2.4 - PRIMARY ANGLE-CLOSURE

Scientific publications on angle-closure have suffered from the lack of a uniform definition and specific diagnostic criteria. Only in recent years has there been recognition of the need to standardize definitions of the various types. Angle-closure is defined by the presence of iridotrabecular contact (ITC). This can be either appositional or synechial. Either can be due to any one of a number of possible mechanisms. Angle closure may result in raised IOP and cause structural changes in the eye. Primary angle-closure (PAC) is defined as an occludable drainage angle and features indicating that trabecular obstruction by the peripheral iris has occurred. The term glaucoma is added if glaucomatous optic neuropathy is present: Primary angle-closure glaucoma (PACG). The main reason to distinguish Primary angle-closure glaucoma from Primary open-angle glaucoma is the initial therapeutic approach (i.e. iridotomy or iridectomy) and the possible late complications (synechial closure of the chamber angle) or the complications resulting when this type of glaucoma undergoes filtering surgery (uveal effusion, cilio-lenticular block leading to malignant glaucoma)\textsuperscript{121,122}.

The prevalence of primary angle closure glaucoma (PACG)
Ethnic background is one of the major factors determining susceptibility to primary angle-closure (PAC). Population surveys show that PAC is more common among people of Asian descent than those from Europe. Among people aged 40 years and over, the prevalence of PAC ranges from 0.1% in Europeans\textsuperscript{123,124} through 1.4% in East Asians\textsuperscript{123,124} and up to 5% in Greenland Inuit\textsuperscript{125}. Of those over 40 years old in European derived populations, 0.4% are estimated to have PACG. Three-quarters of cases occur in female subjects. There are 1.60 million people in Europe and 581 000 people in the USA with PACG\textsuperscript{126}.

Primary glaucoma cases should be examined and the anterior chamber angle shown to be open on gonioscopy before PACG is excluded\textsuperscript{127}.

**Provocative Tests**
In general provocative tests for angle-closure provide little additional information since even when negative they may not rule out the potential for angle-closure. In addition they may be hazardous, triggering an acute angle-closure attack even while the patient is monitored.
2.4.1 Primary Angle-Closure (PAC)

Angle-closure is defined by the presence of iridotrabecular contact (ITC). Gonioscopy remains the standard technique for identifying ITC. Primary angle-closure (PAC) results from crowding of the anterior segment, and as such, usually occurs in eyes with smaller than average anterior segment dimensions. Pathological angle-closure is defined by the presence of ITC combined with either elevated intraocular pressure (IOP) or peripheral anterior synechiae (PAS), or both. The absence of ocular diseases which may induce the formation of PAS such as uveitis, iris neovascularisation, trauma and surgery, defines primary angle-closure. Additionally, angle-closure resulting from the action of forces at the level of the lens or behind the lens is usually regarded as secondary (i.e. cataract, massive vitreous haemorrhage, and silicone oil or gas retinal tamponade) as the successful management is aimed at the underlying lens or posterior segment pathology. Angle-closure may impair aqueous outflow through simple obstruction of the trabecular meshwork (TM), or by causing irreversible degeneration and damage of the TM.

2.4.1.1 Natural History of PAC

PAC becomes more likely as the separation between the iris and TM decreases\(^{128}\). The risk of iridotrabecular contact in a “narrow” angle begins to increase once the iridotrabecular angle is \(\leq 20^\circ\). With angles of \(20^\circ\) or less, signs of previous angle-closure, such as PAS or iris pigment on the TM, should be carefully sought as signs of previous closure. Most angle-closure occurs asymptptomatically. Although symptoms of pain, redness, blurring of vision or haloes may help identify people with significant angle-closure, the sensitivity and specificity of symptoms for identifying angle-closure are very poor. The most commonly identified sign which indicates that treatment is required is ITC. There is not a precise extent of gonioscopically evident ITC which will dictate the indication to treatment for all cases. An international group of experts reached a consensus that 2 quadrants or more of ITC is an indication for prophylactic treatment\(^{130}\) [II,D]. Clearly, in established disease with high IOP, established PAS or glaucomatous optic neuropathy, any potential for angle-closure should be considered and treated on individual merits.

