DOI: 10.7860/JCDR/2026/80548.23099

Review Article

Ophthalmology Section

Surgical Management of Microspherophakia in Young Patients and Associated Visual Outcomes: A Narrative Review

SUYASH SINGH[1], SACHIN DAIGAVANE[2]

ABSTRACT

Microspherophakia (MSP) is a rare crystalline lens development abnormality characterised by increased anteroposterior thickness and decreased equatorial diameter. It is connected with a variety of ocular and systemic diseases, the most serious of which is secondary glaucoma. Early identification, proper lens and glaucoma care, and adequate visual rehabilitation can all help to avoid glaucoma-related blindness. Because MSP mostly affects young people, a multidisciplinary approach with ongoing follow-up is strongly advised. The rarity of this illness has hampered the availability of large-scale prospective investigations, with current information primarily obtained from case reports and retrospective analysis. This makes the development of standardised treatment guidelines problematic. The purpose of this review is to examine the surgical aspects of managing MSP in young patients and to evaluate the associated visual outcomes. To alleviate zonular weakness, surgical treatment options include techniques such as lens extraction with or without Intraocular Lens (IOL) implantation, as well as more sophisticated approaches such as scleral-fixated IOLs, iris-claw lenses, and capsular tension devices. Recent research suggests that prompt surgical intervention can greatly improve visual acuity and overall quality of life in afflicted patients. Nonetheless, long-term treatment must include possible problems such as glaucoma, zonular instability, as well as IOL displacement. This review emphasises the significance of individualised surgical methods, coordinated multidisciplinary therapy, and long-term follow-up in achieving excellent visual results in young people with MSP. It also emphasises the importance of bigger prospective studies and the creation of standardised surgical guidelines for improving evidence-based clinical practice.

Keywords: Intraocular lens, Ocular devices, Quality of life, Treatment modalities, Visual acuity, Young adults

INTRODUCTION

The MSP is a rare eye condition present from birth that significantly alters the lens of the eye. In MSP, the lens becomes unusually small and spherical, meaning its width is reduced while its thickness increases [1]. The prevalence rate of MSP in children can reach 1.5%, and the incidence is more than one per 100,000 people [2]. MSP has a spherical lens and loose zonules, related subluxation/dislocation of the crystalline lens, which is a hallmark feature. The lens has a spherical form due to its reduced equatorial diameter and increased anteroposterior diameter, which makes the lens equator visible when complete mydriasis is attained [3]. The defining trifecta of MSP in a young patient is: a) lenticular myopia; b) shallow anterior chamber; and c) Angle Closure Glaucoma (ACG) [4]. Most individuals have a solitary MSP, which is often bilateral. The two most often documented systemic conditions of MSP are homocystinuria and Weill-Marchesani Syndrome (WMS) [5,6].

Nearly 50% of the eyes affected by this illness have glaucoma, which is the most prevalent sight-threatening consequence. These eyes are more susceptible to ACG because of the shallow anterior chamber caused by the spherical and anteriorly positioned lens [7]. A pupillary block brought on by an anteriorly subluxated or displaced spherophakic lens may result in acute angle closure [8]. Glaucoma and chronic synechial angle closure can result from recurrent pupillary block episodes caused by spherophakic lenses [9]. In eyes with MSP, secondary open-angle glaucoma caused by a drainage angle developmental abnormality has been reported; it is less common [10].

Therefore, this narrative review aimed to understand various surgical modalities among young adults and the associated visual outcomes for MSP.

Pathophysiology of Microspherophakia (MSP)

The lens of a foetus is spherical by nature. An adequately functioning ciliary body and zonules are thought to never exert tension on the lens in spherophakia. The lengthy zonules are frequently detached from the ciliary processes, particularly on the posterior lens surface. These lenses are vulnerable to displacement inside the anterior chamber/ vitreous cavity due to the zonules' weakness, which can happen spontaneously or with minor trauma [11].

The spherical lens may migrate forward due to aberrant zonules, resulting in acute pupillary block glaucoma. ACG can also be caused by persistent, undiagnosed intermittent pupillary blockages that cause irreversible damage to the trabecular and peripheral anterior synechiae [4,12]. A safer first step to alleviate pupillary obstruction is via Laser Peripheral Iridotomy (LPI). In certain circumstances, pharmacologic mydriasis, intravenous mannitol, and placing the patient in a supine posture may aid in posterior lens relocation when there is high intraocular pressure and a displaced lens in the anterior chamber. In order to alleviate the pupillary block and avoid corneal decompensation, the individuals might need an emergency lensectomy.

There are several reports on secondary open angle caused due to glaucoma and developmental anomalies of the angle [4,13]. Even in eyes with open angles, LPI is thought to be beneficial because it prevents occasional pupillary obstruction. Since there are other glaucoma mechanisms besides pupillary block, laser and medication treatment are frequently insufficient to manage the intraocular pressure [6].

