

# Classification Of Primary Angle Closure Disease

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Primary angle-closure disease (PACD) is the leading cause of irreversible blindness in East Asia and is responsible for half of the glaucoma related blindness.<sup>1</sup>

The classification of PACD is often considered confusing, due to inconsistencies in terminology and nomenclature various described and recently used classifications are following.

## Classification of PACD :

Angle closure or narrow angles are the essential components of PACD, while elevated intraocular pressure (IOP) is secondary to angle closure. PACD can be classified in different ways, based on clinical presentation, natural history, anatomy, etiology, etc.

### Clinical classification/ Clinical Spectrum of PACD :

It is based on the clinical course of primary angle closure glaucoma (PACG) and therefore, it revolves around the course of the disease and time of onset.<sup>2</sup>

- a) *Latent PACG*: These are the eyes where pigmented trabecular meshwork is not visible in more than 2 quadrants without indentation or manipulative gonioscopy. There are no other symptoms or signs like any evidence of gonioscopic abnormalities and raised IOP.
- b) *Subacute / Intermittent PACG*: These are the eyes with sudden closure of entire angle for a short period of time under some physiological factors like reading in dim light, entering into a darkened room etc. but spontaneous resolution of pupillary block is the rule. Therefore, these have prodromal symptoms of mild headache, blurred vision, colored haloes that resolve spontaneously. Anterior segment may show subtle signs of angle closure like pupillary ruff atrophy, iris atrophic patches etc. and Gonioscopic abnormalities like patchy pigmentation/synechia might be present but with normal IOP in interparoxysmal phase.<sup>2</sup>
- c) *Acute PACG*: In this stage the sudden complete closure of entire angle leads to sudden rise of IOP to very high levels. This results in the sudden onset of classical symptoms of redness, pain, watering, photophobia,

colored haloes and classical signs of circumciliary congestion, corneal edema, mid dilated pupil, glaukomaflecken etc.

- d) *Chronic PACG*: As the name suggests, the closure of trabecular meshwork by the iris is slow and steady. This may be due to creeping angle closure or following acute PACG or due to recurrent subacute attacks. These eyes have chronically raised IOP with PAS in more than two quadrants. As these eyes have enough time to accommodate for raised IOP, therefore the symptoms are not at all dramatic.

The main concern in this classification is that at certain occasions subjects with “latent angle closure glaucoma” have been classified as cases of established glaucoma, despite having normal visual function. This resulted into misinterpretation of the estimates of visual morbidity attributable to glaucoma.<sup>3</sup> Moreover, difference in the definition of PACG and POAG made it difficult to compare the prevalence and study risk factors in epidemiological glaucoma research.<sup>4</sup>

### Pathogenic classification:

This classification is based on anatomical levels of obstruction to aqueous flow in primary and secondary angle closure glaucoma.<sup>5</sup> According to this the angle closure may be due to forces acting at four anatomical levels:

- a) *Level I*: The forces acting at the level of iris that includes-
  - pupillary block
  - non-pupillary block
  - angle crowding mechanisms
- b) *Level II*: The forces acting at the level of ciliary body, including plateau iris configuration and iridociliary cysts.
- c) *Level III*: The forces acting at the level of lens, including thick, anteriorly positioned and subluxated lens.
- d) *Level IV*: The vector forces posterior to lens – that includes aqueous misdirection, choroidal effusion, space-occupying lesions etc.

However, clinical application of this classification is difficult at various occasions as in many ACG patients multiple “levels” may simultaneously or consecutively play a role. Moreover, estimate of visual morbidity cannot be correlated.

Thus, it was thought that a full re-evaluation of the definition is mandatory, with emphasis on visual morbidity

rather than symptomatic disease or pathogenic mechanism.

Hence, At the biennial congress of the International Society for Geographical and Epidemiological Ophthalmology (ISGEO) held at Leeuwenhorst, the Netherlands, in June 1998, a group interested in glaucoma epidemiology met to propose the new Epidemiological classification based on progression of the disease.<sup>4</sup> The recent American Academy of Ophthalmology (AAO) guidelines also follows the similar classification.<sup>3</sup>

### **Epidemiological classification**

The fundamental concept of this classification is that the term 'glaucoma' is reserved for people with established, visually significant and with end organ damage.<sup>4</sup> So, this classification can also be used conveniently for cross sectional epidemiological research.

(a) *Primary angle closure suspect (PACS)*: PACS was defined as an eye with occludable angle (pigmented trabecular meshwork not visible for  $\geq 180^\circ$  under static gonioscopy without peripheral anterior synechiae, PAS) and IOP lower than 21 mmHg, and no glaucomatous optic neuropathy (GON).

(b) *Primary angle closure (PAC)*: PAC is defined as an eye with an occludable angle and gonioscopic features indicating that trabecular obstruction by the peripheral iris has occurred (e.g. iris whorling, PAS, "glaucomflecken", lens opacities or excessive pigment deposition on the trabecular surface) or raised IOP ( $>21$  mm Hg) but the optic disc does not have GON.

Thus, in this new concept, PAC includes both asymptomatic people with occludable angles who either have not or have had an acute attack, that was treated promptly but suffered no detectable GON damage.

(c) *PACG*: These were the eyes with PAC and GON (defined

as a vertical cup/disc (C/D) ratio  $>0.7$  and/or C/D asymmetry  $>0.2$  and/or focal notching), with compatible visual field loss on static automated perimetry.

This classification is not intended to indicate that those with PAC do not require treatment. It is intended to differentiate between those with and without damaged visual function attributable to GON. Moreover, it is seen that both are likely to benefit from iridotomy, but the former (PAC) are likely to be cured, while the later will require more intensive follow up and treatment much like the treatment for POAG.<sup>2,4</sup>

To summarize, the epidemiological classification has been accepted universally and is used to classify PACD. It differentiates well between those with and without visual morbidity and therefore assists in defining the treatment protocol.

### **References:**

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## **Novel Cyclosporine A Formulation shows promise in patients with Vernal Keratoconjunctivitis**

Investigators of this phase 3 trial assessed the efficacy and safety of an investigational cationic emulsion Cyclosporine A (CsA CE) for severe vernal keratoconjunctivitis. They randomized 169 pediatric patients to a high or low dose of CsA CE drops or a vehicle control. Both treatment groups showed significant improvements over the vehicle group in corneal fluorescein staining scores and rescue Dexamethasone use. Additionally, the high-dose arm reported improved keratoconjunctivitis symptoms and quality of life compared with vehicle. The novel formulation was well tolerated with no unexpected safety findings. *Ophthalmology*, May 2019