·Clinical Research·

A comparison study of the clinical features between Chinese and Indian primary congenital glaucoma patients

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Abstract

• AIM: To summarize the clinical features of Chinese primary congenital glaucoma (PCG) and to investigate the discrepancies of the clinical features between the Chinese and Indian patients.

• METHODS: Clinical records of 40 Chinese PCG patients were reviewed. The clinical features were summarized as several quantifiable clinical parameters and the severity of the disease was evaluated. Both the quantified clinical features and severity were statistically compared with those of the Indian patients, which were cited from the previously published articles.

• RESULTS: Forty Chinese and 43 Indian patients were included in the study. In Chinese patients, sex ratio (male to female)was about 2:1, family history was presented in 3 patients (7.5%) and consanguinity was found in one patient (2.5%). The main symptoms and signs observed in Chinese patients spanned a wide spectrum of manifestations. The most frequent signs noted in the initial examination were enlarged eyeball (42.5%) and decreased visual acuity (VA) (35.0%). Compared with Indian patients, Chinese patients had a later onset, a delayed diagnosis , more severe corneal changes and more severe optic nerve damages (P <0.01). The combined trabeculectomy and trabeculotomy operation was preferred by both Chinese and Indian doctors whereas a higher proportion of Indian patients received the combined operation (P<0.01). The proportions of the severity grade were different between Chinese and Indian patients. Most Chinese patients were in the severe grade while most Indian patients were in the very severe grade (P <0.01).

• CONCLUSION: Chinese PCG patients were sporadic and non-consanguineous. Compared with Indian patients, Chinese patients had a relative later onset, a delayed diagnosis and treatment. More attempts are needed in Chinese PCG prevention and treatment.

• KEYWORDS: primary congenital glaucoma; clinical features; Chinese; Indian

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INTRODUCTION

Primary congenital glaucoma (PCG) is a severe form of childhood glaucoma and is characterized by increased intraocular pressure(IOP), corneal enlargement accompanied with corneal edema and ruptured Descemet's membrane, photophobia, blepharospasm, and excessive tearing [1]. The disease is disastrous not only because it will cause optic nerve damages that may lead to permanent blind in some sick children, but also because it is difficult to be diagnosed and treated promptly before the severe ocular damages occur. Fortunately, molecular genetic researches discovered at least 3 PCG causing gene loci [2-4] and more than 50 mutations in CYP1B1 gene were identified in PCG patients with different ethnic origins [5], which implies the coming possibility of early diagnosis by detecting the underlying defect genes. The genetic linkage analyses on PCG, from pedigree analyses to molecular investigations, confirmed the genetic heterogeneity of PCG[5]. Inbred populations showed a higher incidence of the disease as seen in 1:1 250 in Slovakia Romany population[6] , 1:2 500 in Saudi Arabians[7] , and 1:3 300 in the state of Andhra Pradesh in India [8]. Most PCG cases from inbred populations were familial and were mostly inherited in an autosomal recessive mode. Parent-to-child transmission (pseudodominance) examples also occured[9].

While in Western countries, where consanguineous marriages were relatively few, the PCG incidence ranged from 1:5 000 to 1:22 000, with most figures being closed to 1:10 000 and most cases were sporadic [10]. The clinical features of PCG also varied among different ethnics, as demonstrated in a comparison between Rom and non-Rom populations in Slovakia conducted by Gencik *et al* [11] in 1989.The two PCG populations showed significant differences in the sex ratio (male to female), age at diagnosis, the proportion of familial to sporadic , bilateral occurrence and the course and prognosis. Observations on other ethnic populations around the world also showed the variability of clinical features of PCG. These clinical discrepancies among different ethnics implied the phenotypic heterogeneity of PCG [2]. However, to the best of our knowledge, investigations on Mongoloid populations have only been carried out in Japanese and Indonesians so far[12] and there were no reports pertaining to the statistical comparison between the PCG patients of Mongoloid populations and other ethnics. The two ethnics investigated by Gencik *et al* were from the same geographic domain and there were no reports about clinical discrepancies between geographically different ethnics. That different genetic factors underlie different clinical features implies those population with different phenotypes may have different genetic etiology and further explorations in such populations are necessary. In this article, we attempt to summarize the clinical features of Chinese PCG patients, and by comparing them with Indian PCG patients [13], we try to find out the discrepancies in PCG clinical features between the two geographically and ethnically different populations.