Building on the review article's aim, which is to evaluate the surgical procedures and visual outcomes among young patients suffering from MSP, it is vital to explain the clinical significance of lensectomy in such cases.

Lensectomy

Lensectomy is an important surgical step in addressing diseases in which the crystalline lens causes substantial vision impairment or subsequent ocular problems. In young individuals with illnesses such as MSP, ectopia lentis, or severe lens-induced glaucoma, the spherical or subluxated lens might disrupt anterior chamber structure, cause angle crowding, and raise intraocular pressure [14,15]. Early lens removal not only alleviates these physical obstacles, but it also aids in visual rehabilitation, especially when combined with IOL implantation or other stabilising devices [16].

Lensectomy offers more than just visual rehabilitation; it also prevents increasing glaucomatous damage, reduces zonular strain, and reduces long-term morbidity. Furthermore, early surgical intervention in younger patients helps to preserve functional eyesight throughout important stages of learning and development, which improves overall quality of life. As a result, rigorous patient selection, personalised surgical planning, and ongoing postoperative monitoring are critical for achieving optimal results and lowering the risk of complications [6].

Clinical Scope of Lensectomy

For young individuals, a refractive lensectomy is a suitable surgical choice to enhance both eyesight and general quality of life. In addition to treating myopia, lensectomy also treats glaucoma caused due to intermittent pupillary obstruction. Among the young-aged patients, maintaining the posterior capsule's ability to keep both posterior and anterior compartments of the eye apart is essential in preventing the vitreous loss, Cystoid Macular Oedema (CME), and Retinal Detachment (RD) [6].

The rationale for lensectomy in individuals with MSP and associated lens abnormalities are varied and frequently occur when conservative therapies fail. Surgery is often recommended when optical correction is rendered ineffective due to substantial diplopia or when ametropia develops as a result of the lens equator extending into the pupillary centre, interfering with normal visual function. Another major indication is progressive lens subluxation, which predisposes patients to severe visual distortion and subsequent problems. Furthermore, substantial lens opacification that causes to vision loss requires surgical removal. Recurrent or sporadic pupillary block, secondary glaucoma, posterior lens displacement, and anterior chamber lens with corneolenticular touch are all examples of more severe problems that may need lensectomy. In situations of approaching complete lens luxation, surgical intervention is crucial to avoid permanent vision-threatening complications and restore ocular stability [14].

There is still uncertainty on the diagnosis, timing, and method of extracting the subluxated or dislocated lens. An appropriate age for intervention has not yet been precisely determined, as suggested by existing research on the topic [17]. According to Rao DP et al., the mean age for lensectomy is 12 years old [18].

Surgical Strategies

Surgical care of MSP in young-aged population necessitates a personalised strategy that includes both visual rehabilitation and avoidance of subsequent problems like as glaucoma and lens displacement. The surgical technique is determined by the degree of lens subluxation, capsular stability, and any concomitant ocular comorbidities.

Options include incision, lensectomy, ophthalmic visco-surgical device, lens extraction with or without IOL implantation, Capsulorhexis, Phacoaspiration, along with advanced procedures which include scleral-fixated or iris-claw IOLs, capsular tension devices, or phased interventions in situations of increasing zonular weakening [8].

Given the specific anatomical constraints presented by spherical lenses, extensive surgical planning and intraoperative changes are required to maximise visual benefits while minimising risks. Long-term follow-up is also critical, since these patients frequently require continued therapy to resolve problems and maintain functional eyesight during their lifetime [18].

For lens subluxation, Hoffman RS et al., suggested the following classification:

1. Mild: 0% to 25% of the dilated pupil is visible through the lens edge.
2. Moderate: 25% to 50% of the dilated pupil is shown by the lens edge.
3. Severe: more than 50% of the pupil is exposed by the lens edge.

The individuals should be examined with the head inclined backwards while seated and in the supine position. The preoperative assessment is essential because a Pars Plana Lensectomy (PPL) is a superior way to treat lenses that seem manageable when the patient is standing up but subluxate more when they are lying down. The individual's young age and the progressive nature of the zonulopathy must be taken into consideration while choosing the best course of action [19].

- **Incision**

Patients who are older than 10 years can be given temporal clear corneal incisions due to their scleral stiffness, which needs to be considered. To guarantee a tight seal, a superior scleral tunnel incision is suggested for younger children. To lessen the stress throughout phacoemulsification, it is recommended that the incision be made in the quadrant far from zonular dehiscence [20]. In a prospective study conducted by Khokhar S et al., several surgical procedures (including scleral-tunnel phacoemulsification and lensectomy approaches) were outlined for spherophakia patients. The authors described a considerable reduction in Best-Corrected Visual Acuity (BCVA) following surgery and highlighted how scleral-tunnel or controlled bigger incisions can be beneficial in extremely small/subluxated lenses by reducing stress on the capsular bag and zonules and facilitating capsular support devices [21]. Another study done by Zheng J et al., studied the predictors of visual prognosis after lens surgery in MSP including preoperative IOP, degree of subluxation, and surgical technique. The authors observed that surgical method (including an incision offered appropriate access while minimising produced trauma/astigmatism) and early visual rehabilitation eventually impacted the ultimate acuity [22].