2.4.1.2 Staging of Primary Angle-closure\(^{123}\)

a) **Primary Angle-closure Suspect (PACS)**
   Two or more quadrants of iridotrabecular contact (ITC), normal IOP, no PAS, no evidence of glaucomatous optic neuropathy (GON).

b) **Primary Angle-closure (PAC)**
   Iridotrabecular contact resulting in PAS and/or raised IOP. No evidence of GON.

c) **Primary Angle-closure Glaucoma (PACG)**
   Iridotrabecular contact causing GON; PAS and raised IOP may be absent at the time of initial examination.
2.4.1.3 Ocular Damage in Angle-closure

Primary angle-closure (PAC) may cause ocular tissue damage in many ways. Corneal endothelial cell loss occurs after symptomatic “acute” angle-closure. With very high IOP values the iris may suffer ischaemic damage to musculature causing iris whirling (distortion of radially orientated fibres) and/or a dilated, unresponsive pupil. The lens epithelium may suffer focal necrosis causing anterior sub-capsular or capsular opacity of the lens associated with focal epithelial infarct called “Glaukomflecken”. The trabecular meshwork can be damaged by the formation of PAS, or as the result of long-standing appositional closure. Optic neuropathy in angle-closure may manifest in at least 2 ways. After an “acute” symptomatic episode, the disc may become pale but flat, suggesting an anterior ischaemic optic neuropathy. Typical glaucomatous optic neuropathy manifests in with an excavated surface and a pattern of visual field loss indistinguishable from open-angle glaucoma. Angle-closure accounts for 50% of all glaucoma blindness worldwide, and is probably the most visually destructive form of glaucoma.

2.4.1.4 Outcome following treatment

In asymptomatic (“chronic”) angle-closure, a high presenting pressure (>35 mmHg), more than 6 clock hours of peripheral anterior synechiae and/or established glaucomatous optic neuropathy are signs that a case of angle-closure will not respond fully to a laser iridotomy and that a trabeculectomy may be needed to control pressure.

2.4.1.5 Mechanisms of angle-closure

It is important to identify secondary causes of narrow or closed-angles, such as phakomorphic, uveitic and neovascular cases, as the management of these cases is initially directed at controlling the underlying disease. In isometropic eyes it is helpful to compare axial anterior chamber depths of the two eyes. Asymmetry of > 0.2 mm (3 standard deviations) is suggestive of a secondary pathological process. A-mode or ultrasound biomicroscopy may be helpful in measuring axial dimensions (length, AC depth and lens thickness) and defining anatomical relationships. In primary angle-closure these will be the same in each eye. Mechanisms responsible for angle-closure are described in terms of anatomical location of obstruction to aqueous flow, successively, at the pupil, the iris and ciliary body, the lens and behind the lens. This is also order of decreasing frequency of each mechanism. Two mechanisms may co-exist, especially levels I and II (i.e. pupil and iris/ciliary body). Often, one mechanism predominates.

I. Pupillary block mechanism

Pupillary block is the predominant mechanism in around 75% of cases of primary angle-closure. Pupillary block is an exaggeration of a physiological phenomenon in which the flow of aqueous from the posterior chamber through the pupil to the
Classification and Terminology

anterior chamber is impeded causing the pressure in the posterior chamber to become higher than the pressure in the anterior chamber. As a result, the peripheral iris bows forward and comes into contact with the trabecular meshwork and/or peripheral cornea.

In a minority of cases, this becomes a self-perpetuating cycle with obstruction of trabecular outflow leading to a rise in IOP up to 50-80 mmHg. When total trabecular obstruction occurs rapidly (within a few hours), it causes the symptoms and signs of acute angle-closure (AAC).

The increased resistance to trans-pupillary aqueous flow is believed to result from co-activation of both sphincter and dilator muscles, causing the pupil margin to grip the anterior surface of the lens. This may occur in response to physiological stimuli, such as reading in poor light, or pharmacologically, such as with miotic therapy and concomitant dilator muscle stimulation by phenylephrine (the Mapstone provocation test)\textsuperscript{132}. In most cases, the predisposition to pupil block is created by a narrow anterior segment and the age-related increase of lens volume (See Ch. 2.5.1 and 2.5.3).

The prevalence of PAC is higher in elderly people women and in some races (especially East Asians). There is a weaker association with hypermetropia, exfoliation syndrome, diabetes and retinitis pigmentosa.

II. Anomalies at the level of the iris and/or ciliary body (“plateau iris configuration”)

This group of anterior, non-pupil-block mechanisms are sometimes erroneously referred to under the umbrella term “plateau iris”. They are the result of variations in iris and ciliary body anatomy that brings the peripheral iris into contact with the trabecular meshwork. These include a thicker iris, a more anterior iris insertion and a more anterior ciliary body position. These anatomical factors predict failure of a laser iridotomy to open an appositionally closed angle\textsuperscript{133}. Anteriorly positioned ciliary processes cause “typical” plateau iris configuration\textsuperscript{134}. Plateau iris “syndrome” should be differentiated from plateau iris configuration. The “configuration” refers to a situation in which the iris plane is flat and the anterior chamber is not shallow axially. In most cases, the angle-closure glaucoma associated with the plateau iris configuration is cured by a peripheral iridectomy. “Plateau iris syndrome” refers to a post-laser condition in which a patent iridotomy has removed the relative pupillary block, but gonioscopically confirmed angle closure recurs without shallowing of the anterior chamber axially. Plateau iris syndrome is rare compared to the configuration, which itself is not common. It usually occurs in a younger age group than pupillary-block angle-closure. The treatment is laser iridoplasty or the long-term use of pilocarpine postoperatively as long as it is needed [II,D]. This syndrome must be considered in the differential diagnosis when the intraocular pressure rises unexpectedly following an adequate peripheral iridectomy procedure for angle-closure glaucoma\textsuperscript{135}. Ideally, treatment should be instituted before synechial closure of the angle occurs [II,D]

