
Grade 1	<1/3 illuminated-nasal iris in complete shadow
Grade 2	1/3 to 2/3 illuminated
Grade 3	>2/3 illuminated
Grade 4	Fully illuminated-no shadow

A referral to glaucoma specialist should be done if any one of the following features are present Grade 1 shadow in Oblique flashlight test
With or without one of the following

1. Symptoms and signs suggesting acute/PAC (intermittent) angle closure.
2. Best corrected visual acuity worse than 20/40
3. IOP higher than 21mmHg
4. Disc suspect on fundus photography with non-mydriatic fundus camera

Evaluation protocol

When the screening test is positive a detailed history and clinical examination can help in the diagnosis and appropriate referral to secondary or tertiary centre for further management.

Patient History

A thorough attention should be paid to elicit symptoms that may suggest prior angle closure attacks. These symptoms include blurred vision, transient loss of vision, mild to severe eye pain, photophobia, congestion, coloured haloes around light and headache with nausea or vomiting. These episodes are often relieved by sleep or exposure to bright light. Checking for a family history of angle closure is also essential.

Clinical examination

- **Refractive status**: Check for refractive error whenever possible.
- **Pupil examination**:
- Note sze and shape (mid dilated, irregular, asymmetric or oval in acute angle closure),
- Check if the pupil reacts sluggishly or Is fixed
- Look forRAPD (may be present in asymmetric optic nerve damage)

Diagnosis

At the primary level it is possible to identify primary angle closure disease even if the precise stage is not identified. The importance is to screen patients at risk for blinding glaucoma

Counseling

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After the screening and initial examination, primary health care providers should provide patients and caregivers with clear, balanced information about:

- The nature and prognosis of the disease.
- The importance of visiting a glaucoma specialist for further evaluation and treatment.
- Possible treatments, including laser peripheral iridotomy and anti-glaucoma medications if recommended.
- The need for lifelong follow-up.
- The low risk of angle closure associated with certain medications (e.g., pupil dilation for fundus exams, antidepressants, anticholinergics).

Patients should also be informed about the genetic risk of glaucoma and encouraged to have their family members screened. Effective counseling can greatly improve the quality of life for glaucoma patients.

Therapy

Once a provisional diagnosis of PACD is made, the patient should be referred to nearest Ophthalmologist for further evaluation and LPI

Secondary care (District Ophthalmologists, General Ophthalmologists)

Objectives

1. To Screen all patients presenting for ophthalmic care (Opportunistic screening) for PACD
2. Diagnose and categorize PACD so identified into PACS, PAC, PACG
3. Identify who would require laser peripheral iridotomy
4. Assess those who would require a referral to tertiary care Institutions for glaucoma surgery

Screening for PACD:

Whom to screen?

- Any person above 40 years at the clinic /hospital for any eye ailment
- Any person complaining of intermittent coloured haloes and blurring of vision or headache.
- Any person with a family history of glaucoma
- All persons with IOP > 20 mm Hg using non-contact/applanation tonometer

How to screen

a. History:

Patients can be asymptomatic when presenting for regular eye check-up, or may present with sudden onset of symptoms like pain, redness, congestion, decreased
vision / frequent change of glasses and even unilateral headache and vomiting which points to the diagnosis of acute/ intermittent angle closure.

- Ask about symptoms suggestive of previous episodes of intermittent angle closure (e.g., blurred vision, halos around lights, eye pain, headache, eye redness, symptoms following stress or dilated eye examination).
- Ask for a family history of glaucoma. All first-degree relatives should be screened for
  glaucoma.
- Highest baseline intraocular pressure, before any treatment
- Systemic and ocular medications being used.
- Rule out secondary causes especially when bilateral by questioning regarding use of oral medications that causes ciliary body edema (e.g., sulfonamides, topiramate). Topical, inhaled, or oral drugs with adrenergic or anticholinergic effects (e.g., ipratropium bromide and salbutamol-containing inhalers, phenothiazines) can cause angle narrowing and potentially precipitate an angle-closure attack.

b. Examination

- Torchlight examination
  - Nasal shadow on shining light from the temporal side
  - Any obvious pupillary abnormalities
  - Any afferent pupillary defect

Van Herick grading and Limbal anterior chamber depth (LACD)

It is a quick, simple noncontact when gonioscopy is not possible, for screening angle closure in a resource-poor setting. This method involves a slit-lamp based comparison of the peripheral anterior chamber depth and the thickness of the cornea. The illumination column

should be offset from the axis of the microscope by 60°, objective magnification is set to 1.6X, and the brightest, narrowest possible vertical beam of light is directed at the temporal limbus, perpendicular to the ocular surface, and viewed from the nasal aspect. The beam is positioned at the most peripheral point of the cornea allowing a clear view of the AC and peripheral iris.

The ACD is then graded as a fraction of the thickness of the adjacent cornea in the following categories (Table 2)

Table 2: Van Herrick's Grading

Van Herick Grade	Fraction of thickness of adjacent cornea	
Grade 1	<1/4	Gonioscopy should be performed
Grade 2	1/4	Gonioscopy should be performed
Grade 3	1/4 -1/2	Incapable of closure
Grade 4	≥full thickness of peripheral cornea	Incapable of closure

1. Those patients who are grade 1 and grade 2 Van Herick are at risk of angle closure and gonioscopy should be performed before dilation. They need to be referred to higher centre for gonioscopy, detailed evaluation and LPI if needed.
    - Intraocular pressure (IOP) measurement using Goldmann applanation tonometer.
    - Comprehensive ophthalmological examination on slit lamp Biomicroscopy
      Look for
        - Peripheral Anterior chamber depth- Van Hericks grading
        - Iris pattern loss- any patchy atrophy, whorling
        - Any iris sphincter atrophy
        - Configuration of pupil- whether vertically oval
        - Any pigment on anterior lens surface
        - Status of lens, which will aid in deciding the surgical management

    - Optic nerve head evaluation preferably using a 90 D/78D lens or fundus photograph
      Look for
        a. Definite signs of Glaucomatous optic neuropathy (Hard Signs)

i. Notching, a localized loss of the neuro retinal rim (NRR).
ii. Optic disc haemorrhage
iii. Retinal nerve fibre layer (RNFL) defect

b. Corroboratory signs (Soft signs)

i. Peripapillary atrophy in the region of NRR loss

ii. Nasalization of vessels

iii. Baring of circumlinear vessels (BCLV)

iv. Asymmetry in the cup disc ratio between the two eyes of > 0.2

v. Thinning or loss of the inferior NRR, against the ISNT rule

vi. Optic Nerve Head pallor can follow an acute attack

- Gonioscopy for visible angle structures and angle width
  - Can be performed using a 2 mirror or preferably 4 mirror with indentation /manipulation
  - Should be performed in primary gaze with the patient looking at a far object to avoid accommodation induced miosis
  - Room illumination should be dimmed and use 1 mm beam at its brightest level.
  - Avoid crossing the pupil or else will cause miosis and false opening of the angles
  - Extent of the iridotrabecular angle in degrees, structures visible in primary gaze before and after indentation/manipulation, presence of PAS, iris contour and amount of pigmentation should be assessed; based on which the disease is graded

2. Once a patient is diagnosed as PACD clinically, a optic disc evaluation and visual field test will confirm whether it is PACG. If available, standard automated perimetry must be done in all patients.

3. Perimetry

Reproducible defects corresponding to the optic disc changes, on more than one automated white on white perimetry having any of the following features, between 10 – 20 degrees from fixation, would be a definitive glaucomatous visual field defect.

- A cluster of 3 contiguous non-edge points significantly depressed, likely to be seen in < 5% of the normal age matched population.
- Glaucoma hemifield test 'Outside normal limits"
- PSD with a p<0.5%

Differential Diagnosis:

Primary angle closure disease tends to be bilateral but may be asymmetrical. The presence of a wide angle in the fellow eye would suggest a secondary cause such as neovascular glaucoma, uveitic glaucoma, subluxated lens, intumescent lens, intraocular tumours and if there was prior surgery, like trabeculectomy, pan retinal photocoagulation, scleral buckling, intraocular gas or silicon oil etc, aqueous misdirection syndrome or pupillary block should be ruled out. (Ultrasound B scan and biomicroscopy evaluation may be needed)

Management

The aims of the treatment of PACD

i. Reversal or prevention of further angle closure

ii. Controlling the IOP to a target level that minimizes further optic nerve damage and vision loss

The treatment modalities include

- Lasers
- Medical Management
- Surgical management

Laser peripheral iridotomy is the first line of treatment for PAC and PACG, for relieving the pupillary block and preventing an attack of acute angle closure. Once the pupillary block is relieved the management of glaucoma depends upon the IOP, extent of angle closure, extent of optic nerve head damage and affordability and tolerance of medications. If IOP is not controlled on maximal tolerated medications, they should be scheduled for a trabeculectomy. Early surgery for a visually significant cataract is also beneficial, when IOP is controlled. This can be performed both in secondary and tertiary centers with appropriate preoperative precautions.

Laser Peripheral Iridotomy

Neodymium:yttrium-aluminium-garnet (Nd: YAG-1064 nm) is the laser commonly employed for LPI.

Indications of laser iridotomy in PACD:

1. All Primary angle closure glaucoma- PAC & PACG
2. Fellow eyes of patients having PACG, APAC, PAC
3. PACS with risk factors.
    a. Family history of PACG
    b. Frequent dilation required, as in diabetic retinopathy
    c. Non-compliant patient, not willing to follow-up, eyes with small discs at risk of NAION
    d. Difficult access to laser

Counseling:

- Patients and caregivers should be counselled about the chances of progression from PACS and PAC to PACG
- Patients should be explained about the treatment modalities; and should be encouraged to receive the same in a timely manner
- Hereditary nature of the disease should be discussed and all the first-degree relatives should be screened.