- **Ophthalmic visco-surgical devices**

It is crucial to employ Ophthalmic Viscosurgical Devices (OVDs) appropriately. Vitreous prolapse can be avoided using dispersives to seal the zonular dehiscence location even before rhexis is started. Prior to phacoemulsification, anterior vitrectomy should be carried out if vitreous prolapse is observed to prevent excessive vitreous traction [19]. For the young individuals with limited scleral stiffness, capsulorhexis under cohesive OVD can assist in flattening the elastic anterior capsule and prevent tearouts [19]. In a prospective interventional series conducted by Khokhar S et al., the authors described the routine use of high-molecular-weight viscoelastic to deepen and stabilise the anterior chamber, preserve the corneal endothelium during extended manipulation, and tamponade the iris/zonal apparatus when inserting hooks or capsular tension devices. The paper emphasises tailoring the surgical approach (incision size/route and OVD selection) to the degree of zonular instability and the need for supplementary capsular support, noting that appropriate OVD use facilitates safer instrumentation and may reduce intraoperative zonular stress as well as endothelial injury [21]. In another study, the authors examined determinants of postoperative visual acuity following lens surgery for isolated MSP (including young patients). The author describes the use of viscoelastic agents intraoperatively to: 1) re-establish and maintain anterior chamber depth during lensectomy/phaco; 2) separate and reopen the

angle in eyes with angle compromise; and 3) protect the corneal endothelium during extended manoeuvres (capsular support device placement, scleral fixation). The study associates improved visual results with controlled surgery and fewer intraoperative problems and the authors particularly mention that judicious use of OVDs to stabilise the eye is a critical technical step, but OVD type was not compared [22].

- **Capsulorrhexis**

Each capsular support device requires an intact capsulorhexis. Avoiding peripheral tearouts can be achieved by erring on the side of modest rhexis and enlarging later, if necessary. It is possible to begin capsulorhexis in subluxated lenses away from the zonular dialysis region, allowing the remaining undamaged zonules to provide sufficient countertraction. Iris hooks or Mackool's capsular support system can be employed for counter-traction and stabilising the capsular bag at the time of capsulorhexis. A cortical cleaving hydrodissection can be carried out either before or during the implantation of the capsular devices for support following the completion of capsulorhexis [19]. In a case report presented by Bhattacharjee H et al., the authors discussed the case of a young patient with MSP and subsequent angle-closure glaucoma bout clear-lens extraction and IOLs implantation. As the lens was unstable and the anterior capsule was small/tightly curled, the authors conducted a bimanual capsulorrhexis, which involved stabilising the capsule edge with an iris hook inserted through a side port while ripping the anterior capsule. They report effective regulation of intraocular pressure along with improved visual acuity after surgery [23].

- **Phacoaspiration**

Multi-quadrant mild hydrodissection can alleviate excessive zonular strain during capsule stabilisation. With low fluidics, low bottle height, and vacuum settings, the majority of spherophakic lenses following lensectomy are clear or have an early cataract, requiring just safe Irrigation Aspiration (I/A) of the lenticular material. After using the Capsular Tension Ring (CTR), the cortex should be removed along a vector perpendicular to the capsular bag fornices to reduce the possibility of further zonular loss. Bimanual I/A works well in these difficult circumstances.

When the bag is empty, it can be inflated using cohesive viscoelastics. If a late CTR implantation is being considered, it should be implanted with adequate counter-traction from capsular hooks which firmly bind the bag to the eyewall. Otherwise, the already weak bag may subluxate even more due to the centrifugal movement of the CTR, resulting in the vitreous loss. To prevent the aspiration of the posterior capsule, which might bounce forward, the anterior chamber must be continually filled with viscoelastics [20]. In the literature, there are mentions of case reports where the aspiration techniques used were not strictly labelled as "phacoaspiration", but use of term "lens aspiration" was coined [24,25].

- **IOL implantation**

Except in very early newborns, aphakia might not be a possibility due to the advent of novel surgical procedures to support the bag and alternate the IOL fixation methods. Additionally, this lessens the strain on the amblyopic child with poorly tolerated glasses and/or contact lenses, placed on the children and their parents. However, the degree of subluxation and the surgeon's experience must be considered while deciding whether to implant an IOL. When the bag's stability is guaranteed by strong CTR/M-CTR support, a foldable, hydrophobic acrylic IOL is the most suitable option. Other IOL attachment methods can be used, though they may not always be feasible [5].