III. Anomalies at the Level of the Lens

The most widely recognised risk factor for primary angle-closure is a shallow anterior chamber. The anterior surface of the lens marks the depth of the anterior chamber, and as such, PAC patients typically have a thicker, more anteriorly positioned lens
than people with wide open angles. Nuclear sclerotic cataract is a frequent finding in primary angle-closure. If a separate pathological or iatrogenic process causes the lens to suddenly increase in thickness (e.g. "classic" diabetic or post-traumatic cataract), become more anteriorly positioned (retinal gas or oil tamponade) or subluxate (Marfan syndrome or trauma), this may cause secondary angle-closure (See Ch. 2.5.1 and 2.5.3).

IV. Anomalies posterior to the Lens (Aqueous misdirection syndrome)

In rare cases, aqueous misdirection can complicate the management of primary angle-closure. This may occur following trabeculectomy, lens extraction, laser iridotomy and other surgical procedures. Forward movement of the lens iris diaphragm causes secondary angle-closure resulting in IOP elevation. These cases, typically have very small eyes (axial length <21 mm) and higher hypermetropic refraction (> +6D). It is believed that the ciliary processes come into contact with the lens equator, and/or a firm zonule/posterior capsule diaphragm, causing misdirection of aqueous into the vitreous. As a consequence, the lens/iris diaphragm is pushed forward and occludes the anterior chamber angle. After iridotomy or iridectomy, the use of miotics raises the IOP, whereas the use of cycloplegics reduces the IOP. This ‘inverse’ or ‘paradoxical’ reaction to parasympathomimetics should be tested only after iridotomy has been performed. Ultrasound biomicroscopy can demonstrate abnormal posterior chamber anatomy in these rare cases (See Ch. 2.5.3).

Asymmetry of anterior chamber depth is a cardinal sign of secondary (types III and IV) angle-closure.

Systemic drugs and angle-closure

Systemic drugs which may induce angle-closure in pre-disposed individuals are: nebulised bronchodilators (ipratropium bromide and/or salbutamol), selective serotonin re-uptake inhibitors (SSRI’s), tricyclic antidepressants, proprietary cold and flu medications, muscle relaxants, anti-epileptics (topiramate) and other agents with a parasympatholytic and sympathomimetic action.

2.4.1.6 Demographic risk factors for Primary Angle-Closure

- Older age
- Female
- Asian and Eskimoan Race

Family history if primary angle-closure: family screening is vital in these families as robust evidence now exists for significant increased risk of angle closure in family members of an affected patient: first degree relatives may have a 1 in 4 risk of a PAC disease requiring treatment.
2.4.1.7 Descriptions of subtypes:

Primary angle-closure has previously been divided into 5 clinical subtypes according to mode of presentation. There is debate on whether this approach to classification is useful in determining the prognosis or optimal management.

- Primary Angle-Closure Suspect (PACS)
- Acute Angle-Closure (AAC)
- Intermittent Angle-Closure (IAC)
- Chronic Angle-Closure Glaucoma (CACG)
- Status Post-Acute Angle-closure Attack

2.4.1.7.1 Primary Angle-Closure Suspect (PACS) or “occludable” angle

Etiology and pathomechanism:
Pupillary block or plateau iris configuration; each component plays different roles in different eyes (See Ch. 2.4.1.5).

Features:
Signs:
- Two or more quadrants of iridotrabecular contact (ITC)
- Normal IOP
- No peripheral anterior synechia (PAS)
- No evidence of glaucomatous optic neuropathy (GON)
- No glaucomatous visual field defect

The fellow eye of a documented non-secondary angle-closure is considered capable of occlusion.

Treatment:
PACS or “occludable angle” is a clinical assessment. Whether to treat or not is the responsibility of the ophthalmologist. There is not a precise extent of gonioscopically evident ITC which will dictate the indication to treatment for all cases.

If a PAC suspect has narrow angle with two or more quadrants of ITC but no synechial angle closure, the treatment to offer the patient is laser peripheral iridotomy (LPI) followed by argon laser peripheral iridoplasty (ALPI) in cases with plateau iris configuration [II,D].

The same applies to fellow eyes of primary angle-closure [I,C]. All cases must be assessed individually [I,D]. In general, the risks of treatment are to be balanced against the perceived risk of angle-closure.