MATERIALS AND METHODS

Materials Clinical records of all the primary congenital glaucoma patients that were admitted between January, 2002 and December, 2004 in the West China Hospital were reviewed by the authors. This hospital is one of the largest hospitals in China and is a teary medical center in West China, serving more than 2 million out-patient people and more than 80 thousand in-patient people every year, most of which are Mid-Western Chinese Han people. For the comparison, articles regarding the clinical features of PCG were retrieved in Pubmed and reviewed. We chose the data of Indian patients reported by Panicker *et al* [13], because (1) Indians are both geographically and ethnically different from Chinese and to our best of knowledge there were no reports about the clinical discrepancies of PCG between such two populations, and , (2)Among the articles we reviewed, only Panicker *et al* 's represented detailed clinical feature data and the Indian sample was large enough to do statistical analysis. Based on the Shaffer-Weiss classification of congenital glaucoma [14], only those patients being verified as the PCG patients were included. Patients with glaucoma associated with congenital anomalies or acquired glaucoma were excluded. The include/exclude criteria of the Indian patients were not reported by Panicker *et al* [13].

Table 1 Clinical data ranges observed in Chinese PCG patients

Clinical Parameters	Ranges Observed
Ages of onset	By birth - 9a
Ages of diagnosis	By birth - 18a
Time to diagnosis	0-144mo
IOP (mmHg)	9-76
Cup-to-disc Ratio of the optic nerve	0.3-1.0
Corneal diameter(mm)	11-17
Last recorded vision	20/20-NLP
Corneal changes	Corneal scar,Haab's striae,edema,buphthalmos
Treatments	Types of treatments

All the Chinese patients were examined by the experienced ophthalmologist Dr. Chen either under general anesthesia (for the uncooperative children) or not (for the cooperative ones). The examination included measuring horizontal corneal diameter, documenting the appearance of the cornea, measuring intraocular pressure (IOP) with a Perkins hand-held tonometer or a Goldmann applanation tonometer, and evaluating the optic disc by a direct ophthalmoscopy when the cornea was clear enough. For those patients who claimed to have a family history of "visual problems" or "eye diseases", we examined their relatives to confirm the family history of PCG. Details about the examination of the Indian patients were not reported by Panicker *et al* [13] .

The clinical features of Chinese PCG patients were collected from the clinical records and summarized as several clinical parameters, which could be quantified for the convenience of statistical comparison with Panicker *et al* 's study [13]. These parameters and their observed ranges were presented in Table 1. Not all the cases included in the present study had a complete record of all the parameters and those unavailable data were treated as missing values. The age of onset was speculated from the chief complains provided by the people who brought the children to the doctor or by the elder patients themselves. Indian PCG patients' data were cited directly from the published articles [13] and were not presented in this article.

Methods Panicker *et al* [13] developed a severity index to grade the various phenotypes of the Indian patients. Clinical

Table 2 Severity index constructed by panicker *et al* for grading various PCG phenotyps[13]

Clinical Parameters	Normal	Mild	Moderate	Severe/Very Severe
Corneal diameter (mm)	Up to 10.5	>10.5-12	>12-13	>13
IOP(mmHg)	Up to 16	>16-20	>20-30	>30
Last C/D ratio	0.-0.4	>0.4-0.6	>0.6-0.8	>0.8
Last recorded visual acuity	20/20	<20/20-20/60	<20/60-20/200	<20/200-20/400,<20/400- NLP*
Corneal clarity	No edema	Mild edema	Severe edema	Severe edema and Haab's striae