- **Iris fixated IOL**

There are advantages and disadvantages of suturing the IOL's haptics to the iris or sclera in cases when there is insufficient capsule or zonular support. Since the effective lens position of iris-fixated posterior chamber IOLs is comparable to that of in-the-bag IOLs, they do not influence the IOL's refractive correction. Additionally, attaching flexible haptics to the iris produces a modest posterior vault that separates the optic from the iris's posterior surface, minimising pigment dispersion and chaffing [26].

The AR40e or a three-piece foldable IOL can be iris-fixed at haptics using Seipser slipknots or McCannel sutures. It is often preferable to use a long, curved needle with a 10-0 polyester or 10-0 polypropylene suture. Due to the absence of scleral or conjunctival incisions and the decreased need for vitrectomy, the IFIOL approach is less taxing than Scleral Sutured IOL (SFIOLs); therefore, it can be performed comfortably by the anterior segment surgeon [6].

Pupil peaking, iris chaffing, and the ensuing pigment dispersion can be prevented by carefully positioning the bites at the mid-peripheral iris without applying undue strain. Though acute pigment dispersion develops perioperatively, progressive pigmentary glaucoma is not yet documented as a prevalent late consequence. If there is inadequate iris tissue support, it is appropriate to avoid using this procedure [19]. In a case report presented by Moshirfar M et al., the case of a 23-year-old patient with idiopathic MSP had bilateral iris-fixed phakic IOL (Verisyse/Artisan) implantation which addressed the excessive myopia. At four years post-op, the patient's uncorrected distance vision was 20/25 (best corrected 20/20-1) in both eyes, with no intraoperative or postoperative problems observed. The authors emphasised the cautious patient selection (adequate anterior chamber depth, no lens displacement) and suggest endothelial cell and anatomical monitoring [27].

- **Scleral fixated IOL**

When there is inadequate iris, capsular, and zonular support, SFIOL may be used. Compared to IFIOLs, this method should result in less pigment dispersion since it makes less contact with the uveal tissue. In a study done by Kumar B, Muni I, two sclerotomies are performed 1.5-2 mm posterior to the limbus following the creation of two partial-thickness scleral flaps and a 6-7 mm scleral incision [20]. To encourage proper scar development and avoid future lens displacement, this landmark is essential for implanting haptics and placing sutures in the ciliary sulcus region. In research done by Mehta R and Aref AA, the use of A PMMA IOL with fixation eyelets can then be fixed to the sclera using a double-armed 10-0 Prolene suture using a straight needle [28]. Subbiah S et al., found that after sutured-SFIOL was employed in their series of eight patients with MSP, there was satisfactory control of IOP without any intraoperative or postoperative problems [29]. Nevertheless, this procedure may result in several problems, including vitreous entrapment, PAS, IOL tilt, suprachoroidal haemorrhage, vitreous haemorrhage, and RD [30].

Suture deterioration, breaking, erosion, knot untying and haptic sliding from the suture are among the possible reasons for displacement of an SFIOL. In SFIOL and capsular support systems, late polypropylene suture deterioration is a significant problem. Suture erosion following sutured SFIOL has been reported to occur in 0-28.5% of cases, which may raise the possibility of suture tract endophthalmitis. According to published literature, up to 24% of sutures rupture [31]. Histopathologic investigations have shown that 10-0 polypropylene sutures can gradually degrade, even when the suture knots are secure, in eyes with late IOL displacement, which occurs 7-14 years following surgery. This deterioration appears to be hastened by mechanical stress on the suture, such as repeated blinking and ocular movements on a suspended, scleral-fixated IOL. Hydrolytic degradation of the polypropylene might cause late IOL decentration or dislocation [32,33].

The Best Method for IOL Fixation and Lensectomy

In these spherophakic eyes with increasing zonulopathy, the anterior segment surgeon conducting the limbal lensectomy requires the assistance of suitable OVDs, hooks, rings, careful fluidics, and IOL

insertion technique to guarantee long-term centration within the IOL-bag complex. Maintaining the posterior capsule's integrity lowers the chance of posterior segment issues, particularly in children with Marfan syndrome. On the other side, a PPL combined with SFIOL somewhat lessens the surgical complexity. Therefore, how does one decide which lensectomy procedure is best?

Yang J et al., conducted a prospective, nonrandomised, interventional case series in which patients with secondary glaucoma and spherophakia were divided into two groups and treated with either PPL + SFIOL or phacoemulsification + CTR + IOL. During a three-year follow-up, the authors discovered that both methods were successful in reducing intraocular pressure and enhancing visual acuity [34]. In a different comparison investigation, 14 children, whose mean ages were 8.06 ± 4.49 years, had 28 eyes treated for lens subluxation using either PPL + bonded SFIOL or phacoaspiration using Cionni ring-assisted PCIOL installation. The choice of either of the procedures might depend on the operating surgeon's competence, experience, and personal taste, according to the authors, who observed no discernible difference in the incidence of complications at one year [35].

