

# Primary chronic angle-closure glaucoma in younger patients

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## Abstract

• **AIM:** To evaluate the clinical outcomes of management in younger patients with primary chronic angle-closure glaucoma (PCACG).

• **METHODS:** Thirty-eight patients (50 eyes) aged 40 or younger with confirmed diagnosis of PCACG in advanced or late stage who received surgical treatment in Zhongshan Ophthalmic Center from January 2000 to December 2005 were retrospectively investigated. All patients underwent trabeculectomy. The mean follow-up was  $23.6 \pm 7.5$  months. Full ophthalmic examinations were performed. The clinical outcomes including clinical presentations, surgical results and complications were evaluated.

• **RESULTS:** The mean age of patients was  $33.5 \pm 6.1$  years old. There was a female preponderance (60.5%). The mean axial length was  $22.4 \pm 3.5$ mm with 18.0% short axis of eyeball and 14% nanophthalmos. There was 60.0% flat anterior chamber depth ( $<1.9$ mm). Ultrasonic Biomicroscopy identified that plateau iris was the most common underlying etiology (80.6%). There was a statistically significant difference in intraocular pressure (IOP) reduction postoperatively *vs* preoperatively ( $P < 0.001$ ). Four eyes failed to control IOP and received second filtration surgery. The main postoperative complications included shallow anterior chamber (20.0%) and malignant glaucoma (12.0%).

• **CONCLUSION:** The younger PCACG patients in advanced or late stage can be effectively managed by trabeculectomy. They have more frequency of postoperative sustained shallow anterior chamber and malignant glaucoma. Careful ophthalmic examinations, delicate surgical procedures and well-managed

technique of complications were suggested on younger PCACG patients.

• **KEYWORDS:** primary chronic angle-closure glaucoma; young; plateau iris; trabeculectomy

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## INTRODUCTION

Glaucoma is the second leading cause of vision loss worldwide, affecting over 50 millions people over the world, almost one third of whom become blind<sup>[1]</sup>. Primary angle-closure glaucoma (PACG) disproportionately affects Asians<sup>[2,3]</sup>. In China alone, about 4.5 millions people have PACG, and PACG accounts for the majority of bilateral glaucoma blindness in China<sup>[3,4]</sup>. PACG can be acute and chronic. The primary chronic angle-closure glaucoma (PCACG) has more tendency threatening elder persons<sup>[1-5]</sup> and is rare in young adults<sup>[5-7]</sup>. Age  $\leq 40$  is defined as younger patient as the age is the onset of presbyopia<sup>[5]</sup>. The younger individual with PCACG is typically associated with structural ocular anomalies that have short axis of eyeball, shallow or flat anterior chamber and narrower drainage angle. PCACG can be effectively managed by trabeculectomy when disease progresses even though it is potentially preventable if peripheral iridectomy is performed in the early stage. Malignant glaucoma is a rare complication following trabeculectomy<sup>[8-10]</sup>. It is characterized with flattening of anterior chamber and elevation of intraocular pressure (IOP). With unusual or abnormal situation of anterior segment, the younger PCACG has a potential risk of malignant glaucoma following filtering surgery<sup>[8-12]</sup> and has poor surgical success rate<sup>[7]</sup>. A careful examination before surgical intervention, excellent surgical procedure and properly postoperative management could promote best surgical results and reduce the risk of complications to minimum. In this study, we retrospectively investigated the clinical outcomes on a group of younger PCACG patients.

## MATERIALS AND METHODS

**Subjects** Thirty-eight PCACG (50 eyes) who were hospitalized to receive trabeculectomy in Glaucoma Division, Zhongshan Ophthalmic Center between January 2000 and December 2005 were retrospectively investigated. All cases were 40 years old or younger with mean age  $33.5 \pm 6.1$  years (ranged from 22 to 40 years). There were 15 males (39.5%) and 23 females (60.5%). The follow-up ranged from 8 to 70 months with an average of  $23.6 \pm 7.5$  months. All cases were diagnosis as advanced stage or late stage which was in conformity with current diagnostic criteria<sup>[13-16]</sup>. The anterior chamber angle closure was confirmed larger than 180 circumference under the gonioscopy (Haaf streit, Bern, Switzerland) and Ultrasound biomicroscopy (UBM, Humphrey 840); the vertical cup-disc ratio was 0.6-1.0 with a mean  $0.71 \pm 0.22$ . IOP was measured by Goldmann applanation tonometry; the axial length was measured by B-scan ultrasonography; the visual field was taken by Humphrey 750 automated static threshold perimeter. Significant improvement of visual acuity was defined as increasing by 2 or more lines. Either topical or systemic IOP-lowering agent was represented as one number of medications.