Obtain informed consent from the patient after discussing the risks, benefits, and expected outcomes if laser iridotomy is planned. Patient has to be explained that there would be photophobia immediately after the procedure. The patients need to be counselled that more than one sitting may be required. There can be transient rise of intraocular pressure which can be managed by giving aqueous suppressants like Timolol eyedrops. Rare possibility of Hyphema (specially in those with anticoagulants) needs to be explained.

<u>Technique of laser peripheral iridotomy:</u>

- Use pre-laser Pilocarpine 2% to facilitate laser iridotomy
- Use topical brimonidine perioperatively to prevent sudden IOP elevation, particularly for patients who have severe disease
- Ensure the patency of the iridotomy by directly visualizing gush of aqueous flow and pigment release from the posterior to the anterior chamber and also by visualizing the anterior lens capsule. Visualization of a red reflex alone is insufficient to confirm patency. Ensure the size of iridotomy is adequate (>200 microns, ideally 500 microns)
- Perform at least one IOP check immediately prior to laser and within 30 minutes to 2 hours
- following laser surgery

Common complications include iritis, hyphema and IOP spikes

- LPI can be challenging in APAC, IOP should be brought down with all possible measures to facilitate LPI after miosis (Flow chart 2)

Follow-up after laser iridotomy (Table 3)

LPI is not fully protective against the development of chronic angle closure in all patients. The patient should be continued on his previous anti-glaucoma medications along with an additional antiglaucoma agent for at least one week after the laser iridotomy. A topical steroid (4 times a day) should also be given for 5-7 days.

Patients are seen at intervals of one hour, one week, one month, three months after the procedure or as needed based on the IOP control.

- Follow-up evaluations in the days and weeks after laser should include the following elements:
    - Confirm the patency of the iridotomy by visualizing the anterior lens capsule or ciliary process
    - Corneal status, IOP, AC reaction
    - Perform dark-room gonioscopy with compression/indentation to assess the extent of PAS

Table 3: Follow up schedule after Laser Iridotomy

	Stage of PACD	**Follow up frequency**	**Referral to higher centre**
1	PACS/PAC with normal IOP and open angles post LPI	IOP every 6 months	Annual referral to reassess optic disc, IOP and visual field
2	PACS/PAC with residual occludable angles, Plateau iris configuration post LPI	Needs IOP and Gonioscopy every 6 months to monitor development of synechiae	if uncontrolled Interim referral if IOP >21mmHg
3	PACG	Target IOP evaluation Review every 4 months Annual Visual field examination	Annual referral to reassess optic disc, IOP and Visual field progression. Interim referral if IOP > Target set with AGM Intolerance or allergy to AGM Significant cataract Decrease in BCVA>20/40

What if the IOP is persistently elevated after a patent iridotomy?

Following iridotomy for PAC, persistent elevations of IOP may occur for several reasons:

- Trabecular damage or formation of PAS may have occurred due iridocorneal apposition prior to laser
- If the iridotomy becomes occluded, pupillary block may recur. Repeat laser is indicated.
- Factors other than pupillary block may lead to angle closure and may have gone unrecognized until after the iridotomy. These include plateau iris syndrome, phacomorphic angle closure, and secondary causes of pupillary block.
- Angle closure may have been superimposed on pre-existing open-angle glaucoma or on another cause of IOP elevation, such as steroid use, or exfoliation syndrome (Mixed mechanism glaucoma)

A lifetime of regular follow-up is to be stressed upon, the frequency of which will vary depending upon the status of glaucoma and IOP as for all glaucoma.

What if the IOP target is not met despite laser iridotomy and medications?

This is often seen among PACG patients with advanced glaucoma and they usually require a Filtering surgery with or without a cataract extraction. It would be desirable for surgery to be performed at the secondary level if the expertise is available, but the patients should be referred to a tertiary centre as soon as possible if care cannot be provided at secondary level.

Indications for Glaucoma Surgery (Trabeculectomy/ Phacotrabeculectomy)

- IOP above 'target' despite maximally tolerated medical therapy
- Inability to review regularly
- Unable to afford medications
- Progression of the disease
- Non-compliance with medical treatment

An overview of the Management protocol is shown in the Flowcharts 2-4

Medical Management

Medical therapy is usually employed to reduce the residual elevated IOP following LPI in PAC and PACG. Controlling the IOP is a must to prevent or delay ONH damage. Post LPI these eyes are treated with topical antiglaucoma medications similar to eyes with POAG. Moreover, medical therapy is essential in eyes with APAC to bring down the IOP and facilitate LPI. Ocular and systemic side effect profile should be considered before initiating medical therapy

- First-line of management includes topical Prostaglandin Analogues (PGA). And they have been shown to be effective even in eyes with 360 degree PAS.[19]

- Topical beta blockers can be considered in patients who cannot afford PGA
- Pilocarpine 2 % or as a combination with Timolol may be very effective in such eyes.
- Post LPI occludable angles constitute plateau iris syndrome / configuaration and respond well to pilocarpine eye drops or peripheral iridoplasty.
- Like in POAG topical alpha agonists and carbonic anhydrase inhibitors constitute second-line
  of medications
- Rock inhibitors can be added as a third line drug to buy time for surgical intervention

Tertiary care (Medical Colleges, Speciality Institutions)

The initial protocols of screening, case detection, diagnosis and management at the tertiary are level are as that detailed in the secondary care management. In addition to the clinical evaluation and laser iridotomy and trabeculectomy, the tertiary care centre will evaluate more complex diseases requiring specialized care.

- A repeat gonioscopy is essential to confirm the diagnosis and extent of TM damage.
- OCT may help identify glaucomatous ONH changes early, as well as preogression in PACG
- Anterior segment Imaging Techniques- Anterior segment optical coherence tomography (ASOCT) and ultrasound biomicroscopy (UBM) are useful to understand the pathophysiology of angle closure. Both are useful tools for screening angle closure[20]; can complement gonioscopy.
  but cannot replace it currently. AS-OCT helps delineate the lens vault (LV) and helps decide whether lens extraction will be required and will help in the management
  These tools are also helpful to differentiate certain forms of secondary angle closure. UBM is useful to assess lens and ciliary body and helps to rule out secondary causes of angle closure.

Complex Situations in PACD

Plateau iris

In Plateau iris configuration, there is typically crowding of the angle with anteriorly placed ciliary process, pushing the peripheral iris anteriorly. This results in elevated IOP and it is termed as plateau iris syndrome. In this condition, performing peripheral iridotomy does not eliminate the pupillary block. IOP typically rises after dilation due to greater occlusion of angle. Post LPI gonioscopy will reveal a typical double hump (caused by lens and ciliary process) or sign wave configuration. UBM is helpful in diagnosing plateau iris.

Acute Primary Angle Closure

APAC is an ophthalmic emergency, as the patients present with acute symptoms of pain, redness, headache, nausea and vomiting. Examination will reveal a congested eye with severe epithelial edema, shallow chambers, there can be minimal cells and flare in the anterior

chamber. Pupil will be typically mid dilated, vertically oval. Lens might reveal glaucomflecken. Gonioscopy of the fellow eye is a must and usually will reveal narrow/occludable angles.

Acute angle closure should be differentiated from the conditions shown in Table 4

Table 4: Differential Diagnosis for Acute Primary Angle Closure

Compromised angles with raised IOP
• Neovascular glaucoma
• Malignant glaucoma
• ICE syndrome
• Plateau iris syndrome
• Suprachoroidal effusion (drug induced)
Open angles with raised IOP masquerading as APAC
• Glaucomatocyclitic crisis
• Herpes simplex keratouveitis
• Pigmentary glaucoma
• Exfoliative glaucoma
• Post-traumatic glaucoma
• Steroid-induced glaucoma

Medical therapy of APAC is initiated with oral acetazolamide, IV Mannitol, topical betablockers and alpha agonists to lower the very high and often unrecordable IOP. After 30 -45 minutes pilocarpine 2% can be added every 6 hours to constrict the pupil. Topical and oral glaucoma therapy is continued until the corneal edema resolves and an iridotomy as detailed above is performed.

Argon Laser Peripheral Iridoplasty (ALPI)

ALPI can be considered in medically unresponsive cases of APAC and also in plateau iris syndrome (PIS). It flattens the peripheral iris and opens up the angles. Despite the advantage of the procedure in altering angle morphology, randomized control trials did not show any benefit over medical management.[21] Hence, it is reserved for resistant cases of APAC and PIS.

Surgery

Surgical management is considered after LPI and if the IOP could not be controlled adequately with these measures or if the patient develops cataract.