* Very severe phenotype

manifestations of the patients were graded from normal to severe. We cited the severity index directly, which was given in Table 2. Clinical manifestations of the Chinese patients included in our study were graded into 4 types of normal, mild, moderate, and severe/very severe in the same way as reported by Panicker *et al* [13]. A patient was graded "very severe" when the last recorded vision ranged between less than 20/400 and no perception of light (NPL)[13]. We further summarized the grading results of the Indian patients [13] and compared them with the grading results of the Chinese patients in the present study.

Statistical Analysis Kolmogorov-Smirnov D test was used to evaluate the distribution type of the continuous variables. To compare the clinical features of Chinese and Indian PCG patients, binomial test was used for the binomial variables, Mann-Whitney U test for non-normally distributed continuous variables, t test for the normally distributed continuous variables and χ^2 test for discrete variables, which were presented in table 3 in detail. SPSS 10.0 was employed to process statistical analysis.

The binomial variable was the gender. Non-normally distributed continuous variables analyzed were age of onset, age of diagnosis and time to diagnosis. Normally distributed continuous variables analyzed were IOP at diagnosis, corneal diameter and C/D ratio. Discrete variables analyzed were the affected eye (monocular or binocular), proportions of manifestation at birth, strategies of treatment and grades of severity. $P<0.01$ was considered to be statistically significant.

Data of age of onset, age of diagnosis and time to diagnosis (time span between onset and diagnosis) were rounded to the nearest number of months. Those ages of onset between birth and 15 days after birth were rounded to 0 month. Unavailable data were treated as missing values and were excluded by SPSS 10.0 program when processing statistical analysis, therefore the sample contents varied among the items compared.

RESULTS

During the 3 year study period, a total of 45 patients were initially diagnosed as "PCG" among the in-patient patients in West China Hospital. 44 of them were Chinese Han origin and 1 is Chinese Yi origin. 5 patients were excluded in our present study because 1 case had been applied dexamethasone solution for half a year before the eyeball became enlarged, 1 case had a blunt trauma on the affected eye, 3 cases, including the Yi patient, had other ocular or systemic defects and were confirmed to be Machsani syndrome, Aniridia and Sturge-Weber syndrome respectively. Therefore, 40 patients were included into this retrospective study. General clinical data of the 40 patients included in this study were given in Table 3. Quantitative clinical data of 43 Indian PCG patients were reported in detail by Panicker *et al* [13]. We just cited the data directly and did not present the Indian data in this article.

Epidemic Data of the 40 Chinese PCG Patients The study group included 40 Chinese PCG patients and 80 eyes. 27 patients were males (75%) and 13 patients were females (25%).The male to female ratio was round to 2∶1 and was significantly different from the expected 1∶1 ratio (P=0.04< 0.05, binomial test). The disease was bilateral in 30 patients (80%). In the 10 unilateral cases, 5 of the affected eyes were the right and 5 were the left. Family history was presented in 3 patients (7.5%), among which parent-to-child transmission was observed in one case. Consanguinity was found in only one patient(2.5%), whose grandmother and grandfather were first cousins. The main symptoms and signs noted on the initial examination were enlarged eyeball in 17 cases, decreased visual acuity in 14 cases, cloudy cornea or decreased cornea luster in 11 cases, photophobia and epiphoria in 9 cases, pain or vomiting in 4 cases and exotropia in 1 case. Most cases had combined symptoms and signs mentioned above. 2 cases were found blind in routine physical examinations and were diagnosed as PCG thereafter. 3 cases had ever been misdiagnosed as keratitis or keratomalacia associated with avitaminosis A in local hospitals .All the Chinese patients underwent surgical procedures and the surgeries varied. 22 eyes underwent the combined operation of trabeculectomy and trabeculotomy, 12 eyes underwent trabeculotomy and 12 eyes underwent trabeculectomy. 1