**Methods** All trabeculectomies had been performed by the skillful surgeons: A limbus-based conjunctival flap was obtained. A 3mm×4mm size with 1/2 thickness scleral flap was dissected; Mitomycin C (MMC, 0.25g/L) was applied for 3-5 minutes beneath the conjunctival and/or scleral flap. Then 150-200mL balanced salt solution (BSS) was irrigated. A small cornea incision (anterior paracentesis) was made into the anterior chamber. Then a 1.5mm×2mm deep scleral flap was cut and an iridectomy was followed. The scleral flap was closed with 2 releasable sutures and 2 fixed sutures. The anterior chamber was reformed with balanced salt solution through the incision of anterior paracentesis.

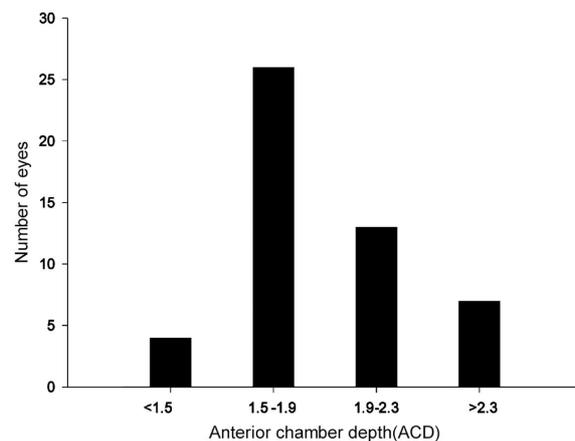
**Statistical Analysis** Software SPSS 11.0 was used. The values were presented as mean  $\pm$ SD of the mean. The comparison of preoperative and postoperative IOP, ACD was tested with paired *t* test. The change of VA was compared by  $\chi^2$  test. All data was taken as a significant difference with  $P < 0.05$ .

## RESULTS

**IOP and IOP -lowering Medications** The mean preoperative IOP was  $42.1 \pm 12.3$  mmHg, the average number of topical and/or systemic antiglaucomatous agents was  $2.3 \pm 1.2$ . The postoperative IOP at last visit was  $12.1 \pm 4.6$  mmHg with adjunctive  $0.4 \pm 0.1$  medications. The difference between preoperative and postoperative IOP was statistically significant ( $t = 15.91$ ,  $P < 0.001$ ). During the follow-up, 4 eyes

**Table 1** The clinical data of nanophthalmos, axial length and refraction in 7 eyes

Age sex	Eye	Axial length(mm)	Refraction(D)
36/M	OS	19.75	+2.00
37/M	OD	20.08	0.00
37/M	OS	19.50	+3.00
36/M	OD	19.50	+6.00
36/M	OS	19.65	+2.25
29/F	OS	20.00	0.00
28/F	OS	19.80	+3.00



**Figure 1** The distribution of preoperative anterior chamber depth in all eyes

failed to control IOP with at least 4 numbers of medications and thus received second trabeculectomy. The mean follow-up of these 4 eyes after second intervention was  $6.2 \pm 1.5$  months; the mean IOP at last visit was  $12.0 \pm 2.2$  mmHg, in which 1 eye adjunctive with topical IOP-lowering agent to drop down IOP to 15.0 mmHg.

**Axial Length and Refractive Error** The mean axial length was  $22.4 \pm 3.5$  mm, in which 9 eyes were  $< 21$  mm (18.0%) including 7 (14.0%) nanophthalmos. The mean refractive error in these 7 nanophthalmos eyes was  $+2.3 \pm 2.0$  diopters (range +0 to +6, Table 1).