Pre-operative Considerations:

- Before any incisional surgery it is always good to perform LPI, which might help in the chamber stability during the surgery and might help reduce the chances of aqueous misdirection
- Bring down the IOP as much as possible with topical AGMs and oral CAI
- It would be good to stop Pilocarpine (if the IOP management allows), to facilitate pupillary dilation. Moreover, pilocarpine can disrupt the blood aqueous barrier increasing the chances of inflammation

Intra-operative Considerations:

- Intravenous mannitol administered in the dose of 1-2 mg/kg body weight helps by deepening the anterior chamber by reducing vitreous volume
- Administer minimal peribulbar block
- Avoid tight speculums in these narrow hypermetropic eyes to avoid positive pressure
- Use air/ high density viscoelastic substances as they help to deepen the chamber as well as protect the corneal endothelium
- Wound integrity and chamber formation should be ensured at the conclusion of the surgery
  as post-operative shallow chamber might increase the chances of aqueous misdirection

Choice of Surgery
A. Trabeculectomy

Trabeculectomy in PACG is more challenging than in eyes with POAG owing to the possible

intra-operative and post-operative complications including a shallow AC, expulsive haemorrhage and aqueous misdirection. Surgical iridectomy should always be performed while doing trabeculectomy in

- PACD. The use of controlled decompression of anterior chamber and care must be taken to
  prevent intra-operative and post-operative hypotony
  Releasable sutures allow maintenance
- It is good to use cycloplegia immediately after the surgery, on the operating table

In APAC, trabeculectomy is considered either alone or in combination with lens extraction if the IOP control is not achieved with conventional medical and laser therapy (Flow chart 2).
However, it has a poor prognosis, for long term function in acutely inflamed eyes, and should be done after congestion subsides[26]

In chronic PACG eyes with uncontrolled IOP and moderate VF loss trabeculectomy alone can be considered, especially if there is no cataract (Flow Chart 3)

B. Combined Trabeculectomy and Cataract surgery

- Phacotrabeculectomy is usually considered in eyes with PACG having co-existing cataract and
  medically uncontrolled IOP.
    - This surgery has the advantage of reducing IOP and offering visual rehabilitation in the same sitting.

- It also has the advantage of deepening the chambers and widening of the angle recess. Can be considered in eyes with APAC after the eye becomes quiet as most of these eyes have lens opacities.

C. Cataract Extraction

Cataract extraction is beneficial in eyes with PACD as increased lens thickness, lens vault contributes to the disease significantly. In PACD, the AC depth increases after cataract extraction and IOL implantation. Increase in AC depth & angle width relates to the exchange of thick cataractous lens

(Avg-5mm) with a thin IOL (Avg-1mm). Cataract surgery alone can be considered in eyes with visually cataract co-existing in PACS or PAC. In PACG eyes with mild to moderate damage with IOP controlled with one or two AGMs, we can consider cataract surgery alone.

Points to keep in mind

- PACD eyes are often hypermetropic with narrow palpebral fissure and shallow anterior chambers
- Can have posterior synechiae, rigid pupil leading to poor intraoperative mydriasis
- Can have compromised endothelium
- High risk of all possible intra and postoperative complications and demands surgical expertise

<u>Evidence regarding lens extraction in PACD Glaucoma</u>

The EAGLE[23] study randomized PACG patients to LPI vs. Clear lens extraction (CLE). CLE showed IOP lowering efficacy of only 1 mmHg over iridotomy. Considering the moderate IOP reduction and various limitations of the study we need more RCTs to decide on clear lens extraction in PACD.

Acute intervention with lens extraction as primary treatment in APAC remains less appealing except in resistant cases due to potential surgical complications that can be avoided by conventional treatment. An elective approach to the lens can be considered, if indicated later. [25] However early lens extraction in the presence of cataract is indicated and would be beneficial.

D. Goniosynechialysis

This involves stripping mechanical stripping of PAS from TM, using viscoelastics or cyclodialysis spatula. A metaanalysis has shown significant benefit.

E. Glaucoma Drainage Implants

These are usually considered as a last resort when there is disease progression even after trabeculectomy and cataract extraction. It is good to avoid in phakic eyes as there is a danger of lens touch and corneal touch due to shallow chambers.

F. Cyclophotocoagulation

The available procedures include

- Transscleral Cyclophotocoagulation
- Micro pulse-transscleral cyclophotocoagulation
- Endocyclophotocoagulation

Transscleral diode cyclodestructive procedures are usually performed when the eye has little visual potential. However, with the recent advances in the technology and the availability of micro pulse-transscleral cyclophotocoagulation it can be tried earlier in phakic eyes with high risk of incisional surgeries.[28]

G. Minimally Invasive Glaucoma Surgeries

While minimally invasive glaucoma surgery (MIGS) has become an important surgical approach for primary open-angle glaucoma, its indications and benefits in PACG are less clear.

Available evidence suggests that cataract surgery in combination with MIGS can be safely and effectively performed in PACG. However, the supporting evidence is largely based on retrospective studies and some small RCTs. Large scale, multicentered RCTs are needed to assess the effectiveness of MIGS in PACG, particularly in eyes with advanced disease.[31]

The diagnosis and management of PACG has been outlined in Flowchart 4.

Regular Review

Patients with PACD should be followed up on a regular basis to ensure prevention or delay in progression and to maintain quality of life.

- PACS patients following LPI, can be monitored on a yearly basis. Those with strong family history of glaucoma, monocular patients and those with occludable angles even after LPI can be followed up once in 6 months. Similarly, PACS patients who are under
  observation (without LPI) can also be followed up yearly
- PAC and APAC patients after iridotomy might need AGM to control IOP, and should be followed up atleast once in 6 months, even if they are stable. Patients who are on glaucoma medications should be followed up in more frequent intervals determined by severity of perimetric damage, to augment medical management or to decide on the surgical intervention (Flow chart 2)
- PACG patients post LPI should be treated similar to POAG patients. However, these patients need more frequent and meticulous review to avoid progression. If they are stable they can be followed up once in 4-6 months. They might need more frequent follow up
- if unstable to decide on further management (Flowchart 3 and 4)

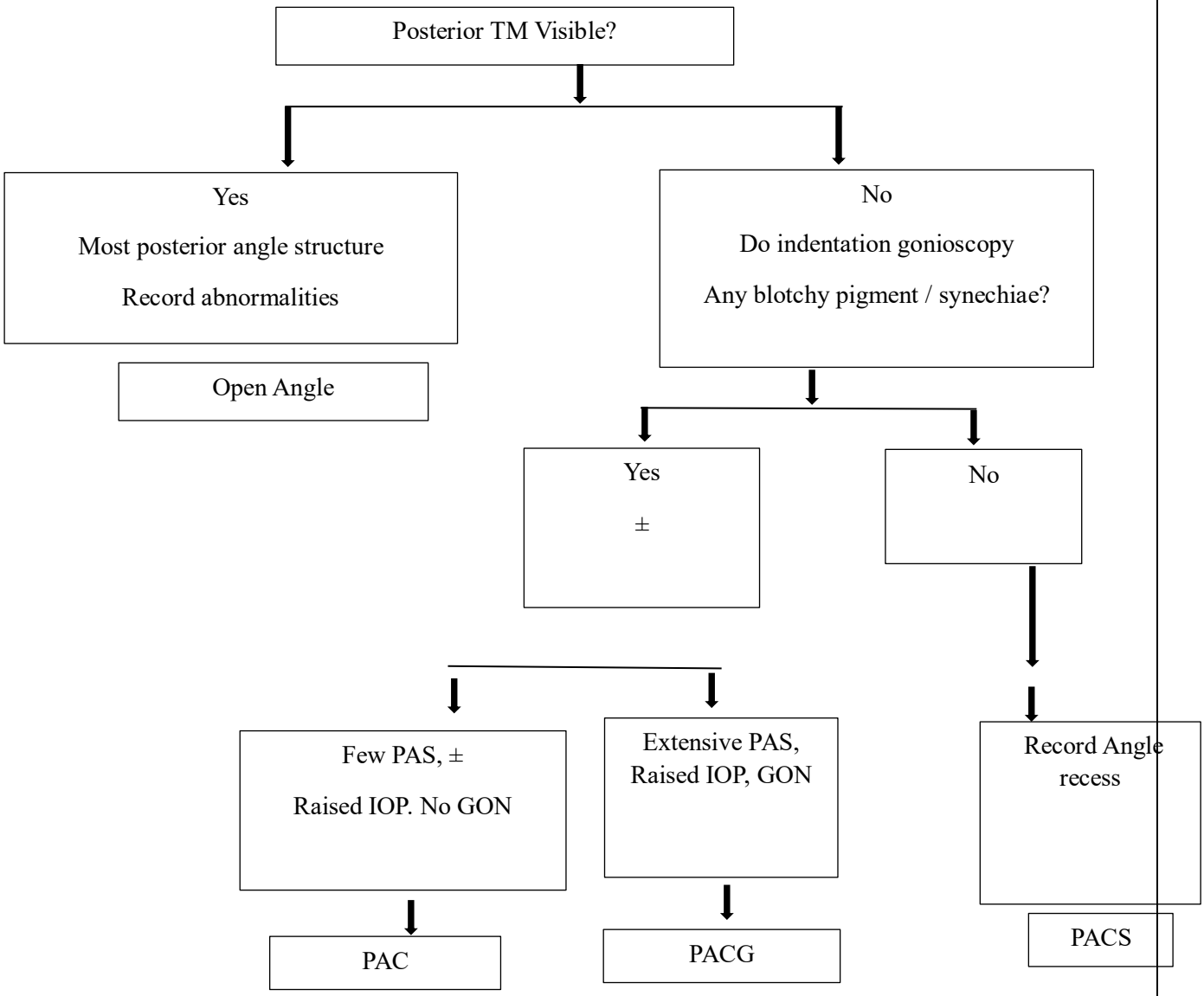
Care and support:

The goal of treatment is preservation of visual function by regular follow-up. One needs to identify patients at risk of acute angle closure. Chronic angle closure glaucoma can have a huge impact on a patient's well-being as both eyes are affected. Such patients will struggle with daily activities such as reading, walking, and even driving a two-wheeler or four-wheeler. The impact is more when glaucoma is severe. They can experience falls within and outside the home and are more likely to be involved in motor vehicle accidents. Quality of life can be affected in all stages of glaucoma.