2.4.1.7.2 Acute Angle-Closure (AAC) with pupillary block mechanism

Etiology:
Circumferential iris apposition to the trabecular meshwork with rapid and excessive increase in IOP that does not resolve spontaneously.
Pathomechanism:
See Ch. 2.4.1.5

Features:

Signs:
- IOP >21 mmHg, often to 50-80 mmHg.
- Decreased visual acuity
- Corneal oedema, initially mostly epithelial oedema. Shallow or flat peripheral anterior chamber
- Peripheral iris pushed forward and in contact with Schwalbe’s line. Gonioscopy: iridotrabecular contact 360°
- Pupil mid-dilated and reduced or no reactivity
- Venous congestion and ciliary injection
- Fundus: disc oedema, with venous congestion and splinter haemorrhages, or the disc may be normal or show glaucomatous excavation
- Bradycardia or arrhythmia
- Gonioscopy clues from the other eye

Symptoms:
- Blurred vision, “halos” around lights
- Pain
- Frontal headache of variable degree on the side of the affected eye
- Nausea and vomiting, occasionally
- Palpitations, abdominal cramps, occasionally

Treatment options:
See also flowchart FC VII-VIII

A. Medical treatment
B. Laser peripheral iridotomy (LPI)
C. Argon Laser Peripheral Iridoplasty (ALPI)
D. Lens Extraction
E. Trabeculectomy
F. Anterior Chamber Paracentesis
G. Goniosynechialysis (GSL)

Iridotomy or iridectomy together with medical treatment is the preferred definitive treatment of acute angle-closure glaucoma with a pupillary block component [I,D]

A: Medical Treatment [I,D]
Medical treatment serves to lower IOP, to relieve the symptoms and signs so that laser iridotomy or iridectomy is possible
Medical therapy aims for
1. withdrawal of aqueous from vitreous body and posterior chamber by hyperosmotics
2. pupillary constriction to open the chamber angle
3. reduction of aqueous production reduction of inflammation.
All the above steps of medical therapy should be implemented concurrently [I,D]

Consider contraindications to each of the medications to be used

- Reduction of aqueous production
  - acetazolamide 10 mg/Kg intravenously or orally. Topical carbonic anhydrase inhibitors (CAIs) are not potent enough to break the pupillary block
  - topical alpha-2 agonists
  - topical beta-blockers

- Dehydration of vitreous body

Hyperosmotics are the effective agents but carry significant systemic risk in some patients: patients must be evaluated for heart or kidney disease because hyperosmotics increase blood volume which increases the load in the heart [IID]. Glycerol may alter glucose blood levels and should not be given to diabetics (FC VII) [I,D]

- glycerol 1.0 - 1.5 g/Kg orally
- mannitol 1.0 - 1.5 g/Kg intravenously

- Pupillary constriction [I,D]
  - pilocarpine 1% or 2% or aceclidine 2% twice or three times within 1 hour
  Note: while the sphincter is ischaemic and the pupil non-reactive to light for sphincter paresis, multiple applications of topical parasympathomimetics is not helpful, will not cause pupillary constriction and may cause forward rotation of the ciliary muscle, thereby increasing the pupillary block. Since miotics in large doses can cause systemic side effects due to trans-nasal absorption leading to abdominal spasms and sweating, intensive topical parasympathomimetics are no longer indicated to treat this condition. Miotics are likely to constrict the pupil only after IOP has been lowered.
  - dapiprazole 0.5%
  - Alpha-1 blockers relax the dilator muscle. They do not reduce pupil size when the sphincter-muscle is paretic.

- Reduction of inflammation
  Topical steroid every 5 minutes for three times, then 4-6 times daily, depending on duration of raised IOP and severity of inflammation.

B: Surgical Treatment

- Neodymium YAG laser iridotomy
  Laser iridotomy should be attempted if the cornea is sufficiently clear [I,C]. Argon laser iridotomy is rarely performed nowadays but thermal laser pre-treatment (e.g., argon) of dark irides reduces total YAG energy required [II,B]

- Surgical iridectomy
  1) Transcorneal approach.
     - No conjunctival scarring
     - A water-tight self-sealing incision is possible
Classification and Terminology

- Disadvantages:
  - Technically more difficult in dilated fixed pupil and flat anterior chamber
  - More traction on iris with increased risk of haemorrhage

2) Corneoscleral approach
- Advantages:
  - Iridectomy can be basal
- Disadvantages:
  - Conjunctival wound may lead to scarring compromising the outcome of a filtering procedure which may become necessary at a later stage insufficient wound closure and aqueous misdirection may occur in rare cases

3) General advantages of surgical iridectomy:
  - It can be performed even when the cornea is cloudy
  - It allows deepening of the anterior chamber, breaking freshly formed PAS

4) General disadvantages of surgical iridectomy:
  - All the potential risks of any intraocular procedure in an eye with angle closure

C: Argon Laser Peripheral Iridoplasty (ALPI)
There is now some evidence from randomised controlled trials that ALPI can break an attack of acute angle closure as or more swiftly than medical therapy\textsuperscript{140}. Many glaucoma specialists now routinely use ALPI if topical treatment + acetazolamide have not broken an attack within an hour, prior to considering hyper-osmotics. ALPI is also a useful procedure to eliminate appositional angle closure resulting from mechanisms other than pupillary block (i.e: plateau iris configuration)\textsuperscript{141}. Diode Laser Peripheral Iridoplasty has greater penetration of an oedematous cornea but has been less extensively studied. Anterior chamber paracentesis is being evaluated to break the attack in cases that are refractory to medical management\textsuperscript{142}.