Table 3 Clinical Feature comparison between Chinese and Indian PCG patients

Clinical features	Chinese PCG patients	Indian PCG patients	Statistical method	*P*
Age of onset(mo)	*n**=36	*n**=42	*Mann-Whitney U test*	<0.01
M	6.0	0.0		
Range	0.0-108.0	0-12.0		
Interquartile Range	36.0	0.0		
Manifestation	*n**=40	*n**=43	*Person* χ^2 *test*	<0.01
at birth(*n*)	16	38		
after later(*n*)	24	5		
Age of diagnosis(mo)	*n**=40	*n**=43	*Mann-Whitney U test*	<0.01
M	36.0	0.0		
Range	0-216.0	0-360.0		
Interquartile Range	58.8	4.0		
Time to diagnosis(mo)	*n**=36	*n**=42	*Mann-Whitney U test.*	<0.01
M	12.0	0.0		
Range	0-144.0	0-120.0		
Interquartile Range	20.76	3.0		
IOP at diagnosis (mmHg)	*n**=61	*n**=75	*t* test equal variance not	<0.01
mean±SD	41.16±13.40	29.28±7.46	assumed	
M	41.00	28.00		
Corneal diameter (mm)	*n**=72	*n**=71	*t* test equal variance not	0.002
mean±SD	13.70±1.41	13.01±1.18	assumed	
M	14.00	13.00		
C/D ratio	*n**=30	*n**=45	*t* test equal variance not	<0.01
mean±SD	0.84±0.22	0.53±0.26	assumed	
M	0.90	0.50		
Affected eye	*n**=40	*n**=43	*Fisher* exact test	0.008
Monocular(cases)	10	1	.	
Binocular (cases)	30	42		
Treatment(eyes)	*n**=70	*n**=85	*Fisher* exact test	<0.01
Trabeculotomy	15	0		
Trabeculectomy	19	0		
Combined operation**	28	78		
Enucleation	3	1		
Medicine only	0	4		
No treatment	5	2		

*Sample contents vary among items because of missing data;**comibined operation of trabeculectomy and trabeculotomy

patient underwent bilateral enucleation due to the unbearable pain.

Clinical Feature Comparison between Chinese and Indian PCG Patients [13] Table 3 describes the differences of clinical features between Chinese and Indian PCG patients. Because age of onset, age of diagnosis and time to diagnosis were not normally distributed, we used the median and interquartile range to describe the data. For those normally distributed variables as IOP at diagnosis, corneal diameters and cup/disc ratio, we used the mean and standard deviation to describe the data. The median age of onset was 6.0 month for Chinese PCG patients and 40% of the cases manifested at birth. 88.4% of the Indian patients manifested at birth, therefore the median age of onset for Indian patients was 0.0 month. The median age of diagnosis was 36.0 month for Chinese patients and 0.0 month for the Indian patients. The median time to diagnosis for the Chinese patients was 12.0 month while most of the Indian PCG patients were diagnosed shortly after the disease onset. The mean IOP at diagnosis for the Chinese patients was 41.16mmHg and was 29.28 mmHg for the Indian patients. The mean cup/disc ratio was 0.84 for Chinese patients and 0.53 for the Indian

patients. The ratio of the cases with unilateral affected eye to the cases with bilateral affected eyes was 10/30 for Chinese patients and 1/42 for Indian patients. Among the 70 Chinese PCG affected eyes studied, 15 eyes received the trabeculotomy, 19 eyes received trabeculectomy, 28 eyes received the combined operation of trabeculotomy and trabeculectomy , 3 eyes were enucleated due to unbearable pain and 5 eyes received no treatment for they were of no light perception. Against the 85 Indian PCG eyes, 78 were exerted the combined operation of trabeculotomy and trabeculectomy, 1 eye was enucleated, 4 eyes received medical treatment only and 2 eyes received no treatment.