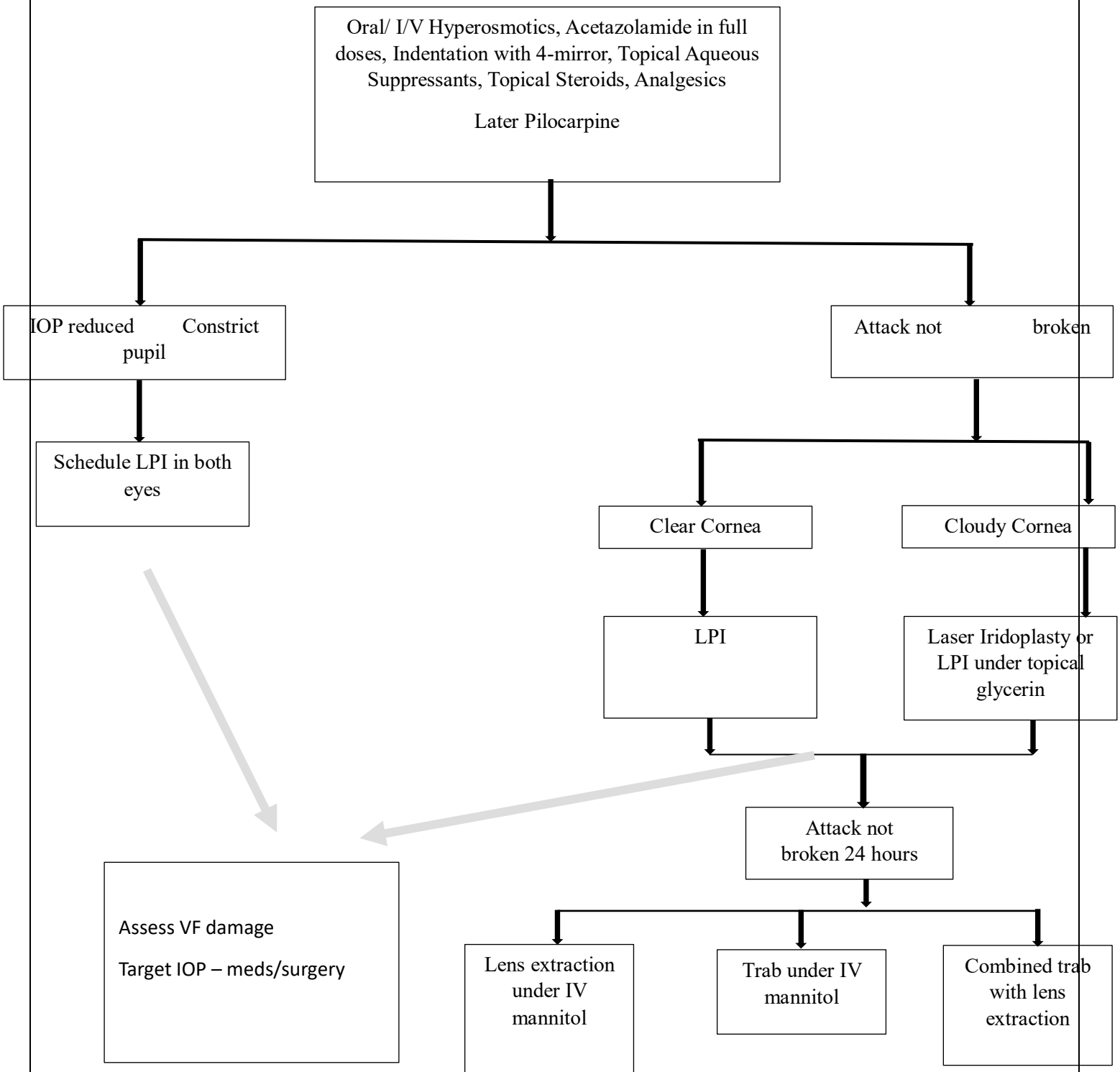
We need to be sensitive to their problems and support the socioeconomically disadvantaged patients to have regular follow up and receive care that is in line with recommended clinical practice guidelines.

It is important to educate patients about glaucoma and engage them in the management of their condition to prevent blindness from this serious disease.

