Original Article

Outcomes of trabeculectomy in microspherophakia

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Purpose: To report the outcomes of trabeculectomy in eyes with glaucoma in microspherophakia. **Materials and Methods:** In a retrospective non-comparative case series, we analyzed 29 eyes of 18 patients with glaucoma in microspherophakia, who underwent primary trabeculectomy between 1998 and 2012. Success was defined as complete if the intraocular pressure (IOP) was ≤21 and >5 mm Hg without any antiglaucoma medication and qualified if IOP ≤21 and >5 mm Hg with or without antiglaucoma medications. Eyes not falling into qualified success criteria were labeled as failure. **Results:** The median age at the time of trabeculectomy was 23 years (inter quartile range: 12, 28). The mean IOP reduced from 31.1 ± 8.6 mm Hg to 14.6 ± 4.4 mm Hg after trabeculectomy over a median follow up of 77 months ($P < 0.001$). The probability of complete success was 96% (95% CI: 77-99%) at one year, 88% (95% CI: 67-96%) at 2 years, which was maintained till 7 years and decreased to 79% (95% CI: 50-92%) at 8 years. The probability of qualified success was 100% till 7 years and decreased to 90% (95% CI: 47-98%) at 8 years. The median number of postoperative medications reduced from 2 to 0 postoperatively **(**$P < 0.001$**).** Five eyes (21%) developed post-operative shallow anterior chamber (AC) requiring anterior chamber reformation, with 2 of these eyes needing lensectomy for resolution of this complication. **Conclusion:** Primary trabeculectomy had good success rate in glaucoma associated with microspherophakia. Post-operative shallow AC was a frequent complication needing additional intervention.

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Microspherophakia is a rare developmental anomaly of the crystalline lens in which the lens assumes a spherical shape with increased antero-posterior and reduced equatorial diameter.[1] The condition is bilateral and can occur either as an isolated anomaly or with associated systemic disorders such as Weill-Marchesani syndrome (WMS) or Marfan's syndrome.[1-3] The hallmark of this condition is visibility of the equator of the lens on full mydriasis [Fig. 1a]. The condition is associated with subluxated or dislocated lens, progressive myopia, defective accommodation, glaucoma and retinal detachment.[4] Glaucoma is the most common sight threatening complication,[4-10] affecting more than 50% of the eyes associated with this condition.[11] The spherical and anteriorly positioned lens results in shallow anterior chamber, predisposing these eyes to angle closure glaucoma. Acute angle-closure can result from pupillary block caused by anteriorly subluxated or dislocated spherophakic lens.[4,5] Recurrent attacks of pupillary block by spherophakic lens can lead to chronic synechial angle closure and glaucoma.[6,7] Though less frequent, secondary open angle glaucoma due to developmental anomaly of the drainage angle has been described in eyes with microspherophakia.[2,6]

Glaucoma associated with microspherophakia is difficult to manage with close to 60% of the eyes failing medical and laser treatment. These eyes often require surgical intervention for IOP control.[11] Various surgical modalities have been described to manage glaucoma in microspherophakia, which include lensectomy, goniosynechiolysis, trabeculectomy, drainage implants or a combination of these procedures. The available evidence so far is from isolated case descriptions with limited follow up.[2,12-15]

The outcomes of trabeculectomy in eyes with glaucoma associated with microspherophakia has not been reported so far and hence this communication.

Materials and Methods

We retrospectively reviewed all charts of subjects with microspherophakia who underwent trabeculectomy at our institute between February 1998 and May 2012. Our institutional review board approved the study. Trabeculectomy was performed in 31 eyes of 20 subjects during the study period. Two eyes of two patients with follow up of less than six weeks were excluded, hence twenty-two eyes of 11 subjects (both eyes) and seven eyes of seven subjects were included in the analysis. Microspherophakia was diagnosed clinically, based on the spherical shape of the lens on slit lamp biomicroscopy and the visibility of entire lens equator on pharmacological mydriasis [Fig 1a].

The preoperative data noted were age, gender, visual acuity, refractive error, intraocular pressure (IOP), gonioscopic findings, condition of lens, optic disc changes, number of anti-glaucoma medications and presence or absence of laser peripheral iridotomy (PI). The operative details of trabeculectomy were noted. Post-operative data noted were visual acuity, IOP, requirement of antiglaucoma medications, complications and interventions if any, at all subsequent visits.