For all the variables analyzed herein, there was statistically significant difference between the Chinese and Indian PCG patients (P <0.01). Chinese PCG patients had relative later onset than the Indian patients and the diagnosis age was later as well. The proportion of cases with manifestation at birth was significant lower than that of the Indian patients. The time span for the patient to be diagnosed was larger in Chinese than that in Indians. When diagnosed, the Chinese patients had higher IOPs, larger corneal diameters and greater cup/disc ratios than the Indian patients did. There were a higher proportion of unilateral affected cases in Chinese PCG patients than that in Indian patients. The combined operation of trabeculotomy and trabeculectomy was more frequently exerted on Indian patients than on Chinese patients.

Severity Grading Comparison The Chinese patient's manifestations were evaluated and graded by using the severity index constructed by Panicker *et al* [13]. Both eyes of every patient included in this study were evaluated since asymmetric manifestations may present in PCG patients[9]. A total of 80 Chinese PCG eyes were graded from normal to very severe and the number of eyes fall into each grade was counted. There were 86 Indian eyes graded by Panicker *et al* [13]. We counted the number of Indian eyes of each grade. The grading results of the Chinese and Indian patients were compared by using the Chi-square test, which is shown in Table 4. There was statistical difference between Chinese and Indian patients in the proportion of severity grades. Most Chinese patients had a severe manifestation of the disease whereas the very severe type accounts for the major part of the Indian PCG patients.

Table 4 Severity grading comparison between Chinese and Indian PCG patients

Severity	Chinese eyes (n)	Indian eyes(n)	P
Normal	7	1	<0.01
Mild	5	6	
Moderate	11	12	
Severe	43	25	
Very severe	14	42	
Total	80	86	

DISCUSSION

PCG accounts for approximately 55% of primary pediatric glaucoma and is an important cause of childhood blindness [15]. Investigations on this disastrous disease in Mongoloid populations are fewer than those in Caucasian populations. We reviewed the clinical records of PCG patients that were admitted between January, 2002 and December, 2004 in West China Hospital, summarized the clinical features with several quantifiable parameters and compared them with those of Indian PCG patients' [13]. We also evaluated the severity of Chinese PCG manifestations by using the severity index constructed by Panicker *et al* [13] and compared severity grading results of the two populations. We confirmed the diagnosis of PCG mainly based on the ocular manifestations and excluded those cases with a diagnostic uncertainty, with any evidence for a secondary origin of glaucoma or with glaucoma as a component of a specific syndrome. We didn't focus much on the age of onset when considering about the include and exclude criteria because (a) onset age was provided by the patients or their relatives with much less medical knowledge in general and would likely be distorted by the potential recall bias; (b) some authors[9]used "primary congenital glaucoma " to refer to all the 3 groups of childhood glaucoma that were recognized at birth , between the age of 1 month and 2 years, and after the age of 2 years. Thus we included all the 3 groups in our present study without a very strict limitation on the patients' onset age.

Studies showed that PCG was an entity with both genetic and phenotypic heterogeneity [2,16]. Most cases of primary congenital glaucoma were sporadic, but 10% to 40% were familial, frequently associated with consanguinity[2] . In most familial cases, transmission was autosomal recessive and pseudo-dominant transmission might exist, with expression and penetrance varying from 40% to 100% [2]. Most of the Chinese PCG cases studied were sporadic and non-consanguinous. Successive transmission was also observed in Chinese PCG population. 48.5% of the Indian families were of a non-consanguineous group and sporadic cases accounted for 80% of the group [13]. The difference in

hereditary background may account for the discrepancies in clinical features compared between these two populations.