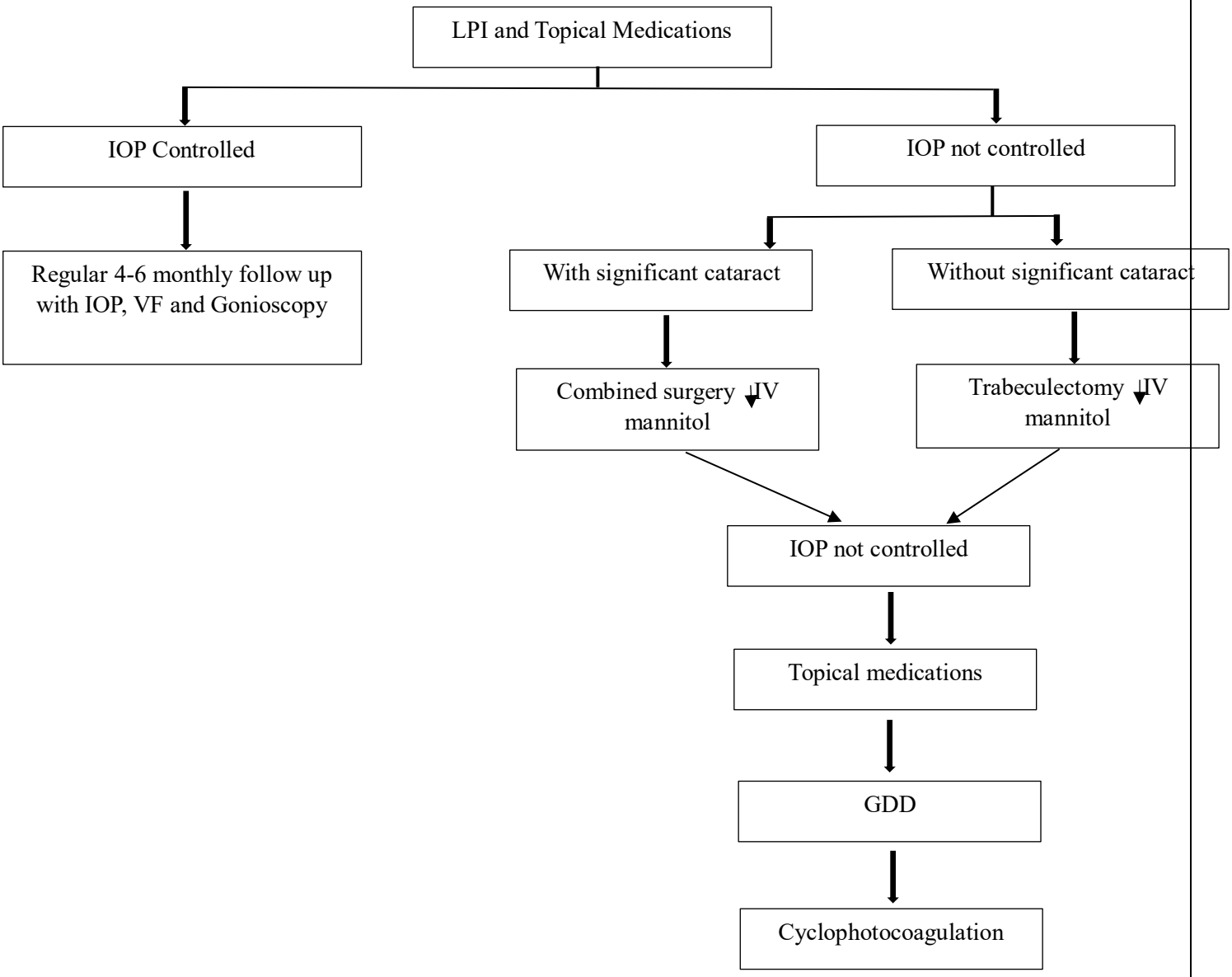
Flowchart 1: Evaluation of Angle



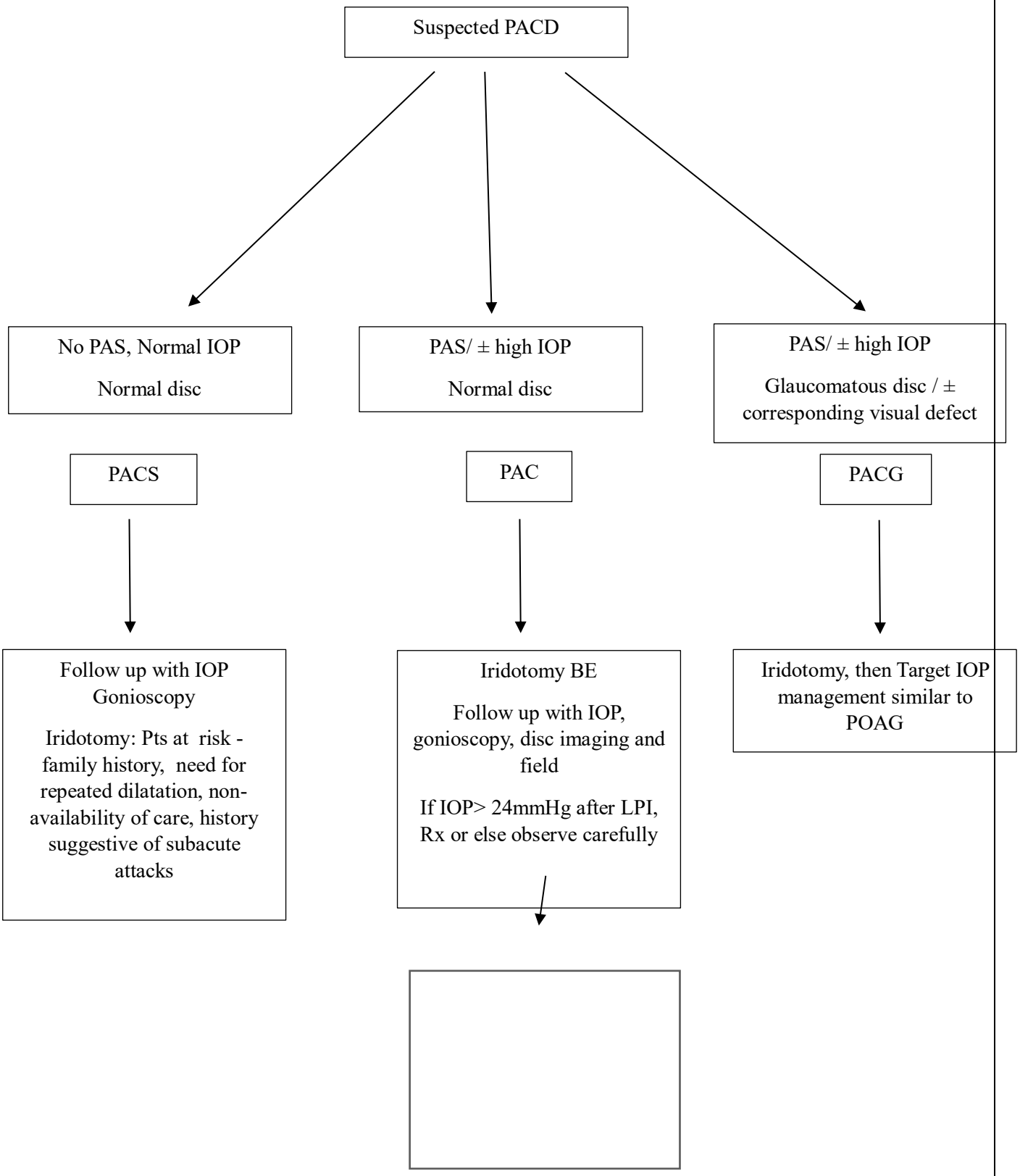
Flowchart 2: Management of Acute Primary Angle Closure



Flowchart 3: Primary Angle Closure Glaucoma Management and Follow up



Flowchart 4: Management overview of Primary Angle Closure Disease



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Secondary Glaucomas

1. Glaucoma in Uveitis

Raised IOP in the setting of Uveitis is estimated to occur in 20% of uveitis patients.

- Can be Open angle glaucoma in Acute setting due to increased viscosity of aqueous, inflammatory cells blocking Trabeculum, Inflammation (and scarring as consequence) of trabeculum or hypersecretion in early uveitis stages.
- Can be closed angle due to peripheral synechiae developing due to inflammation or due to pupillary block (Iris bombe) by seclusio or occlusio pupilae.

Uveitis is to be treated as per cause/type of uveitis.

Medical Rx preferred for IOP. Aqueous suppressants preferred as 1st line (order of topical Rx if not contraindicated in the given patient – Beta blockers→ Alpha agonists→ Carbonic anhydrase inhibitors → ROCK inhibitors → Pg analogues). For short term one can use Acetazolamide tablets and if IOP very high IV Mannitol.

If not controllable medically one can opt for Antimitotic Augmented trabeculectomy or a Tube shunt procedure. If there is a cataract the same can be dealt with also. Failure rates for filtration surgery is higher in Uveitic glaucoma. Late hypotony can occur with eventual Ciliary shut down also.

Specific Situations

(i) **Posner Schlossman Syndrome**- Acute episodic loss of vision with mild discomfort and relatively white eye. Non granulomatous uveitis with small -medium sized white KPs and open angle glaucoma. Corneal edema and IOP in 50s often. In acute phase Uveitis and glaucoma are to be treated medically. Often returns to normal in a week or so. Repeated episodes can cause disc and field changes.

(ii) **Herpes Group**- Uveitis due to HSV, HZV and CMV can have glaucoma. HSV being commoner in this situation. Very high IOP with corneal edema is the rule and can be recalcitrant to medical Rx and often need surgery. Oral antivirals may be required long term.

(iii) **Fuchs heterochromic uveitis** - Associated with an open angle glaucoma that is slightly more aggressive than POAG. Treatment is same as for POAG. Rarely can have new vessels in the angle and may lead to failure of glaucoma surgery. These eyes will need retinal consult and may benefit with implant surgery for IOP control.

(iv) **Grant syndrome** – Inflammatory precipitates on Trabeculum. Granulomatous Uveitis. Otherwise quiet eye. Responds to topical steroids well. Can lead to synechial closure over many years.

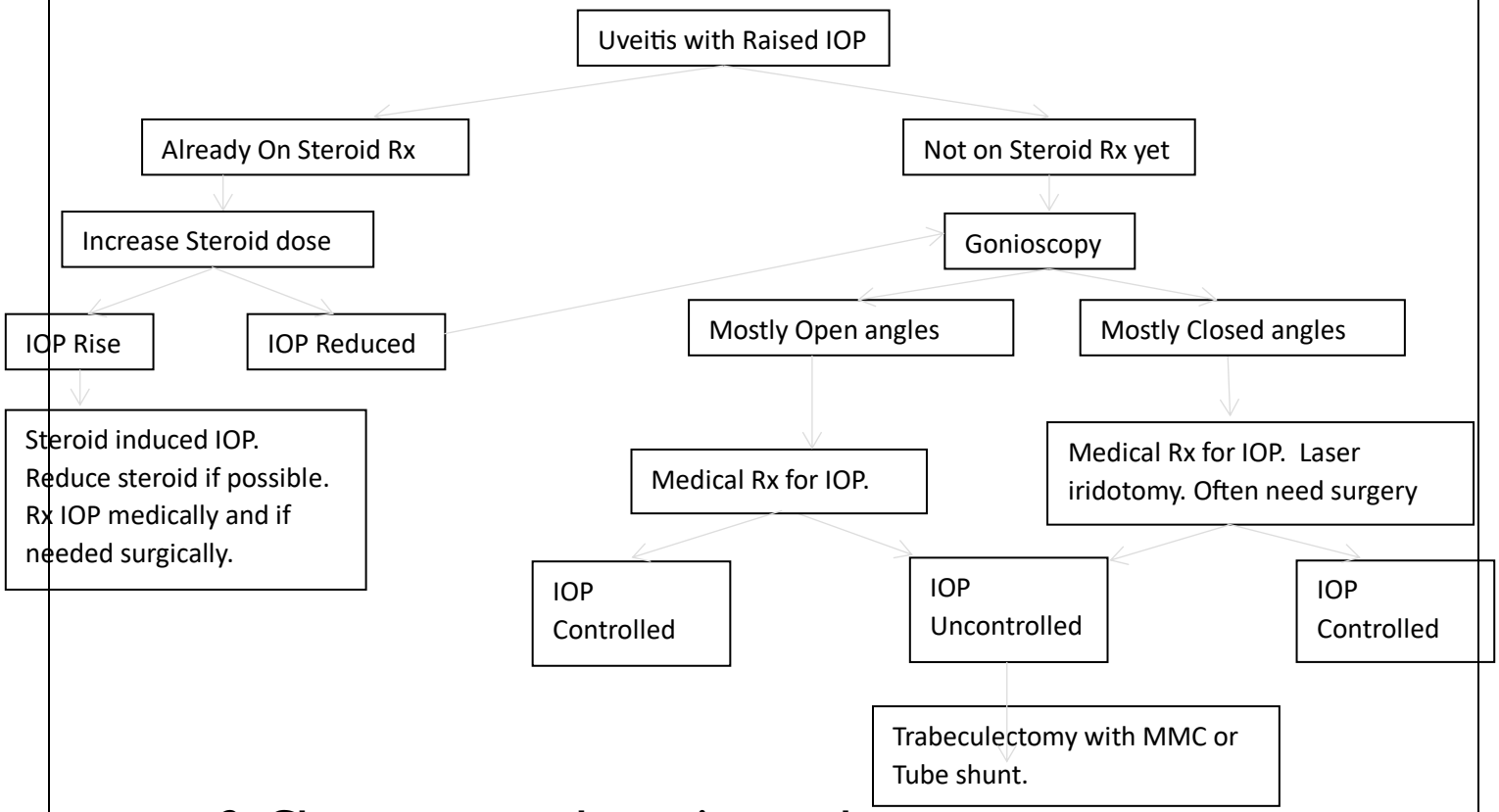
Pointers to diagnosing

- **Symptoms to suspect glaucoma in Uveitis**
  Symptoms - Halos, headache with vomiting, diminished vision more than expected from uveitis.
  Signs- Shallow Anterior chamber, Iris bombe, Steamy looking cornea.
- **Investigations**
  IOP, Gonioscopy and disc evaluation.
- **If IOP high in Uveitis**
  Manage medically to start with. Avoid Prostaglandin analogues and pilocarpine as far as possible. Aqueous suppressants are preferred (Beta blocker, Brimonidine and Carbonic anhydrase inhibitors). For very high pressures (above 30mmHg) use **oral** acetazolamide in the short term.

Systemic evaluation would be needed to rule out an infective etiology or a immunologic etiology. Appropriate investigations would be needed to establish the etiology and treat the cause.

When do we need to refer to higher centres?
IOP not getting controlled in reasonable time (say 2-4 weeks under 30 or Above 30 for few days) as surgery may be needed.
If there is advanced disc changes.
Shallow anterior chambers as Iridotomy may be needed.