**ACD and Anterior Chamber Angle** The mean ACD measured was  $1.9 \pm 0.4$  mm, in which 4 eyes  $< 1.5$  mm, 26 eyes 1.5-1.9 mm, 13 eyes 1.9-2.3 mm and 7 eyes  $> 2.3$  mm. The flat anterior chamber accounted for 60.0% ( $< 1.9$  mm) (Figure 1). The mean postoperative ACD was  $1.9 \pm 0.2$  mm. By comparing the difference of ACD between preoperative and postoperative, there was no significance ( $t = 0.142$ ,  $P > 0.05$ ). Thirty-eight eyes underwent UBM examination. There were 29 eyes presenting plateau iris configuration with anteriorly positioned ciliary processes (80.6%, Figure 2), 7 eyes (19.4%) were pupillary block (Figure 3) and 2 eyes present mixed two elements of angle appearance. The anterior chamber angles were not seen significantly changes postoperatively vs preoperatively (Figure 4A, B). The filtering bleb was well observed under the UBM (Figure 4C).



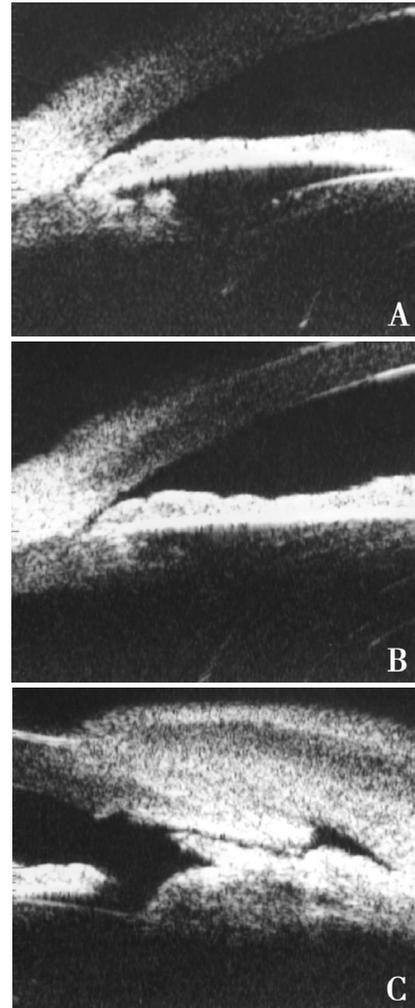
**Figure 2 Plateau iris configuration was demonstrated by UBM**  
The ciliary body touched the posterior iris. The anterior chamber was relatively deep, the iris contour was essentially planar, the posterior chamber was very small, and the ciliary's processes were centrally displaced



**Figure 3 Pupillary block was seen between the lens and iris**

The pressure in the posterior chamber was increased with forward shift of the peripheral iris and closure of the anterior chamber angle. UBM showed that the iris contour was convex, the posterior chamber distended, and a prominent ciliary's sulcus was present.

**Complications and others** No vitreous prolapsed happened during trabeculectomy in all cases. No filtering bleb leakage, bleb perforation or bleb infection occurred postoperatively. Three eyes sustained hypotonus (between 5 and 7mmHg)  $4.2 \pm 1.3$  months, in which 2 eyes formed thin-wall bleb. Ten eyes (20.0%) presented shallow anterior chamber, in which accompanied with choroidal detachments 1 eye, malignant glaucoma 6 eyes (12.0%). We identified that the corresponding ACD in these 6 malignant glaucoma were between 1.5 mm and 1.9 mm in 5 eyes, <1.5mm in 1 eye. Three in 6 eyes were relieved after medication treatments and IOP was controlled with adjunctive 2 numbers of antiglaucomatous agents. The other 3 eyes received surgical managements including aspiration of fluid



**Figure 4 The UBM images before and after trabeculectomy**  
The anterior chamber angle was not seen significant changes postoperatively (B) vs preoperatively (A). The functional filtering bleb was well formed postoperatively (C)

from the vitreous by a pars plana incision in 1 eye, anterior pars plana vitrectomy in 1 eye, phacoemulsification plus intraocular len implantation (Phaco+ IOL) and anterior vitrectomy in 1 eye. 10g/L Atropine solution was applied in 3 eyes for  $13.3 \pm 3.2$  months to maintain ACD to  $2.8 \pm 0.3$ mm, 1 eye was plus adjunctive with Tropicamide 8 months to maintain the anterior chamber to 3.1mm. (Table 2, Figure 5).