The American Academy of Ophthalmology determined that there was not enough data to prove that one kind of lens or fixation site is better than another after reviewing the scientific literature regarding open-loop ACIOL, IFIOL, & SFIOL implantation without capsule support [36]. Choosing the right IOL fixation technique requires careful consideration of the patient's age, corneal condition, angle, iris, and concurrent glaucoma [6].

DISCUSSION

The MSP is a congenital lens-zonal defect marked by a smaller, spherical crystalline lens, increased lenticular myopia, and zonular instability that often affects children or young adults [37]. The lens's underdeveloped zonular apparatus and small equatorial diameter make it prone to forward displacement, pupillary block, and secondary angle-closure glaucoma, in addition to progressive lens subluxation and dislocation.

These anatomical and refractive problems increase the risk of amblyopia, uncontrolled intraocular pressure, and long-term optic nerve injury if surgery is delayed or performed inadequately [37,38]. Many young-aged patients have shown significant improvements in best-corrected visual acuity after having the aberrant lens surgically removed and receiving visual rehabilitation with an IOL, capsular support device, or aphakia correction. However, the procedure is technically demanding because of the tiny capsular bag, flexible zonules, and risk of glaucoma. Incision design, capsulorhexis integrity, IOL fixation, and postoperative refractive rehabilitation are key predictors of prognosis. Preoperative glaucoma, lens subluxation severity, and surgical technique remain important predictors of ultimate vision [5,31,38].

In a study which was conducted by Muralidhar R et al., it was found that eight patients had isolated MSP, whereas others had systemic connections. Although plasma homocysteine levels could only be measured in two cases, homocystinuria was identified as the most often related systemic disease. Additional syndromic relationships included WMS and Marfan syndrome, which were previously described. One patient also presented with Tourette syndrome; however, since MSP had not previously been linked to this illness, the cohabitation was considered accidental rather than causal. There were total of sixteen eyes (44.4%) that required lensectomy for the dislocated crystalline lens. Eventually, it was observed that lensectomy had no impact on the intraocular pressures [5].

WMS and homocysteinuria are thought to be common causes of pupillary block glaucoma, which results in angle closure. No prior reports of glaucoma incidence have been reported. There are several ways that glaucoma develops in MSP [9]. Peripheral anterior synechiae may occur because of persistent pupillary obstruction. Other processes that have been documented in a study done by Harasymowycz P et al., included angle anomalies with agenesis of angle structures, chronic pupillary block without full angle closure, and crowding of angle by spherophakic lens [39]. Even though the majority of patients in the research, which was undertaken by Cumba RJ et al., had patent Peripheral Iridotomy (PI), the incidence of glaucoma was only 44.4%. Before a PI was done, several of these individuals most likely already had angle closure and peripheral anterior synechiae. The fact that half of these individuals had high IOP at presentation supports the result [40]. Remarkably, open-angle glaucoma was identified in only one paediatric case, which was done by Khokhar S et al., this patient had no angle abnormalities and required multiple surgeries to lower her right eye's intraocular pressure. It is challenging to control glaucoma in general, and the majority of patients are required to take long-term medications in addition to surgery [41]. Cyclo-destructive treatments were necessary to regulate the intraocular pressure in three eyes. Intraocular pressures were unaffected by lensectomy, and two eyes experienced elevated IOPs throughout follow-up [8].

In another study, which was undertaken by Senthil S et al., trabeculectomy showed an excellent success rate in treating glaucoma linked to MSP, and it effectively reduced intraocular pressure for an extended period of time. At one year, the chance of complete success was 96%; at two years, it decreased to 88%; this percentage was maintained for seven years. Trabeculectomy appears to be more successful than other secondary glaucomas (54%), despite the fact that success rates cannot be directly compared because data on the procedure in eyes with MSP were insufficient [8]. According to Senthil S et al., an earlier study, glaucoma affected young individuals and was linked to MSP in 51% of the eyes at the time of presentation. A 20% of the glaucoma-affected eyes at presentation and 30% at the final follow-up were blind as a result of glaucomatous optic neuropathy [8]. This underscores the critical need for early detection and timely management of glaucoma to prevent the vision loss and blindness associated with the disease [8,10].

A few trials with shorter follow-up times showed that simple lensectomy or lens extraction helped lower intraocular pressure [8,42]. Longer follow-up studies have shown that glaucoma filtering surgery is necessary eventually during the time of follow-up to regulate the IOP in eyes having glaucoma in MSP, since lensectomy alone is not insufficient [42,43]. After a follow-up of one to two years, it was demonstrated that a combination of lens aspiration, goniosynechialysis, and peripheral iridoplasty could effectively manage the intraocular pressure in two eyes with glaucoma and synechial angle closure [44].

According to Venkataraman P et al., comparing studies that show outcomes and implications for lensectomy in MSP is difficult due to the small number of patients, the numerous reasons of lens subluxation, and the different periods of follow-up. The authors eventually concluded that though MSP is rare among young adults, it increases the likelihood of vision impairment and glaucoma. Early detection and a systematic lens- and glaucoma-directed therapy approach would result in a considerable improvement in the visual outcomes after lensectomy. Poor visual acuity before to surgery, as well as the existence of glaucoma at the time of presentation, is major indicators of a worse visual outcome. As a result, quick intervention and ongoing interdisciplinary follow-up are needed [6].