Management protocol is given in the attached Flow Chart.



2. Glaucomas secondary to intraocular surgery

Post-surgical glaucoma is a secondary glaucoma that can develop following various intraocular surgeries. Pre-existing glaucoma can also be worsened by surgery.

Acute IOP elevation: In the immediate postoperative phase, due to inflammation, hyphema, uveitis, or retained viscoelastic. This may be painful and associated with decreased vision.

Chronic insidious elevations in IOP: Over a longer term this could be due to PAS, especially after hypotony and flat anterior chamber

(i) Primary ophthalmic care

Know when to refer the patient and when to treat.

- Detailed history:
  - History of pain, redness, coloured haloes, decreased vision
  - History for nausea, vomiting, or headache
  - History of previous eye illness or surgery, systemic illness and use of recent / chronic medications.
- Vision assessment
- Torchlight examination:
- Note whether the external eye structures appear normal in anatomy.
- Look for any signs of high IOP:
  - High digital eye pressure
  - Apparent signs of recent surgery: redness, sutures, scar, conjunctival hemorhhage/chemosis
  - Loss of corneal transparency/ graft opacification/ corneal sutures
  - Shallowing of anterior chamber, blood in anterior chamber, air bubble or silicon oil in AC
  - Non reacting / irregular pupil / surgical or laser iridotomy/ pupillary membrane/ hypopyon
  - Intraocular lens position, subluxation
- Measure IOP whenever possible using non-contact tonometry
- If no apparent pathology or abnormal vision, give symptomatic treatment and counsel.

Refer if:

- Any patient post-surgery who complains of decreased vision, redness, pain, watering and discharge must be referred to higher centre.
- High index of suspicion in post vitreoretinal surgery and corneal transplant patients; and children who have undergone any intraocular surgery.
- Most important differential diagnosis is endophthalmitis. Both, endophthalmitis and post-surgery glaucoma are sigh threatening emergencies.
- In teleconsultation with higher centre, start antibiotics or anti-glaucoma medication as advised, immediately. Counsel patient/ caregiver about emergency, and refer to higher centre.

(ii) Secondary ophthalmic care

- Thorough history taking, visual acuity assessment, detailed slit lamp examination, IOP measurement, fundus evaluation, perimetry, and gonioscopy (if possible). Remember, IOP measurement in eyes following vitreoretinal and corneal surgery may not always be accurate.
- Imaging modalities: USG B scan, UBM, ASOCT, ONH imaging
- Appropriate treatment and follow up advice.

- In case of intractable glaucoma, or if IOP not controlled by maximal tolerable medical therapy, consider glaucoma surgery.
- In case pupillary bock, consider laser peripheral iridotomy, in case of malignant glaucoma or aqueous misdirection, consider laser disruption of the posterior capsule and anterior hyaloid face.
- Surgery for secondary glaucomas is best performed in tertiary care centres. This includes trabeculectomy with anti-metabolites, primary shunt surgery or cyclodestructive procedures.

Refer if:

- Diagnosis and treatment difficult.
- Surgery required.

(iii) Tertiary care

- Review the history, conduct a comprehensive eye examination and conduct further investigations, including electrophysiological tests.

Key Guidelines:

- Diagnosis requires a strong index of suspicion even if there is no increased IOP in the immediate post-surgical period. Special care post vitreoretinal surgery, post keratoplasty and following paediatric cataract surgery.
- In early onset glaucoma, consider inflammation, hyphema, uveitis, or retained viscoelastic.
- In case of late onset glaucoma, consider steroid response, uveitic glaucoma, coexistent POAG, secondary angle closure and malignant glaucoma, as well as epithelial downgrowth and fibrous ingrowth.
- Primary objective: prevent vision damage
  - Control IOP to Target to prevent glaucomatous optic neuropathy
  - Control inflammation
- Anti glaucoma medication:
  - First-line treatment: topical beta-blocker, alpha-2 agonist, carbonic anhydrase inhibitor, ROCK inhibitors, or their combination. Prostaglandin analogues and pilocarpine may increase inflammation, so may be used as reserved drugs.
  - Topical and oral, to be stepped up as per the need after assessing the efficacy. If the IOP is high, start with multiple agents simultaneously, withdraw treatment stepwise.
  - Use of intravenous mannitol/hyperosmotic agents for vitreous shrinkage and posterior displacement of lens-iris diaphragm, if indicated.
- Topical, periocular and systemic corticosteroids for inflammation. Cycloplegics to control pain.
- Rule out pupillary block (perform LPI) ad aqueous misdirection (consider laser disruption of posterior capsule/ anterior hyaloid). The latter may require vitrectomy.
- Glaucoma surgery is best performed at a tertiary centre- trabeculectomy with anti-metabolites or primary shunt surgery. The latter is usually preferred for secondary glaucomas.
- Long-term treatment and follow up may be needed for all cases of post-surgery glaucoma.

- Surgical interventions, superspecialist care, and visual rehabilitation, as indicated.

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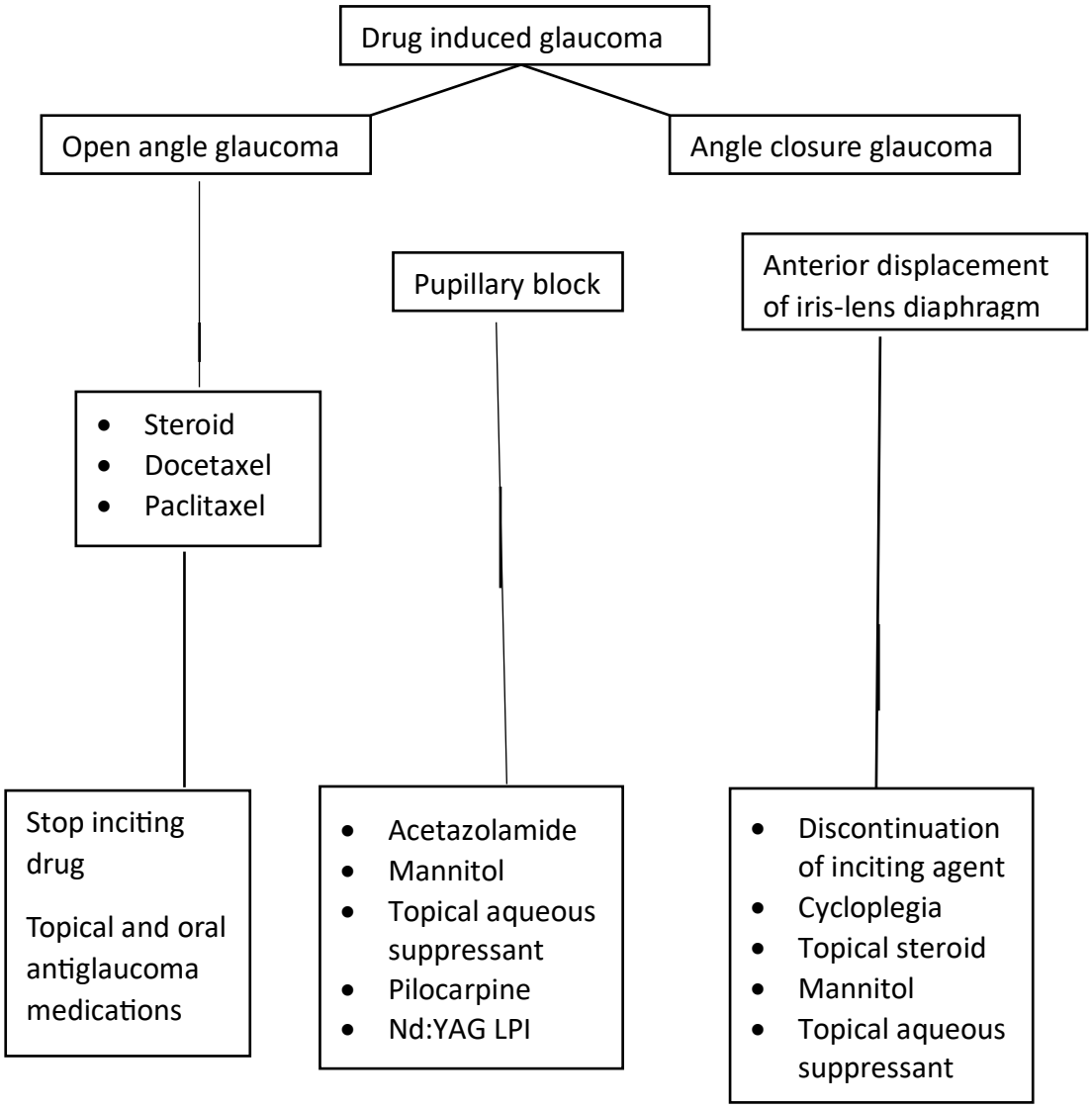
3. Drug Induced Glaucoma

- Incidence of drug induced glaucoma is uncertain.
- It can lead to secondary open angle glaucoma or secondary angle closure glaucoma.
- Drugs that cause or exacerbate open-angle glaucoma are mostly glucocorticoids (discussed in steroid induced glaucoma).
- Drug induced acute angle closure glaucoma can develop in a susceptible individual by various classes of drugs.
- Various classes of drugs precipitating acute angle closure attack are listed in Table 1.

Acute angle closure- Broadly, there are two mechanisms of drug-induced acute angle closure (AAC) with different management protocols. The bilateral acute angle closure can be primary or secondary.