**Visual Acuity and Visual Field** Compared postoperatively vs preoperatively, the VA at last visit did not change significantly ( $\chi^2 = 55.02, P > 0.05$ ): decreased in 6 eyes (12.0%), remained in 40 eyes (80.0%) and improved in 4 (8.0%). Prior to surgery, no lens opacity was seen in all eyes. However, during the postoperative follow-up, 3 eyes developed visible cataract. One of them underwent Phaco+IOL when malignant glaucoma coincidentally happened in this eye afterwards. Twenty-two eyes underwent visual field examination before and after surgery. Most of



**Figure 5 10g/L atropine instillation for malignant glaucoma** The pretreatment (A), after Atropine instillation (B, C). The anterior chamber depth was deeper after mydriasis treatment (C)

**Table 2 The clinical information of malignant glaucoma in 5 cases (6 eyes)**

<i>n</i>	Age/Sex	Eye	Postoperative occurring time	Management	ACD (mm)	IOP (mmHg)
1	37/F	OD	23 <sup>rd</sup> mo	10g/L Atropine 17months	2.6	9.0
		OS	21 <sup>st</sup> mo	Phaco+IOL plus Anterior vitrectomy	3.2	12.0
2	37/M	OD	1 <sup>st</sup> mo	Anterior pars plana vitrectomy	2.5	10.0
3	40/F	OD	1 <sup>st</sup> wk	10g/L Atropine 7months	2.8	11.5
				When stopped 1month	1.8	14.6
				10g/L Atropine again 4 months	2.5	15.0
				10g/L Atropine again 8 months plus Tropicamide 8 months	3.1	18.0
4	28/F	OS	1 <sup>st</sup> wk	10g/L Atropine 12months	2.6	7.0
5	22/F	OS	2 <sup>nd</sup> wk	Aspiration of fluid from the vitreous by a pars plana incision	2.3	12.5

them didn't show significant progression during follow-up, however, 4 eyes presented visual field deterioration.

## DISCUSSION

Primary chronic angle-closure glaucoma (PCACG) is a disease of elders [1-5], and it is rare in children and young adults [5-7]. The mechanisms causing angle closure glaucoma in PCACG can be pupillary-block, plateau iris, and mixed mechanism [5,12-18]. Approximately 90.0% of elder individuals with angle closure have pupillary block as the underlying mechanism while hyperopic eyes have more tendency [5,19]. Laser or surgical peripheral iridectomy can successfully manage the situation when disease was in early stage. Plateau iris is illustrated as an angle appearance in which the iris roots angulated forward and centrally and inserted anteriorly on the ciliary face [18]. The angle is shallow and narrow; the iris contour is planar; the anterior chamber depth within the normal range. Patients with plateau iris tend to be female, younger and less hyperopic than those with pupillary block angle-closure glaucoma [5]. Some of patients with plateau iris could have an element of pupillary block simultaneously.

Age 40 or younger was defined as "younger" patients because of the onset of presbyopia [5]. Anterior chamber narrowed with increasing age and hyperopia. Thus, in younger patients the mechanism of angle closure differs from those in elder patients in frequency. Younger person has characteristics in short axis of eyeball, shallow or flat

anterior chamber angle, thick lens and forward positioned ciliary [5-7]. The Lowe's coefficient <0.19 or the depth of anterior chamber <1.9 mm was once indicated in the textbook as a predisposing factor for happening angle closure or malignant glaucoma following filtering surgery [8-12]. The data from our study was similar with that described in literatures [5-7]: there was a clear female frequency (60.5%); 18.0% has shorter axial length; 14.0% were nanophthalmos; 60.0% has flat anterior chamber (<1.9 mm); plateau iris was the predominant underlying mechanism (80.6%).