In our patients studied, the male to female ratio was 2:1. The sex ratio of the Indian patients was not reported[13]. Sex ratios (male to female) of the non-consanguineous PCG populations were different significantly from the expected 1:1 [11] as demonstrated in Japanese (2:3), the United States(3:2) , Europe (3:2)[9] and non-Gypsy populations of Slovakia (1.55:1)[11]. The sex ratios in these PCG populations were not consistent with an autosomal recessive segregating mode of the disease. In Gypsy populations of Slovakia with high consanguinity rate, the sex ratio (male to female) was 1.15:1[11], which can be explained by the autosomal inheritance. Unequal gender distribution and successive generations transmission may be explained by the genetic heterogeneity that was confirmed by linkage analysis and molecular investigations in recent years [2,16].

Main symptoms and signs observed in our patients were diverse, spanning almost the whole manifestation spectrum of the primary congenital glaucoma [9]. The most frequent signs noted on the initial examination were enlarged eyeball and decreased VA. According to a previous study [17], progressive ocular enlargement ceased by the age of 3 to 4a, stretching the cornea, lengthening the ocular axis and developing typical buphthalmos in severe cases. It is worth noting the fact that cases with decreased VA as the initial examination signs accounted for 35.0% of the population (14 cases out of 40 cases), which was higher than that of the well known signs of photophobia and epiphoria (22.5%, 9 cases out of 40 cases). Exotropia was the initial sign that made the parents brought the children to the doctor in 1 case and, even in 2 cases, children were not diagnosed as PCG until they were found blind in routine physical examinations. The median diagnosis age of the cases with decreased VA as the initial examination signs was 66 months, which is significantly greater than that of the whole Chinese PCG population (P = 0.003, Mann-Whitney U test) . This may make the clinicians think about the possibility of PCG when examinating an elder child complaining about the decreasing of visual acuity or when confronted with strabismus children. Because children are often unable to describe their feelings clearly, nausea was observed in 10% of the patients (4 cases out of 40 cases). The cluster changing of the cornea may lead to the misdiagnosis of keratitis or keratomalacia associtated with avitaminosis A [17], which were seen in 3 cases in our studied group. All these make the careful examinations rather important when diagnosing the disease. Since almost all the Indian patients were bilaterally affected while 25% of the Chinese patients were unilateral cases, asymmetric expression should be suspected in clinically apparent unilateral cases[9].

We found that Chinese PCG patients had a relative later onset than the Indian patients. However, all the Indian patients' onset age was under 12 months [13]. Because we could not access to details about the selection of the Indian PCG patients in the published article [13], we had to hold the guess that Panicker *et al* might have only chose those patients with onset age under 12 months. But we don't think this tentative selection bias would likely to affect our analysis much for we have conducted a Mann-Whitney U test which is based on median and median would be much less affected by the actual value of the data than mean[18]. To confirm the relative later onset of Chinese PCG patients further, we compared the proportions of the cases manifesting at birth between the two populations. The Indian PCG patients had a much higher proportion of manifesting at birth than the Chinese, which implied a relative later onset of the Chinese PCG indirectly. Similar scenarios occurred in the comparison between Gypsy and non-Gypsy populations[11], whose proportions of manifesting at birth were approximately 80% and 35% respectively. Both the Gypsy and Indian populations studied had high consanguinity rates of 40.8% [11] and 48.5% [13] respectively while the Chinese and the non-Gypsy populations' consanguinity rates were 7.5% and 5%[11] respectively. This implies an earlier disease onset in consanguinity PCG populations. Earlier onset cases had been confirmed to be associated with poor outcome of surgery by both surveys [1,19] and statistical analysis [20], which suggests that PCG cases with earlier onset may have more severe developmental defects in the aqueous humor drainage system. Age of onset may be one of the important clinical features to predict the prognosis of the disease.