D: Lens extraction See FC VII
Clinical reports of phacoemulsification with posterior chamber intraocular lens implantation in the treatment of acute, chronic, and secondary angle-closure +/- glaucoma describe very favourable results. The appropriate role for lens extraction in the management of primary angle closure, however, still remains unproven. The first case-series study showed that cataract extraction was associated with a good reduction in IOP and a reduction in the number of medications required to control IOP\textsuperscript{143}. A few prospective case series or randomized clinical trials have been performed\textsuperscript{143-146} or are ongoing\textsuperscript{147} to determine the value and comparative risks and efficacy of lens surgery, both clear lens extraction and cataract surgery, versus medical therapy, laser peripheral iridotomy, laser iridoplasty, and filtration procedures for the treat-
Classification and Terminology

ment of acute and chronic primary angle closure and for the prevention of chronic angle-closure glaucoma, both after and instead of laser peripheral iridotomy. Cataract surgery in PACG is generally more challenging and prone to complications than in normal eyes or eyes with POAG because of the shallow AC, larger lens, corneal oedema, poorly dilated or miotic pupil, extensive posterior synechiae, lower endothelial cell count, weaker zonules, especially after an acute angle closure attack.

In an eye with a clear lens: laser PI first. If the angle does not open and IOP not well controlled with unquestionable glaucomatous damage, consider to proceed with phacoemulsification and IOL implantation [I,D].

E: Trabeculectomy
Trabeculectomy in chronic PACG is also associated with higher risk of postoperative anterior chamber shallowing, malignant glaucoma, and a significant rate of cataract formation compared to POAG. Even when filtration surgery has successfully reduced the IOP, the ailing trabecular meshwork does not regain its function, and so the disease is not cured.

Combined lens extraction and trabeculectomy
In a study in CACG eyes with coexisting cataract, combined phacotrabeculectomy resulted in significantly more surgical complications than phacoemulsification alone. Visual acuity or disease progression did not differ between the 2 treatment groups.

F: Anterior Chamber Paracentesis
- Rapidly lowers IOP in APAC
- Instantaneous relief of symptoms
- Prevention of further optic nerve and trabecular meshwork damage secondary to the acutely elevated IOP
- The IOP-lowering benefit may decrease by 1 hour after the procedure
- Anti-glaucoma medications are necessary to maintain IOP control.

Paracentesis will not directly break the pupillary block but can allow the cornea to clear permitting to perform LPI

Possible complications include
- Excessive shallowing of the anterior chamber
- Puncture of iris, lens
- Choroidal effusion
- Haemorrhage due to the sudden decompression

G: Goniosynechialysis
Often performed with other procedures such as lens extraction, to detach synechia from the angle, in eyes with minimal to moderate optic nerve damage. The procedure may be complicated by:
- hyphema
- fibrinous inflammation and
- recurrent synechial closure of the angle
2.4.1.7.3 Acute Angle-Closure (AAC) with plateau iris configuration (See FC VII)

In plateau iris configuration the iris plane is flat and the anterior chamber is not shallow axially. (See above under Staging of Primary Angle-closure).

Medical treatment [II,D]:
- Pupillary constriction to pull the peripheral iris centripetally
- In plateau iris configuration, a modest pupillary constriction may prevent further angle-closure
  - pilocarpine 1%, aceclidine 2%, carbachol 0.75%
  - dapiprazole 0.5%

Surgical treatment [I,D]:
- Iridotomy is essential to confirm the diagnosis because it eliminates any pupillary block component
- Argon Laser Peripheral Iridoplasty (ALPI) stretches the iris and widens the chamber angle

“Plateau iris syndrome” refers to a post-laser iridotomy condition in which a patent iridotomy has removed the relative pupillary block, but gonioscopically confirmed angle closure recurs without central shallowing of the anterior chamber. Isolated plateau iris syndrome is rare compared to the plateau configuration, which itself is not common. It usually occurs in a younger age group than pupillary-block angle-closure. The treatment is laser iridoplasty or the long-term use of pilocarpine postoperatively [II,D]. This condition must be considered in the differential diagnosis when the intraocular pressure rises unexpectedly following an adequate peripheral iridectomy procedure for angle-closure glaucoma.