The complications following trabeculectomy surgery in PCACG were reported in literatures [10] as scarring of the filtering bleb (47.6%), choroidal detachment (18.6%), shallowing of the anterior chamber (8.0%), fibrinous reaction (5.3%), overfiltration (4.4%), reduced flow under the scleral flap (4.4%), corneal dellen (3.5%), prolapse of the iris (3.5%), external fistulation (2.7%), increase of the IOP due to steroids (1.8%), detachment of the ciliary body (1.8%) and malignant glaucoma (0.9%). Our data presented that only 4 eyes (8.0%) failed to control IOP due to scarring filtering bleb and received second surgical treatment. Mitomycin C (MMC) application had been more standardization and rationalization nowadays in antiglaucomatous surgeries, however, there were still some eyes happening hypotony in our study. It implied that the reaction to MMC surely had individual difference. Although adjunctive MMC

enhance the success of trabeculectomy, their risk/benefit ratio should be assessed for each individual patient prior to use. Malignant glaucoma is a rare complication following trabeculectomy and is reported to complicate 0.6% to 4.0% [8-10]. In our patients, there were 20.0% shallow anterior chamber and 12.0% malignant glaucoma which were much higher than that described in the literatures [8-10].

Preoperative ophthalmic examinations including axial length, lens thickness and ACD are necessary for management younger PCACG. Gonioscopy and UBM were practical technique to diagnosis and follow-up [20,21]. Careful and exquisite surgical procedures could reduce the complications to minimum. With our experiences, a limbus-based conjunctival flap and a relative bigger and thicker scleral flap (3-4mm×4-5mm, 1/2-2/3 thickness) and a larger iridectomy were recommended for flat anterior chamber individuals. A paracentesis was suggested before deep layer scleral flap excision. There were two purposes to perform paracentesis: to release the preoperative IOP so as to help performing following procedure; and to help judge if leakage when the anterior chamber was reformed. Deep scleral flap was excised to clear cornea to prevent unexpected ciliary body bleeding. Scleral flap was sutured tightly and left two releasable sutures and this way could decrease the chance of postoperative shallow anterior chamber in early stage. Finally, after closing the scleral flap, balanced salt solution was injected to anterior chamber through incisional paracentesis to judge if the anterior chamber stable and aqueous humor leakage lightly.

The mydriatic-cycloplegic treatment could effectively prevent shallow anterior chamber sustained and malignant glaucoma happening as it could pull the lens back by tightening the zonules and break a ciliary block. With 10g/L atropine and tropicamide together, we found it very helpful to maintain the anterior chamber and prevent recurrences. Surgical interventions including aspiration of fluid from the vitreous by a pars plana incision, anterior pars plana vitrectomy or posterior continuous curvilinear capsulorhexis (PCCC) and anterior vitrectomy and/or phacoemulsification plus intraocular lens implantation (Phaco+IOL) were alternative approaches for malignant glaucoma [8,9,22-25]. Phacoemulsification could successfully manage malignant glaucoma in some extent as removal of the lens could release the causes of misdirection of aqueous humor [22]. Our patients received these treatments and achieved good surgical outcomes.

In summary, the current study showed that the younger PCACG patients in advanced or late stage were effectively managed by trabeculectomy even though the mechanisms causing angle closure differ from those in elder patients in frequency. These kinds of patients have a relatively high risk of postoperative shallow anterior chamber and malignant

glaucoma. A careful ophthalmic examination, delicate surgical procedure and well-managed technique of complications were suggested on younger PCACG patients to prevent complications to minimum.