Glaucoma in microspherophakia was diagnosed based on one or more of the following criteria; (a) untreated IOP >22 mm Hg, recorded twice on different occasions (b) history of using anti-glaucoma medications or (c) glaucomatous optic nerve

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damage in the form of rim thinning, notch, asymmetric disc cupping (difference in cup to disc ratio >0.2 between 2 eyes) with corresponding visual field changes.

Our algorithm for glaucoma management in eyes with microspherophakia has already been described.[11] In brief, trabeculectomy was indicated in eyes with IOP >21 mm Hg despite maximum medical therapy, in the absence of gross subluxation or dislocation of the lens. To improve the outcomes of trabeculectomy in this complex condition, we have suggested a few modifications in the surgical technique. Anti-metabolites were not used to augment trabeculectomy either intra-operatively or post-operatively.

Trabeculectomy was performed either by a fornix based or limbus based conjunctival approach. All procedures were performed by one of the four senior glaucoma surgeons (SS, HLR, AKM, GCS). A 4 × 4 mm triangular scleral flap was dissected up to clear cornea, a 2 × 2 mm deep scleral block was excised and peripheral iridectomy was performed. The scleral flap was closed with one to three (depending on the amount of filtration) 10-0 nylon sutures and conjunctiva was closed with 8-0 vicryl suture in a continuous fashion or winged sutures based on conjunctival incision. Anterior chamber reformation was performed with air, whenever required as per surgeon's discretion to avoid postoperative shallow anterior chamber (AC) [Fig 1b]. This was performed using a 26/29 gauge needle through a long paracentesis tract under local or topical anesthesia and the needle gently withdrawn and sealed with a cotton bud to prevent air escape. Topical cycloplegics were started intraoperatively (to deepen the anterior chamber by cycloplegia and posterior pull of the iris lens diaphragm) and the same was continued in the postoperative period along with topical steroids and antibiotics.

Statistical analysis

Descriptive statistics included mean and standard deviation (SD) for normally distributed variables and median and inter-quartile range (IQR) for non-normally distributed variables. Generalized estimating equations were used during comparisons between preoperative and postoperative parameters, as both eyes of 11 subjects were included in the analysis.

Kaplan-Meier survival curves were used to assess cumulative probability of success. Complete success of trabeculectomy was defined as IOP ≤21 and >5 mmHg without any antiglaucoma medication. Qualified success was defined as IOP ≤21 and >5 mm Hg with or without antiglaucoma medications. Eyes not falling into qualified success criteria were labeled as failure. Associations between failure and preoperative factors such as age at surgery, gender, refractive error, preoperative IOP and number of preoperative medications were assessed using Cox proportional hazard regression model. A $P \leq 0.05$ was considered statistically significant. Statistical analysis was performed using commercial software (Stata ver. 11.0; StataCorp, College Station, Tx).

Results

The demographic and clinical features of 29 eyes of 18 subjects with microspherophakia that underwent trabeculectomy are shown in Table 1.

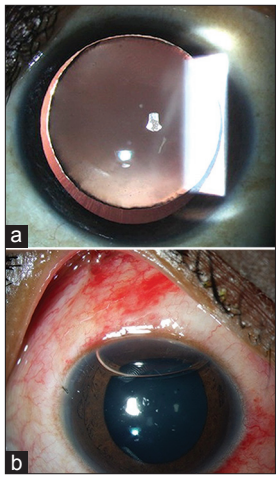

Figure 1: (a) Clinical picture of microspherophakia with the lens equator visible in a dilated pupil. (b): A post-trabeculectomy day three picture of a microspherophakic eye with primary air injection showing a diffuse bleb, well-formed anterior chamber and a small residual air bubble

Table 1: Demographic and clinical details of patients (29 eyes of 18 subjects)

	Median	Interquartile range
Age (years)	23	12, 28
Gender (male: female)	10:8	
Follow-up (months)	77	35, 117
Refractive error (diopters)	-12.75	-7.0, -16.50
Cup-disc ratio	0.9	0.8, 0.9
	Mean±SD	Range
Preoperative IOP (mmHg)	31.1±8.6	18 to 46
Postoperative IOP (mmHg)	14.6±4.4	6 to 31

SD: Standard deviation, IOP: Intra ocular pressure

Seven of the 18 subjects had systemic features suggestive of Weill-Marchesani syndrome. Pre-operative gonioscopy was available in 26/29 eyes, of which 21 eyes had closed angles and 5 eyes had open angles.