2.4.1.7.4 Intermittent Angle-Closure (IAC)

Etiology:
Similar but milder clinical manifestations than AAC, it resolves spontaneously.

Pathomechanism:
See above Ch. 2.4.1.5

Features:
- Signs:
  - May vary according to amount of iridotrabecular contact of chamber angle and mimic acute angle-closure in a mild form
  - When not on miotics, pupil is round and reactive
  - The optic disc rim may show atrophy with an afferent pupillary defect

Symptoms:
- Mild, intermittent symptoms of acute angle-closure type
Classification and Terminology

Treatment:
Pupillary constriction, iridotomy, iridoplasty or lens extraction are to be considered according to the main mechanism determining angle occlusion [II,D]

---

**FC VII - Management of Acute Primary Angle Closure Attack**

**Medical Procedures**
- Decrease AH* production
  - Topical therapy: β-blockers / α-agonists
  - Systemic therapy (IV / oral): Acetazolamide / Mannitol (repeat if necessary)
- Re-open the angle
  - Pharmacologically: Pilocarpine 2%
  - Mechanically: Corneal indentation (4 mirror lens)
- Reduce inflammation
  - Topical Steroids

**Laser / Surgical procedures**
- Break pupillary block + re-open angle
  - Iris procedures
    - Clear Cornea ——— Try topical glycerin 10% ——— Cloudy Cornea
      - Iridotomy / Iridecemy
  - Lens extraction

*Remember prophylactic iridotomy in the other eye!

---

* Aqueous Humor

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2.4.1.7.5 Chronic Angle-Closure Glaucoma (CACG) (See FC VIII)

**Etiology:**
Permanent synechial closure of any extent of the chamber angle as confirmed by indentation gonioscopy.

**Pathomechanism:**
See Ch. 2.4.1.5

**Features:**
**Signs:**
- Peripheral anterior synechiae of any degree at gonioscopy
- IOP elevated to a variable degree depending on the extent of iridotrabecular contact, above 21 mmHg
- Visual acuity according to functional status (may be normal)
- Damage of optic nerve head compatible with glaucoma
- Visual field defects "typical" of glaucoma may be present
- Superimposed intermittent or acute iridotrabecular contact possible

**Symptoms:**
- Visual disturbances according to functional states.
- Usually no pain; sometimes discomfort
- Transient "halos" when intermittent closure of the total circumference causes acute IOP elevations

**Treatment:**
Medical treatment alone is contraindicated as all patients require relief of pupil block by iridotomy, iridectomy or lens extraction [I,D]. If the synechial closure is less than half the circumference, iridectomy/iridotomy may be sufficient. Since complications of iridotomy are uncommon, its use as the initial procedure is justified in practically every case [I,D].
Argon laser trabeculoplasty is contraindicated as it may increase synechial angle-closure [I,D]. Lens removal may be considered at all stages and can lead to relief of pupil block and sufficient IOP control [II,D].
If IOP cannot be controlled medically after breaking pupil block (with or without lens extraction), a filtering procedure is indicated [II,D].
These eyes are more frequently prone to develop posterior aqueous misdirection and the necessary precautions must be taken when considering surgery.

2.4.1.7.6 Status Post-Acute Angle-closure Attack

**Etiology:**
Previous episode of acute angle-closure attack

**Pathomechanism:**
See Ch. 2.4.1.5
Classification and Terminology

Features:

Signs:
- Patchy iris atrophy
- Iris torsion/spiralling posterior synechiae
- Pupil either poorly reactive or non-reactive
- “Glaukomflecken” of the anterior lens surface
- Peripheral anterior synechiae on gonioscopy
- Endothelial cell count can be decreased

Therapy:
Management according to angle, lens, IOP and disc/visual field. In case of cataract surgery, non dilatable pupil, low endothelial cell count and loose zonules are of concern.

FC VIII - Management of Chronic Angle Closure

IDENTIFY THE PATHOPHYSIOLOGICAL MECHANISM(s) RESPONSIBLE

Make sure a patent iridotomy is present/made before considering mechanisms other than pupillary block

<table>
<thead>
<tr>
<th>Pupillary Block</th>
<th>Plateau Iris</th>
<th>Lens-induced</th>
<th>Aqueous Misdirection</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Aqueous suppression/vitreous dehydration (acetazolamide/mannitol and/or α-2 agonists and/or β-blockers)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>Pupil constriction (pilocarpine 1-2% or aceclidine 2% or dapiaprazole)</td>
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<tr>
<td>2</td>
<td>Indotommy</td>
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<td>3</td>
<td>Consider lens extraction</td>
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<tr>
<td></td>
<td>Filtration</td>
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</tr>
</tbody>
</table>

Cycloplegia (atropine or cyclopentolate)

Laser capsulotomy Vitreolysis Diode cyclodestruction

Lens extraction

Vitrectomy*

Consider lens extraction

* Combined with zonulectomy+iridectomy in pseudophakia

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2.5 - SECONDARY ANGLE-CLOSURE

There are many different causes of secondary angle-closure and the clinical signs vary according to the underlying condition. For example in secondary acute angle-closure, the chamber angle is closed by iridotrabecular contact that can be reversed, whereas in chronic secondary angle-closure, the angle-closure is irreversible due to peripheral anterior synechiae.