1. Primary due to pupillary block secondary to thickening of iris base with mydriasis
2. Secondary due to anterior displacement of the lens–iris diaphragm (due to ciliary body edema, ciliary detachment, blood or uveal effusion)

- **Symptoms**: Headache, nausea, vomiting, blurring of vision, severe eye pain, halos and mostly bilateral involvement.
- **Patient's history** must be suggestive of inciting medications.
- **Sign**: usually bilateral involvement, elevated IOP, corneal edema, congested conjunctiva, and shallow anterior chamber. mid-dilated and sluggishly reacting pupil in primary and almost normal pupil in secondary. Myopic shift in refraction in secondary AAC and hyperopia or emmetropia in primary AAC
- **Investigations**: IOP, gonioscopy, disc examination, UBM to look for anterior rotation of ciliary body
- **Management of AAC due to pupillary block (Primary):**
  → Acetazolamide, mannitol, topical aqueous suppressant, topical steroid, pilocarpine, and Nd:YAG laser peripheral iridotomy (LPI)
- **Management of AAC due to anterior displacement of lens–iris diaphragm (secondary):**
  → Immediate discontinuation of inciting agent
  → Cycloplegia, topical corticosteroids, and mannitol and topical aqueous suppressant
  → Acetazolamide should be avoided, if inciting agent is sulpha drug
  → Pilocarpine is contraindicated
  → LPI will not help and should be avoided



- In most of the cases patients can be managed with available treatment option after taking detail clinical history (to find out the etiological agent)
- After conformation of the inciting drug, patient need to be educated about the risk related with the drug.

Primary Ophthalmic Care

- Diagnose an abnormality – fall in vision, corneal edema, raised IOP and refer
- Detailed history of ophthalmic and systemic disease along with detailed history of recent drug intake or instillation of eye drops or hospital admission. It helps to identify the inciting agent.
- Comprehensive ophthalmic examination including visual acuity, IOP, anterior chamber depth assessment (gonioscopy) and disc evaluation.
- Management of high IOP with topical and systemic anti-glaucoma medications.

Secondary Ophthalmic Care

- Thorough systemic and ophthalmic history taking and history related with drug usage and comprehensive ophthalmic examinations (assessment of visual acuity, detailed slit lamp examination- corneal clarity, AC depth, AC reactions, pupillary reaction and dilatation), measurement of IOP with applanation tonometer, gonioscopy to rule out NVA and undilated fundus evaluation is to be done.
- Immediate cessation of inciting drugs.
- Control of IOP based on mechanism (pupillary block or anterior displacement of iris-lens diaphragm).
- Avoid pilocarpine and YAG-LPI in cases of anterior displacement of iris-lens diaphragm.
- After IOP control repeat all the eye examinations. Dilated fundus examination after YAG-LPI in pupillary block and after deepening of AC in forward shift of iris lens diaphragm.
- Keep the patient on close follow up.
- In cases of suspicious anterior shift of iris-lens diaphragm and IOP failed to control with maximum medical management, may occur due to permanent angle closure due to long standing insult, may even have severe inflammation and hypotony in cases with delayed diagnosis of SAAC
- In almost all cases, IOP are controlled after all above mentioned therapy.
- Educate the patient and his relatives about the precipitating drugs and take opinion from the physician to stop the drug and switch on other medication, if necessary, with close ophthalmic follow up.

Need referral if:

- IOP is medically uncontrollable.
- Vision fails to improve due to choroidal effusion and necessitate surgical retinal intervention.

Tertiary Ophthalmic Care

- Review of history to identify the causative agent.
- Comprehensive eye examination.
- Stop the inciting drugs.
- Control of IOP depending on the mechanism of raised IOP (pupillary block or anterior displacement of iris-lens diaphragm)
- Vitreo-retinal consultation for intervention in unresponsive choroidal effusion.

Table 1: Classes of drug causing acute angle closure.

Class of Drugs	Drugs	Mechanism
Sulpha drugs	Topiramate, Acetazolamide, Hydrochlorothiazide and Cotrimoxazole	Anterior displacement of lens–iris diaphragm
Adrenergic agonists	Epinephrine, Norepinephrine, Ephedrine, Phenylepinephrine, Amphetamines, and Cocaine	Pupillary block
Cholinergics	Pilocarpine, Acetylcholine, Carbachol	Anterior displacement of lens–iris diaphragm

Anticholinergics	Atropine, Homatropine, Cyclopentolate, Tropicamide and Ipratropium bromide	Pupillary block
Serotonergic agents	Aripripazole, Sumatriptan, Venlafaxine, Bupropion, Mirtazapine, Escitalopram	Pupillary block/anterior displacement of lens–iris diaphragm
Anti-histaminic	Promethazine, Cetirizine, Chlorpheniramine, Ranitidine, and cimetidine	Pupillary block
Anticoagulants	Low molecular weight heparin (enoxaparin, warfarin)	Anterior displacement of lens–iris diaphragm

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4. Traumatic glaucoma

- It is a type of secondary glaucoma that develops following blunt or penetrating ocular trauma.
- It usually entails several mechanisms that interplay to produce increased intraocular pressure (IOP).
- It is important to consider all the possible mechanisms leading to increased IOP as they influence patient management.

Primary ophthalmic care

The most important aspect of managing an ophthalmic patient at primary level is knowing when to refer the patient and when to treat.

- Vital signs should be checked first and monitored.
- Detailed history:
  - History of type of trauma, duration since injury, other associated complaints.
  - History for loss of consciousness, vomiting, headache or any ENT bleed.
  - History of previous eye illness or surgery, systemic illness and use of recent / chronic medications.
  - Particularly note if the patient is on any antihypertensive agents and blood thinners.
- Brief assessment of other injuries must be performed.
- Once the patient is found to be stable, ophthalmic evaluation can be done.
- Torchlight examination:
  - Note whether the external eye structures appear normal in anatomy.
  - Look for any signs of penetrating injury.

Look for

- Loss of transparency in cornea
- Shallowing of anterior chamber
- Blood in anterior chamber
- Non reacting / irregular pupil
- Subluxation of lens
- Restricted ocular motility.

- Measure IOP using non contact tonometry if available and possible.
- If no apparent pathology is present and patient has normal vision, give symptomatic treatment and counsel.
- Provide protection to globe from further injury if necessary.

Refer if:

- History suggestive of head injury
- Penetrating trauma
- Torch light examination shows any abnormal findings
- Vision not improving to 6/6
- Difficulty in opening eyes, proptosis, periorbital edema, and afferent pupillary defect – which constitute an ocular emergency.

- Tele-ophthalmic consultation can be done with higher centre if necessary.

Secondary ophthalmic care

- Thorough history taking, visual acuity assessment, detailed slit lamp examination, IOP measurement and fundus evaluation is to be done.
- Do a gonioscopy if possible - deferred in case of a ruptured globe or intraocular foreign body.
- Ultrasonography of posterior segment, CT scan or MRI maybe done as necessary.
- If penetrating injury is present, primary repair has to be done.
- Appropriate treatment has to be initiated and follow up advised accordingly.
- Patient and relatives need to be explained the importance of adherence to treatment and possible prognosis.

Refer if:

- Diagnosis difficult.
- Multiple interventions needed.

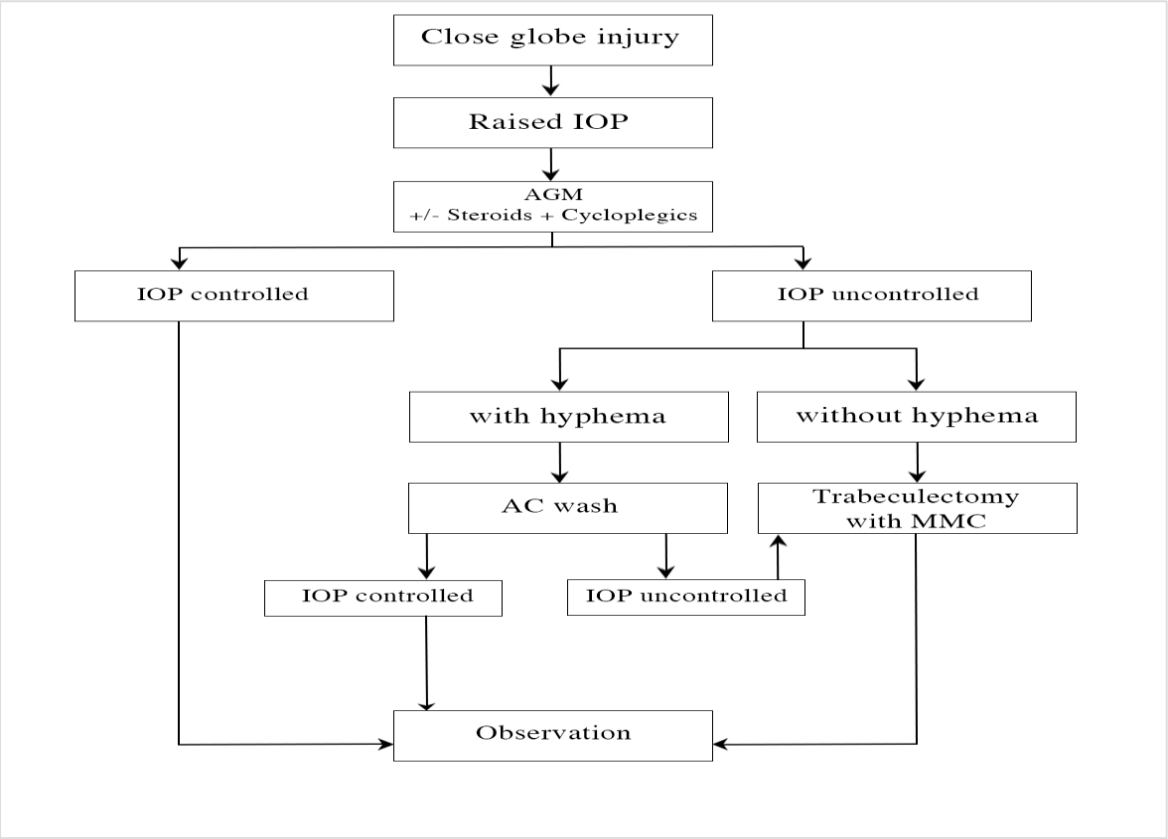
Tertiary care

- Review the history
- Conduct a comprehensive eye examination and look for any missed findings like secondary glaucoma / angle closure.
- Review or conduct further investigations.
- All patients need to undergo gonioscopy at appropriate times to look for angle recession and foreign body.
- Electrophysiological tests can be done in cases of unexplained visual loss.