Median preoperative log MAR visual acuity was 0.3 (IQR: 0.2 and 0.6) and median postoperative log MAR visual acuity was 0.3 (IQR: 0.2 and 0.9). There was no statistically significant difference ($P = 0.90$, Wilcoxon signed-rank test) between the pre-operative and post-operative visual acuity. At the final follow up visit after trabeculectomy, visual acuity remained stable or improved in 26 eyes and deteriorated in three eyes. Two of the three eyes had deterioration in vision following trauma (scleral tear in one and non resolving vitreous hemorrhage in the other) and one eye had decrease in vision due to advanced glaucoma despite IOP ranging between 14 and 17mm Hg throughout the follow up period.

The probabilities of complete and qualified success after trabeculectomy are shown in Figs. 2 and 3. The probability of complete success was 96% (95% confidence interval (CI): 77-99%) at one year, was 88% (95% CI: 67-96%) at 2 years, which was maintained till 7 years, it reduced to 79% (95%CI: 50-92%) at 8 years and was maintained until 14 years. The probability of qualified success was 100% till 7 years and

decreased to 90% (95% CI: 47-98%) at 8 years and remained so till the last follow up. In our study, both eyes of 11 subjects were included and it is possible that this may have influenced the results. However, we found that the results were similar when we included only the left eye of subjects who contributed both eyes for the analysis, the probability of complete success was 100% at one year, was 93% (95% CI: 59-98%) at 2 years, it reduced to 77% (95%CI: 31-94%) at 8 years and was maintained until 14 years.

The mean (± SD) preoperative IOP in these eyes was 31.1 ± 8.6mm Hg and the mean postoperative IOP was 14.6 ± 4.4 mmHg ($P < 0.001$, Wilcoxon signed-rank test). The median percentage reduction of IOP following trabeculectomy was 54% (IQR: 40-64%). Fig. 4 shows the scatter plot comparing the pre and post-operative IOP. The median number of antiglaucoma medications reduced significantly from 2 preoperatively to 0 postoperatively ($P < 0.001$, Wilcoxon signed-rank test).

Primary AC reformation with air at the conclusion of trabeculectomy was performed in 21/29 eyes. Significant complication noted during the first post-operative week was shallow AC, which was seen in five eyes (3 of these eyes had primary AC reformation with air). The shallow AC was associated with irido-corneal touch in three eyes and irido-lenticular touch in two eyes, which persisted even after conservative management. None of these eyes had wound leak and IOP ranged from 5-12mm Hg during this time. Four of these five eyes had gonioscopically-closed angles preoperatively. All 5 eyes underwent AC reformation with air; two of these eyes continued to have flat anterior chamber despite AC reformation and required lensectomy with anterior vitrectomy, one eye in less than one week and other eye 6 weeks after trabeculectomy.

Thirteen of the 29 eyes underwent lensectomy during the follow up period. Two of these within 6 weeks, one eye developed anterior dislocation of the lens after 18 months, 7 eyes had progressive subluxation of the lens and required lensectomy with a median time to lensectomy of 49 months (IQR: 7.4, 130 months), 3 eyes developed cataract and required lens extraction after an average of 39 months. The IOP was controlled without any antiglaucoma medications in these 13 eyes even after lensectomy until the last follow-up.

Four eyes in our series developed IOP ≥22 mm Hg during the follow-up period. IOP was controlled with topical medications in 3 of these eyes and one eye failed to respond to medical treatment and was termed failure. This eye developed raised IOP 8 years after trabeculectomy following blunt injury to the eye, presented with vision of counting fingers close to face, subluxated lens, vitreous hemorrhage and IOP of 40 mm Hg, was started on antiglaucoma medications and topical steroids, and was lost to follow up after this visit.

None of the pre-operative factors, namely age at surgery, gender, refractive error, preoperative IOP, number of preoperative medications, preoperative angle status and need for AC reformation were significantly associated with failure of trabeculectomy by Cox proportional hazard regression model ($P > 0.10$ for all associations).

Discussion

In our study, the success rate of trabeculectomy in glaucoma associated with microspherophakia was good and resulted in an effective control of IOP over a long term. The complete success probability was 96% at the end of one year, which reduced to 88% at 2 years, which was maintained till 7 years and qualified success was 100% at one year, which was dropped to 90% at 8 years. Though the success rates cannot be compared directly due to lack of reports on trabeculectomy in eyes with microspherophakia, the success of trabeculectomy seems to be better than that of other secondary glaucomas (54%).[16]

As shown by Senthil *et al*., in their previous publication,[11] glaucoma was associated with microspherophakia in 51% of the eyes at presentation and affected younger individuals (median age of 16 (IQR: 9, 25.5 years). Of these eyes with glaucoma, 20% of the eyes at presentation and 30% of the eyes at last follow up were blind due to glaucomatous optic neuropathy. This emphasizes the need to detect and treat glaucoma early and appropriately, to prevent visual impairment and blindness associated with this condition.