A complete discussion of these topics is outside the scope of this text.

2.5.1 Secondary Angle-Closure With Pupillary Block

Etiology:
The following is a limited list of other etiologies for relative or absolute pupillary block:
- Enlarged, swollen lens (cataract, traumatic cataract)
- Anterior lens dislocation (trauma, zonular laxity; Weil-Marchesani’s syndrome, Marfan’s syndrome etc.)
- Posterior synechiae, seclusion or occlusion of the pupil
- Protruding vitreous face or intravitreal silicone oil in aphakia
- Microspherophakia
- Miotic-induced pupillary block (also the lens moves forward)
- IOL-induced pupillary block; anterior chamber IOL, phakic intraocular lens (PIOL), anteriorly dislocated posterior chamber intraocular lens (PC-IOL)

Pathomechanism:
Pupillary block pushes the iris forward to occlude the angle. In iritis or iridocyclitis, the development of posterior synechiae may lead to absolute pupillary block with consequent forward bowing of the iris or “iris bombé”. Acute secondary angle-closure glaucoma may result.

Features:
- IOP > 21 mmHg
- Disc features compatible with glaucoma

Treatment:
Several steps may be considered, according to the clinical picture of causative mechanisms [II,D]
- Topical and systemic IOP lowering medication
- Nd:YAG laser iridotomy
- Peripheral surgical iridectomy
- Lens extraction, vitrectomy
- Discontinuing miotics in miotic-induced pupillary block
- Pupillary dilation
- Nd:YAG laser synechiolysis of posterior synechiae
2.5.2 Secondary Angle-Closure With Anterior “Pulling” Mechanism Without Pupillary Block

Pathomechanism:
The trabecular meshwork is obstructed by iris tissue or a membrane. The iris and/or a membrane are progressively pulled forward to occlude the angle.

Features:
- IOP>21 mmHg
- Disc features compatible with glaucoma

2.5.2.1 Neovascular glaucoma

The iridotrabecular fibrovascular membrane is induced by ocular microvascular disease with retinal ischemia; initially the neovascular membrane covers the angle, causing a secondary form of open angle glaucoma (See Ch 2.3 Secondary Open Angle Glaucoma)

Treatment [II,D]:
- a) Topical atropine or equivalent
- b) Topical steroid initially
- c) Topical and systemic IOP lowering medication as needed
- d) Retinal ablation with laser or cryotherapy
- e) Cyclodestruction
- f) Filtering procedure with antimetabolites
- g) Aqueous drainage devices
- h) Miotics are contraindicated

The intravitreal injection of anti-VEGF molecules has shown benefit for this indication [II,C] and is in widespread use.

2.5.2.2 Iridocorneal endothelial syndrome

Iridocorneal Endothelial (ICE) Syndrome, with progressive endothelial membrane formation and progressive iridotrabecular adhesion.
Peripheral anterior synechiae, due to prolonged primary angle-closure; this is theoretically a primary angle-closure.

Treatment [II,D]:
- a) Topical and systemic IOP lowering medications as needed
- b) Filtering procedure, with antimetabolite according to risk factors
- c) Aqueous drainage device
2.5.2.3 Posterior polymorphous dystrophy

Treatment [II,D]:
- a) Topical and systemic IOP lowering medication as needed
- b) Filtering procedure, with antimetabolite according to risk factors

2.5.2.4 Epithelial and fibrous ingrowth after anterior segment surgery or penetrating trauma

Epithelial and fibrous ingrowth after anterior segment surgery or penetrating trauma
Inflammatory membrane.

Treatment [II,D]:
- a) Topical and systemic IOP lowering medication as needed
- b) Excision, destruction of the immigrated tissue
- c) Filtering procedure, with antimetabolite according to risk factors
- d) Aqueous drainage device
- e) Cyclodestruction

2.5.2.5 Inflammatory membrane

Treatment [II,D]:
- a) Anti-inflammatory medications and cycloplegics
- b) Topical and systemic IOP lowering medication as needed
- c) Filtering procedure with antimetabolite
- d) Aqueous drainage device
- e) Cyclodestruction

2.5.2.6 Peripheral anterior synechiae after ALT and endothelial membrane covering the trabecular meshwork late after ALT

After argon laser trabeculoplasty (ALT), early and late peripheral anterior synechiae and endothelial membrane covering the trabecular meshwork

Treatment [II,D]:
- a) Topical and systemic IOP lowering medication as needed
- b) Filtering procedure

2.5.2.7 Aniridia

Treatment [II,D]:
- a) Topical and systemic IOP lowering medication as needed
- b) Trabeculotomy
2.5.3 Secondary Angle-Closure With Posterior ‘Pushing’ Mechanism Without Pupillary Block

2.5.3.1 Aqueous misdirection (also known as cilio-lenticular block, ciliary block or malignant glaucoma)

Etiology:
Angle-closure is caused by the ciliary body and iris rotating forward. Aqueous misdirection, or malignant glaucoma, is a rare type of secondary angle-closure glaucoma most commonly encountered after filtering surgery. The syndrome, also known as ciliary block glaucoma, can occur spontaneously or following any type of intraocular surgery.