MSP may necessitate early surgical intervention to reduce IOP and avoid glaucoma damage. If left untreated, the spherical lens increases the risk of pupillary block, secondary angle-closure glaucoma, and optic nerve injury [8]. Lensectomy, with or without IOL implantation, can ease pupillary block, deepen the anterior chamber, and stabilise IOP, hence maintaining vision [45,46].

Preliminary examinations of lens shape, anterior chamber depth, and IOP in children can guide the prompt diagnosis and referral to tertiary

centres for advanced evaluation and effective surgery planning [33]. Overall, early detection and recommendation by family physicians is critical for avoiding vision loss. A multidisciplinary strategy that includes both ocular and systemic evaluations improves long-term results for young children with this uncommon but potentially blinding illness.

CONCLUSION(S)

The MSP should be suspected if there is bilateral high myopic refractive error, subluxation, or crystalline lens dislocation. According to our research, following a lensectomy, individuals with MSP have a notable improvement in their vision. The presence of glaucoma at the time of diagnosis, high intraocular pressure, and corrected visual acuity at the initial visit were all substantial risk factors for poor visual outcomes after lensectomy in patients experiencing MSP. An uncommon but potentially blinding condition, MSP has a wide range of concomitant systemic symptoms and clinical manifestations. Blindness from glaucoma can be avoided with early disease detection, prompt vision rehabilitation, and proper lens and glaucoma care. It is advised to have multidisciplinary care with lifetime monitoring.

Authors' contribution: SS: Compiled and wrote the article. SD: Guided the writing of the article. All authors reviewed the article.

Availability of data and materials: The English-language papers that were accessible were compiled from a variety of databases, including PubMed, Scopus, WoS, Springer Nature, and Google Scholar. To handle MSP, a multidisciplinary approach employing diagnostic instruments, knowledge, and experience is required.