Various surgical modalities have been recommended based on isolated case reports for the management of this condition either by lensectomy,[2,17,18] trabeculectomy,[12,15] with or without goniosynechiolysis or shunt surgeries.[15]

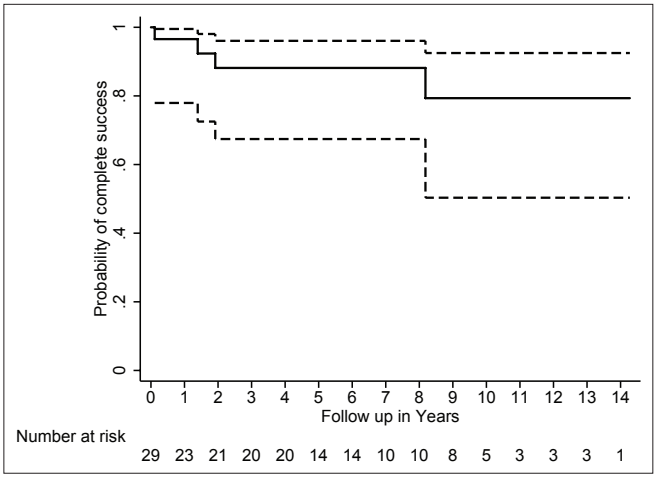

Figure 2: Kaplan-Meier survival showing complete success of trabeculectomy in microspherophakia with 95% confidence intervals (dotted lines)

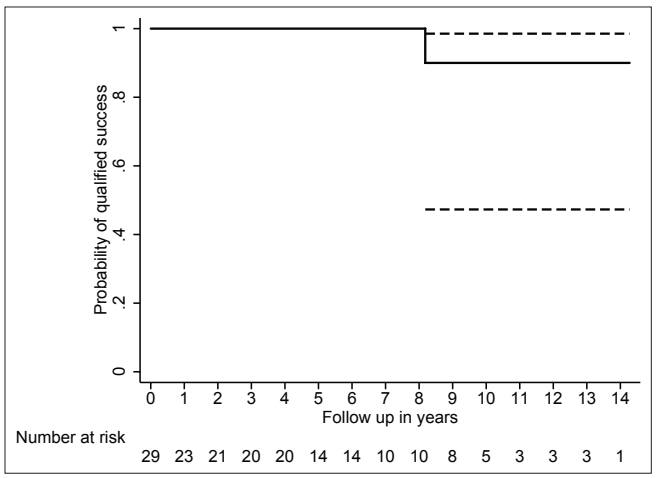

Figure 3: Kaplan-Meier survival showing qualified success of trabeculectomy in microspherophakia with 95% confidence intervals (dotted lines)

Simple lensectomy or lens extraction was shown to be effective in controlling IOP in a few studies, but with shorter follow up.[2,17,18] Studies with longer follow up have reported that lensectomy alone was not sufficient in controlling the IOP in eyes with glaucoma in microspherophakia and glaucoma filtering surgery was required later during the follow up to control the IOP.[12] Combined lens aspiration and goniosynechialysis followed by peripheral iridoplasty was shown to successfully control the IOP in two eyes with synechial angle closure and glaucoma with a follow up of 1-2 years.[13]

There are limited and conflicting reports on the surgical outcome of trabeculectomy alone in the treatment of glaucoma associated with Microspherophakia. Asaoka *et al*., reported good IOP control with trabeculectomy in a spherophakic eye with secondary angle closure glaucoma, with 4 months follow-up.[19] Paul Harasymowycz *et al*., have reported trabeculectomy in one eye, which required lensectomy, vitrectomy with scleral fixated intraocular lens and Molteno tube shunt implant for the management of persistent flat anterior chamber following trabeculectomy.[15] They resorted to primary lensectomy with intraocular lens and tube implant in all the other three eyes in their series with good IOP control and visual rehabilitation.