PCG has an early onset. Being promptly diagnosed and treated, substantial cases can have better prognosis [21]. The diagnosis age of Chinese PCG patients was later than that of the Indians'. This may be attributed partly to the relative later onset of the disease. The comparison of the time to diagnosis between Chinese and Indian patients confirmed a delayed diagnosis existing in Chinese PCG population. The main reason may be the poor recognition of the cases by their

parents. On the other hand, since PCG is a rare disease and some of the clinical manifestations mimic corneal diseases, misdiagnosis might be made without careful inspections and delayed the right treatment strategies.

The IOP at diagnosis, corneal diameter and the C/D ratio are main parameters for describing the severity of PCG. In general, for normal infant eyes, the IOP ranges from 11 to 14mmHg and the horizontal corneal diameter is no greater than 12mm[17]. AC/D ratio greater than 0.3 was found in 68% PCG infants [22] and in less than 2.6% normal newborns [23]. Though the severity grading showed more Indian cases were categorized into the "very severe " grade, we would rather believe that this group of Chinese patients was more severe affected than the Indian patients for the comparison of the 3 parameters implied a more severe clinical situation in Chinese PCG patients than in Indian patients. The parameter for differentiating the "severe" and "very severe" categories herein was the last recorded VA and it is well known that usually the VA won't be damaged until glaucoma progresses to its final stage. We conducted a retrospective study and no records of follow-up were available, thus the "last recorded visual acuity" and the "last C/D ratio " were actually recorded when the patients were released from hospital. The Indian patients had been followed up for several years by the time they were studied and it is reasonable to believe that the last VA and the last C/D ratio were recorded long after the initial diagnosis. Since the "last recorded visual acuity" of the two populations would most probably be recorded in quite different stages, it should not be taken much into consideration when comparing the severity. The large proportion of "severe" cases in Chinese patients may progress into the "very severe" stage after several years of follow-up, given the VA be the parameter of grading the very severe cases. The two populations would be more comparable if both of them had been graded by using the clinical classification constructed by AI-Hazmi *et al* that involved only the IOP, corneal diameter and corneal clarity [24].

The management of PCG is primarily surgical. Medication alone is rarely effective but plays a secondary role as an adjunctive treatment [1]. Strategies of surgeries include goniotomy, trabeculectomy, trabeculotomy and the combined trabeculectomy and trabeculotomy [25]. Goniotomy requires the good visibility of the chamber angle structures [20]. This may be the reason why neither the Chinese PCG patients nor the Indian patients underwent this procedure. Other studies[24,26], including an Indian one[26] ,showed the combined operation gave the best results for PCG patients. This may be an explanation of the preference to the combined operation in Indian patients studied herein. However, the association of the effectiveness with the type of the surgeries applied remains unclear for some other studies showed success rate did not differ significantly among the 3 procedures of trabeculectomy, trabeculotomy and the combined trabeculectomy and trabeculotomy [20]. Investigators tended to agree that individual course and severity of the disease governed the prognosis of the surgeries [20,24]. Major risks for surgery failure may include the patients onset age under 3 months, the ocular axial length of 24mm or more [20,27-29], parental consanguinity[29] , initial high IOP and high C/D ratio[25]. We chose surgeries depending on the integrity of the limbus, exerting trabeculot- omy or the combined operation on those with a relative intact limbus and trabeculectomy on those with abnormally stretched limbus. The higher proportions of no treatment eyes and enucleated eyes in Chinese patients than in Indian patients implied the higher proportion of delayed treatments in Chinese PCG patients.

Studies comparing the difference of clinical features between two populations have to be evaluated with caution, especially when there are some missing values as it does here. On the other hand, PCG represents an extraordinary rare disease and it is difficult to conduct a prospective study between two nations. We can treat this comparison as a gross one and further dedicated designed follow up is to be done in Chinese PCG population.

In conclusion, most Chinese PCG patients were sporadic and had few associations with consanguinity. Compared with the Indian patients, Chinese patients have relative later onset and delayed diagnosis and treatments. More attempts are needed in Chinese PCG prevention and treatment.

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