REFERENCES

[1] Yu X, Chen W, Xu W. Diagnosis and treatment of microspherophakia. J Cataract Refract Surg. 2020;46(12):1674-79. Doi: 10.1097/j.jcrs.0000000000000334.
[2] Liu Y, Sun Y, Huo Q, Song L, Wang X, Shen X, et al. Genetic landscape and ocular biometric correlations in microspherophakia: Insights from a comprehensive patient cohort. Hum Genomics. 2025;19(1):22. Doi: 10.1186/s40246-025-00729-6.
[3] Chan RT, Collin HB. Microspherophakia. Clin Exp Optom. 2002;85:294-99. Doi: 10.1111/j.1444-0938.2002.tb03085.x.
[4] Macken PL, Pavlin CJ, Tuli R, Trope GE. Ultrasound biomicroscopic features of spherophakia. Clin Exp Ophthalmol. 1995;23:217-20. Doi: 10.1111/j.1442-9071.1995.tb00160.x.
[5] Muralidhar R, Ankush K, Vijayalakshmi P, George VP. Visual outcome and incidence of glaucoma in patients with microspherophakia. Eye (Lond). 2015;29(3):350-55. Doi: 10.1038/eye.2014.250.
[6] Venkataraman P, Haripriya A, Mohan N, Rajendran A. A systematic approach to the management of microspherophakia. Indian J Ophthalmol. 2022;70(7):2262-71. Doi: 10.4103/ijo.IJO_2888_21.
[7] Jensen AD, Cross HE, Paton D. Ocular complications in the Weill-Marchesani syndrome. Am J Ophthalmol. 1974;77(2):261-69. Doi: 10.1016/0002-9394(74)90685-0.
[8] Senthil S, Rao HL, Hoang NT, Jonnadula GB, Addepalli UK, Mandal AK, et al. Glaucoma in Microspherophakia: Presenting features and treatment outcomes. J Glaucoma. 2014;23(4):262-67. Doi: 10.1097/IJG.0b013e3182707437.
[9] Feiler-Ofry V, Stein R, Godel V. Marchesani's syndrome and chamber angle anomalies. Am J Ophthalmol. 1968;65(6):862-66. Doi: 10.1016/0002-9394(68)92211-3.
[10] Senthil S, Rao HL, Babu JG, Mandal AK, Addepalli UK, Garudadri CS. Outcomes of trabeculectomy in microspherophakia. Indian J Ophthalmol. 2014;62(5):601-05. Doi: 10.4103/0301-4738.129785.
[11] Dietlein TS, Mietz H, Jacobi PC, Krieglstein GK. Spherophakia, nanophthalmia, hypoplastic ciliary body and glaucoma in brachydactyly-associated syndromes. Graefes Arch Clin Exp Ophthalmol. 1996;234:187-92. Doi: 10.1007/BF02343070.
[12] Willi M, Kut L, Cotlier E. Pupillary-block glaucoma in the Marchesani syndrome. Arch Ophthalmol. 1973;90(6):504-08. Doi: 10.1001/archopht.1973.01000050504020.
[13] Veeraraghavan N. Adult eye conditions: Primary open-angle glaucoma and cataract. FP Essent. 2022;519:19-23. Available from: https://pubmed.ncbi.nlm.nih.gov/35947132/.
[14] Wu-Chen WY, Letson RD, Summers CG. Functional and structural outcomes following lensectomy for ectopia lentis. J AAPOS. 2005;9(4):353-57. Doi: 10.1016/j.jaapos.2005.03.004.
[15] Malik KP, Goel R, Jain K, Nagpal S, Singh S. Management of bilateral microspherophakia with secondary angle closure glaucoma. Nepal J Ophthalmol. 2015;7(1):69-73. Doi: 10.3126/nepjoph.v7i1.13174.
[16] Maharana PK, Sahay P, Mandal S, Lakshmi CC, Goel S, Nagpal R, et al. jks, w. Indian J Ophthalmol. 2022;70(7):2432-38. Doi: 10.4103/ijo.IJO_255_22.
[17] Simon MA, Origlieri CA, Dinallo AM, Forbes BJ, Wagner RS, Guo S. New management strategies for ectopia lentis. J Pediatr Ophthalmol Strabismus. 2015;52(5):269-81. Doi: 10.3928/01913913-20150714-02.
[18] Rao DP, John PJ, Ali MH, Kekunnaya R, Jalali S, Garudadri CS, et al. Outcomes of lensectomy and risk factors for failure in spherophakic eyes with secondary glaucoma. Br J Ophthalmol. 2018;102(6):790-95. Doi: 10.1136/bjophthalmol-2017-310861.
[19] Hoffman RS, Snyder ME, Devgan U, Allen QB, Yeoh R, Braga-Mele R. Management of the subluxated crystalline lens. J Cataract Refract Surg. 2013;39(12):1904-15. Doi: 10.1016/j.jcrs.2013.09.005.
[20] Kumar B, Muni I. Scleral Fixation of Intraocular Lenses. [Updated 2023 Jul 3]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2025 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK570585/.
[21] Khokhar S, Pillay G, Sen S, Agarwal E. Clinical spectrum and surgical outcomes in spherophakia: A prospective interventional study. Eye (Lond). 2018;32(3):527-36. Available from: https://doi.org/10.1038/eye.2017.229.
[22] Zheng J, Cheng L, Chen Z, Chen T, Jiang Y. Factors related to visual outcomes after lens surgery in isolated microspherophakia. J Ophthalmol. 2022;2022:9089203. Doi: 10.1155/2022/9089203.
[23] Bhattacharjee H, Bhattacharjee K, Medhi J, DasGupta S. Clear lens extraction and intraocular lens implantation in a case of microspherophakia with secondary angle closure glaucoma. Indian J Ophthalmol. 2010;58(1):67-70. Doi: 10.4103/0301-4738.58477.
[24] Karmoun S, Aboulanouar A, Hajji Z, Elmarzouqi B, Elhassan A, Berraho A. High myopia associated with advanced glaucoma due to microspherophakia: A case report. Austin J Clin Ophthalmol. 2020;7(1):1108.
[25] Kanamori A, Nakamura M, Matsui N, Tomimori H, Tanase M, Seya R, et al. Goniosynechialysis with lens aspiration and posterior chamber intraocular lens implantation for glaucoma in spherophakia. J Cataract Refract Surg. 2004;30(2):513-16. Doi: 10.1016/S0886-3350(03)00670-9.
[26] Stem MS, Todorich B, Woodward MA, Hsu J, Wolfe JD. Scleral-Fixated Intraocular Lenses: Past and Present. J Vitreoretin Dis. 2017;1(2):144-52. Doi: 10.1177/2474126417690650.
[27] Moshirfar M, Meyer JJ, Schliesser JA, Espandar L, Chang JC. Iris-fixated phakic intraocular lens implantation for correction of high myopia in microspherophakia. J Cataract Refract Surg. 2010;36(4):682-85. Doi: 10.1016/j.jcrs.2009.08.043.
[28] Mehta R, Aref AA. Intraocular lens implantation in the ciliary sulcus: Challenges and risks. Clin Ophthalmol. 2019;13:2317-23. Doi: 10.2147/OPTH.S205148.
[29] Subbiah S, Thomas PA, Jesudasan CA. Scleral-fixated intraocular lens implantation in microspherophakia. Indian J Ophthalmol. 2014;62:596-600. Doi: 10.4103/0301-4738.129787.
[30] Solomon K, Gussler JR, Gussler C, Van Meter WS. Incidence and management of complications of transsclerally sutured posterior chamber lenses. J Cataract Refract Surg. 1993;19(4):488-93. Doi: 10.1016/s0886-3350(13)80612-8.
[31] Cheung CS, VanderVeen DK. Intraocular lens techniques in pediatric eyes with insufficient capsular support: Complications and outcomes. Semin Ophthalmol. 2019;34(4):293-302. Doi: 10.1080/08820538.2019.1620809.
[32] Price MO, Price FW, Jr, Werner L, Berlie C, Mamalis N. Late dislocation of scleral-sutured posterior chamber intraocular lenses. J Cataract Refract Surg. 2005;31(7):1320-26. Doi: 10.1016/j.jcrs.2004.12.060.
[33] McAllister A, Hirst L. Visual outcomes and complications of scleral-fixated posterior chamber intraocular lenses. Journal of cataract and refractive surgery. 2011;37(7):1263-69. Doi: 10.1016/j.jcrs.2011.02.023.
[34] Yang J, Fan Q, Chen J, Wang A, Cai L, Sheng H, et al. The efficacy of lens removal plus IOL implantation for the treatment of spherophakia with secondary glaucoma. Br J Ophthalmol. 2016;100(8):1087-92. Doi: 10.1136/bjophthalmol-2015-307298.
[35] Thapa BB, Agarwal A, Singh R, Gupta PC, Ram J. Phacoaspiration with a Cionni ring versus pars plana lensectomy, vitrectomy and sutureless transscleral IOL fixation in pediatric patients with a subluxated lens. Graefes Arch Clin Exp Ophthalmol. 2016;254(5):901-09. Doi: 10.1007/s00417-016-3297-y.
[36] Wagoner MD, Cox TA, Ariyasu RG, Jacobs DS, Karp CL. Intraocular lens implantation in the absence of capsular support: A report by the American Academy of Ophthalmology. Ophthalmology. 2003;110(4):840-59. Doi: 10.1016/s0161-6420(02)02000-6.
[37] Şimşek T, Beyazyıldız E, Şimşek E, Öztürk F. Isolated Microspherophakia Presenting with Angle-Closure Glaucoma. Turk J Ophthalmol. 2016;46(5):237-40. Doi: 2016 Oct 17. PMID: 28058167; PMCID: PMC5200837.
[38] Kaur K, Gurnani B. Microspherophakia. [Updated 2023 Jun 11]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2025 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK576395/.
[39] Harasymowycz P, Wilson R. Surgical treatment of advanced chronic angle closure glaucoma in Weill-Marchesani syndrome. J Pediatr Ophthalmol Strabismus. 2004;41 (5):295-99. Doi: 10.3928/01913913-20040901-08.
[40] Cumba RJ, Nagi KS, Bell NP, Blieden LS, Chuang AZ, Mankiewicz KA, et al. Clinical outcomes of peripheral iridotomy in patients with the spectrum of chronic primary angle closure. ISRN Ophthalmol. 2013;2013:828972. Doi: 10.1155/2013/828972.
[41] Khokhar S, Pangtey MS, Sony P, Panda A. Phacoemulsification in a case of microspherophakia. J Cataract Refract Surg. 2003;29(4):845-47. Doi: 10.1016/s0886-3350(02)01617-6.
[42] Yasar T. Lensectomy in the management of glaucoma in spherophakia: Is it enough? J Cataract Refract Surg. 2003;29(6):1052-53. Doi: 10.1016/s0886-3350(03)00397-3.