Trabeculectomy in eyes with Microspherophakia is not only a surgical challenge but post-operative management is equally critical. Careful pre-operative assessment of the anterior chamber depth, angle status, the lens position and stability would help plan surgery appropriately and prevent complications. Meticulous surgical technique, by avoiding sudden decompression of the anterior chamber, careful deep block excision and iridectomy, tight scleral flap closure and reformation of the anterior chamber with air at the conclusion of the surgery would help prevent post-operative shallow AC and other complications. Also taking precautions to avoid wound leak by ensuring a tight conjunctival closure and if needed suturing the paracentesis wound may help to prevent postoperative shallow AC. Care should be taken to avoid intra-operative complications like vitreous loss (lack of zonular and peripheral lens support predispose these eyes to vitreous disturbance during or after iridectomy) that are specific to eyes with microspherophakia. Figs. 5a and b are slitlamp photographs showing a diffuse bleb and a well-formed anterior chamber in an eye with microspherophakia one year following trabeculectomy.

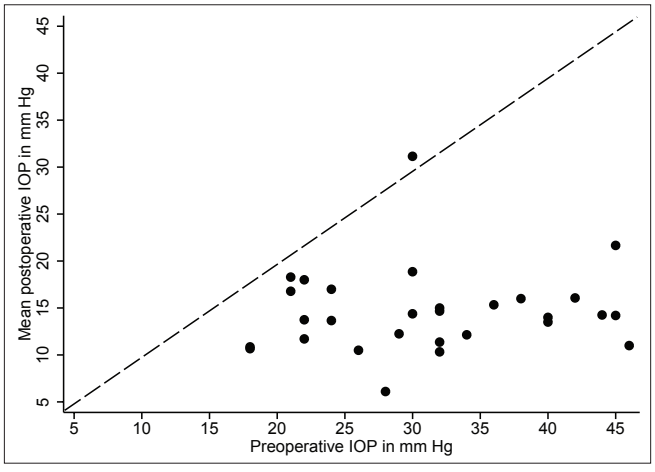

Figure 4: Scatter plot showing the preoperative and the postoperative intraocular pressures in the study eyes

Despite intra-operative precautions taken during trabeculectomy, 5 out of 29 eyes required AC reformation to manage post-operative shallow AC. Although 3 eyes resolved with AC reformation alone, the other 2 eyes required lensectomy with vitrectomy for persistent flat AC. Among the eyes with post-operative shallow AC, 4 eyes had preoperative closed angles and one eye had open angle and interestingly both the eyes that required lensectomy for persistent flat chamber had subtle subluxation of the lens preoperatively. One needs to be cautious in performing trabeculectomy for eyes with glaucoma in the presence of a subluxated lens. Combined lensectomy and trabeculectomy in these eyes could have been a better option. One of the eyes that had intervention for postoperative shallow AC developed raised IOP one month postoperatively and was controlled with one antiglaucoma medication.

It is important to note that 45% of the eyes after trabeculectomy required lensectomy during the follow up period. This observation highlights the fact that after trabeculectomy, apart from the IOP control these eyes need close follow up to assess the lens stability as these eyes are prone for progressive lens subluxation due to weak zonules.

None of the preoperative factors were significantly associated with failure of trabeculectomy in our series. This may be due to the small sample size.

This, to our knowledge is the first report on the outcomes of trabeculectomy in this rare condition with a long follow up and we believe this would add new knowledge to the existing literature. The limitation of this study is its retrospective design and a small sample size with variable follow up. However, for a rare condition like microspherophakia, retrospective study could be the only appropriate design to answer some important clinical questions.

Conclusion

In our series, the outcome of trabeculectomy for the management of secondary glaucoma in microspherophakia was good. Postoperative shallow AC was a significant complication and required surgical intervention for its

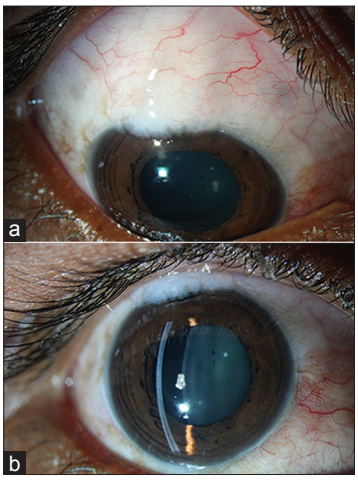

Figure 5: One-year postoperative clinical picture of a microspherophakic eye following trabeculectomy showing a diffuse bleb (a) and a well-maintained anterior chamber (b)

management. Nearly half the eyes required lensectomy during the follow up.

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