Key Guidelines:

- Management depends on interval between injury and presentation, clarity of ocular media and degree of IOP elevation.
- Rule out contra-indications for any topical / systemic drugs.
- Topical, periocular and systemic corticosteroids are used in the setting of inflammation without infection.

- Cycloplegics, short-acting and long-acting, are used to control pain.
- IOP lowering agents, topical and oral, are used in the setting of IOP elevation. Avoid prostaglandin analogues and pilocarpine. Medicines to be stepped up as per the need after assessing the efficacy – similar to the protocol for primary open angle glaucoma.
- Topical, oral and intravenous antibiotics should be used depending on the injury.
- Anti glaucoma surgery is better done at a tertiary centre - trabeculectomy with anti metabolites or primary tube surgery can be done.
- Long-term treatment and indefinite follow up may be needed.



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5. Neovascular Glaucoma

Neovascular glaucoma (NVG) is a form of vision threatening secondary glaucoma characterized by elevated intraocular pressure (IOP) and neovascularization of the angle, culminating in optic nerve damage. The main pathology is retinal ischemia leading to an impaired balance between angiogenic and anti-angiogenic factors and the main causes are:

- Proliferative diabetic retinopathy
- Central retinal vein occlusion
- Central retinal artery obstruction
- Ocular ischemic syndrome
- Chronic retinal detachment
- Chronic uveitis

Management is divided into:

- Treatment of retinal ischemia through PRP or intravitreal Anti VEGFs + PRP.
- Control of high IOP by topical and systemic antiglaucoma medications or surgical interventions in presence of uncontrolled IOP and progressive damage to the optic nerve.

Primary Ophthalmic Care

The most important aspect of managing an ophthalmic patient at primary level is knowing when to refer the patient and when to treat.

- Refer longstanding diabetics for an Ophthalmic review/ fundus photography and teleconsult
- Refer painful eyes to secondary center
- Detailed systemic history and work up to identify the cause for NVG.
- Detailed ophthalmic examination: visual acuity, slit lamp biomicroscopy to look for NVI, gonioscopy to look for NVA, tonometry (non-contact / applanation) and detailed fundus evaluation for cause.
- Management of high IOP using topical and systemic drugs. Avoid pro-inflammatory drugs like prostaglandin analogues and pilocarpine. Systemic acetazolamide after understanding the renal and systemic status of the patient.

Topical steroids to reduce inflammation in case of hyphema or severe inflammation

- Topical cycloplegics like atropine to reduce pain.

*__Refer__ all *patients with NVG to secondary center at the earliest unless it is a longstanding NVG (with a known cause) and patient is asymptomatic*.

Secondary Ophthalmic Care

- Thorough history taking, visual acuity assessment, detailed slit lamp examination, IOP measurement (applanation), gonioscopy to rule out NVA and determine stage of NVG and fundus evaluation is to be done.
- Systemic assessment and control of the underlying comorbidities.
- FFA to identify the ischemic area after ruling out any renal impairment.
- Intravitreal Anti-VEGF injections and PRP to curb down the VEGF stimulation.
- Control of IOP to Target depending on severity of damage, and if uncontrolled, plan trabeculectomy with antimetabolite or a drainage device surgery after antiVEGF and or PRP.
- Close follow up.

- Counseling of patient and relatives about the importance of compliance and prognosis.

Refer if:

IOP is medically uncontrollable and primary surgery is likely to fail.

- There is an apparent need for cyclodestructive procedures.
- Vision is very low because of severe retinopathy, macular edema or vitreous haemorrhage, which may necessitate expert surgical intervention.

Tertiary Ophthalmic Care

- Review of history
- Comprehensive eye examination to detect cause.
- Systemic assessment and control of the underlying morbidities.
- Antiglaucoma medications, steroids and cycloplegics to control IOP
- Intravitreal Anti VEGF injections and PRP with additional anterior retinal cryopexy if required.
- Augmented trabeculectomy or drainage device or cyclodestructive procedure as per the overall patient profile.
- Expert vitreo-retinal advice and intervention.

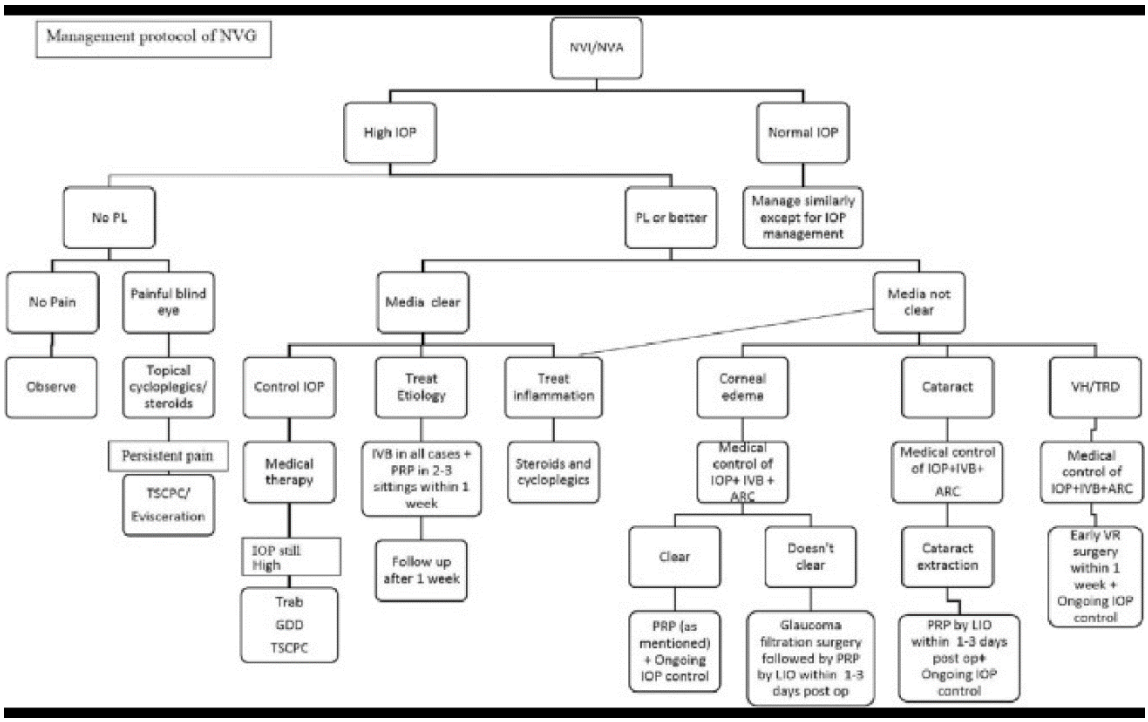
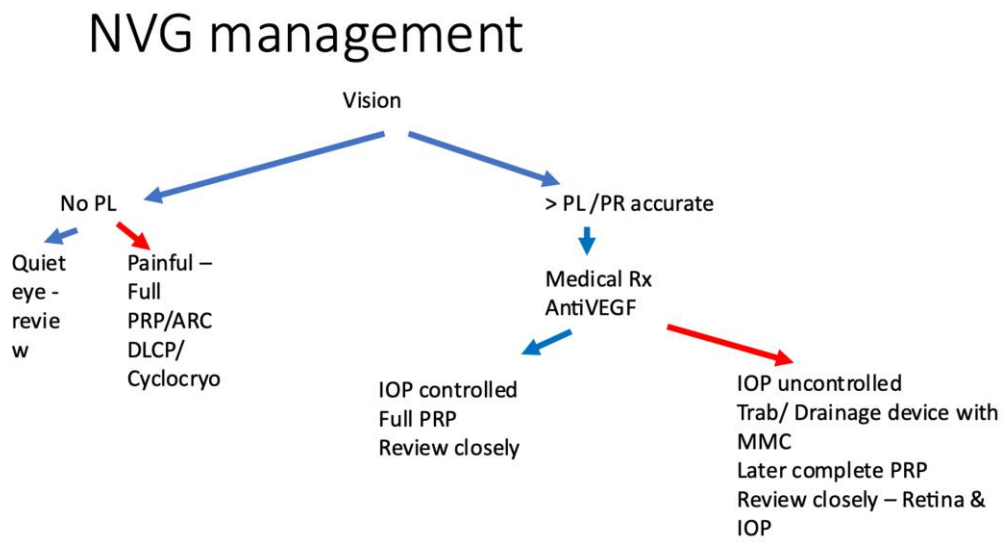


Image courtesy: Senthil S, Dada T, Das T, Kaushik S, Puthuran GV, Philip R, Rani PK, Rao H, Singla S, Vijaya L. Neovascular glaucoma - A review. Indian J Ophthalmol. 2021 Mar;69(3):525-534.

NVG Management



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