Pathomechanism:
- The lens may be proportionally abnormally large or swollen, “phacomorphic glaucoma”
- Aqueous humour accumulates in the vitreous body (posterior aqueous humour misdirection) or behind and around the crystalline lens (perilenticular misdirection) or behind the iridocapsular diaphragm or posterior chamber intraocular lens (PCL) after extracapsular cataract surgery, with or without PCL implantation, “retrocapsular misdirection”
- Frequently precipitated by ocular surgery and flat anterior chamber
- Predisposition may be similar in both eyes particularly in small eyes

Treatment:
Medical treatment
- Parasympatholytics (atropine, cyclopentolate) both initially and for long-term pupillary dilation and cycloplegia [I,C]
- Aqueous production suppressants given orally and/or topically [I,D]
- Hyperosmotics (Ch. 3.3.1.3) [I,D]
- Miotics are contraindicated!

Surgical treatment
- A patent iridotomy must be present or, if not present, iridotomy should be performed [I,D]
- YAG laser vitreolysis/capsulotomy, especially in aphakia, pseudophakia [II,C]
- Anterior vitrectomy, especially in aphakia, pseudophakia [II,C]
- Cyclo diode laser
- In selected cases lens extraction [II,D]
2.5.3.2 Iris and ciliary body cysts, intraocular tumors

Treatment:
- a) Tumour irradiation or excision
- b) Filtering surgery
- c) Cyclodestruction

2.5.3.3 Silicon oil or other tamponading fluids or gas implanted in the vitreous cavity

Treatment:
- a) Topical/systemic IOP lowering medications as needed
- b) Silicon oil or gas aspiration
- c) Filtering surgery
- d) Drainage device
- e) Cyclodestruction

2.5.3.4 Uveal effusion

It is due to:
- Inflammation as in scleritis, uveitis, HIV infection
- Increased choroidal venous pressure as in nanophthalmos, scleral buckling, panretinal photocoagulation, central retinal vein occlusion, arterio-venous communication
- Tumor

Treatment:
- a) Anti-inflammatory medication (for 1)
- b) Topical and systemic IOP lowering medication as needed (for 1, 2 and 3)
- c) Relaxation of scleral buckling; vitrectomy, sclerectomy in nanophthalmus (for Tumor excision or irradiation (for 3)
- d) Cyclodestruction

2.5.3.5 Retinopathy of prematurity (stage V)

Features:
  Signs and Symptoms:
  - Variable discomfort, pain, redness, corneal oedema IOP ≥ 21 mmHg
  - Axially shallow anterior chamber

Treatment:
- a) Topical and systemic IOP lowering medications
- b) Cyclodestruction
- c) Filtering procedure with or without antimetabolite
- d) Drainage devices
2.5.3.6 Congenital anomalies that can be associated with secondary glaucoma

These conditions are extremely variable in pathogenesis, clinical presentation and required management; an extensive discussion is outside the scope of this chapter.

**Etiology:**
Familial iris hypoplasia, anomalous superficial iris vessels, aniridia, Sturge-Weber syndrome, neurofibromatosis, Marfan's syndrome, Pierre Robin syndrome, homocystinuria, goniodysgenesis, Lowe's syndrome, microcornea, microspherophakia, rubella, broad thumb syndrome, persistent hyperplastic primary vitreous.

**Pathomechanism:**
Angle-closure is caused by pushing forward the ciliary body and iris.
Increase of volume of the posterior segment of the eye.

**Features:**
- **Signs and Symptoms:**
  - IOP > 21 mmHg
  - Pain, redness, corneal oedema
  - Axially shallow anterior chamber
  - Laser iridotomy and surgical iridectomy are not effective

**Some differential diagnoses:**
- Acute IOP elevation with corneal oedema but open-angle may result from Posner Schlossman syndrome (iridocyclitic crisis), or from endothelitis/trabeculitis as in disciform herpetic keratitis.
- Neovascular glaucoma may be associated with an open or closed-angle and may mimic some signs and the symptoms of acute angle-closure.

**Treatment:**
Treatment to be adapted to the primary anomaly, the mechanism of IOP elevation and the quality of life of the patient.
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