#### REFERENCES

- 1 Quigley HA. Number of people with glaucoma worldwide. *Br J Ophthalmol* 1996;80(5):389-393
- 2 Quigley HA, Broman AT. The number of people with glaucoma worldwide in 2010 and 2020. *Br J Ophthalmol* 2006;90(3):262-267
- 3 Foster PJ, Johnson GJ. Glaucoma in China: how big is the problem? *Br J Ophthalmol* 2001;85(11):1277-1282
- 4 Congdon NG, Friedman DS. Angle-closure glaucoma: impact, etiology, diagnosis, and treatment. *Curr Opin Ophthalmol* 2003;14(2):70-73
- 5 Ritch R, Chang BM, Liebmann JM. Angle closure in younger patients. *Ophthalmology* 2003;110:1880-1889
- 6 Fivgas GD, Beck AD. Angle-closure glaucoma in a 10-year-old girl. *Am J Ophthalmol* 1997;124(2):251-253
- 7 Sidorov EG, Perchikova OI. Characteristics of trabeculectomy in the treatment of some forms of glaucoma in young patients. *Vestn Oftalmol* 1992;108(2):5-7
- 8 Lowe RF. Malignant glaucoma related to primary angle closure glaucoma. *Aust J Ophthalmol* 1979;7:11
- 9 Fellman R, Budenz D. Malignant glaucoma. *J Glaucoma* 1999;8(2):149-153
- 10 Picht G, Mutsch Y, Grehn F. Follow-up of trabeculectomy. Complications and therapeutic consequences. *Ophthalmologe* 2001;98(7):629-634
- 11 Razeghinejad MR, Amini H, Esfandiari H. Lesser anterior chamber dimensions in women may be a predisposing factor for malignant glaucoma. *Med Hypotheses* 2005;64(3):572-574
- 12 Quigley HA, Friedman DS, Congdon NG. Possible mechanisms of primary angle-closure and malignant glaucoma. *J Glaucoma* 2003;12(2):167-180
- 13 He M, Foster PJ, Johnson GJ, Khaw PT. Angle-closure glaucoma in East Asian and European people. Different diseases? *Eye* 2006;20(1):3-12
- 14 Foster PJ, Oen FT, Machin DS, Ng TP, Devereux JG, Johnson GJ, Khaw PT, Seah SK. The prevalence of glaucoma in Chinese residents of Singapore. A cross-sectional population survey in Tanjong Pagar district. *Arch Ophthalmol* 2000;118:1105-1111
- 15 Glaucoma Division, Chinese Society of Ophthalmology. The diagnostic guideline for early diagnosis of primary glaucoma. *Chin J Ophthalmol* 1987;23:127
- 16 Lowe RF. Clinical types of primary angle closure glaucoma. *Aust N Z J Ophthalmol* 1988;16(3):245-250
- 17 Lee DA, Brubaker RF, Ilstrup DM. Anterior chamber dimensions in patients with narrow angles and angle-closure glaucoma. *Arch Ophthalmol* 1984;102:46-50
- 18 Ritch R. Plateau iris is caused by abnormally positioned ciliary processes. *J Glaucoma* 1992;1:23-26
- 19 Van Rens GH, Arkell SM. Refractive errors and axial length among Alaskan Eskimos. *Acta Ophthalmol* 1991;69:27
- 20 Schroeder W, Fischer K, Erdmann I, Guthoff R. Ultrasound biomicroscopy and therapy of malignant glaucoma. *Klin Monatsbl Augenheilkd* 1999;215(1):19-27
- 21 Pereira FA, Cronemberger S. Ultrasound biomicroscopic study of anterior segment changes after phacoemulsification and foldable intraocular lens implantation. *Ophthalmology* 2003;110(9):1799-1806
- 22 Sharma A, Sii F, Shah P, Kirkby GR. Vitrectomy-phacoemulsification-vitrectomy for the management of aqueous misdirection syndromes in phakic eyes. *Ophthalmology* 2006;113(11):1968-1973
- 23 Ge J, Guo Y, Liu Y, Lin M, Zhuo Y, Chen B, Chen X. New management of malignant glaucoma by phacoemulsification with posterior chamber foldable intraocular lens implantation. *Yanke Xuebao* 1999;15(3):162-168
- 24 Harbour JW, Rubsamen PE, Palmberg P. Pars plana vitrectomy in the management of phakic and pseudophakic malignant glaucoma. *Arch Ophthalmol* 1996;114(9):1073-1078
- 25 Hu N, Gong Q, Guan H. Mechanism and treatment of malignant glaucoma. *Chin Med J* 1998;78(3):225-226