[43] Sun Y, Chen Z, Liu Y, Huo Q, Jia WN, Zhao Z, et al. Novel classification for microspherophakia and its related surgical implications: A retrospective cohort study. BMJ Open Ophthalmol. 2025;10(1):e002063. Doi: 10.1136/bmjophth-2024-002063.
[44] Nie L, Pan W, Fang A, Li Z, Qian Z, Fu L, et al. Combined phacoemulsification and goniosynechialysis under an endoscope for chronic primary angle-closure glaucoma. J Ophthalmol. 2018;2018:8160184. Doi: 10.1155/2018/8160184.
[45] Ardjomand N, Kölli H, Vidic B, El-Shabrawi Y, Faulborn J. Pupillary block after phakic anterior chamber intraocular lens implantation. J Cataract Refract Surg. 2002;28(6):1080-1. Doi: 10.1016/s0886-3350(01)01114-2.
[46] Alió JL, Abdelrahman AM, Javaloy J, Iradier MT, Ortuño V. Angle-supported anterior chamber phakic intraocular lens explantation causes and outcome. Ophthalmology. 2006;113(12):2213-20. Doi: 10.1016/j.ophtha.2006.05.057.

PARTICULARS OF CONTRIBUTORS:
1. Postgraduate Student, Department of Ophthalmology, Datta Meghe Institute of Higher Education and Research, Wardha, Maharashtra, India.
2. Professor and Head, Department of Ophthalmology, Datta Meghe Institute of Higher Education and Research, Wardha, Maharashtra, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Suyash Singh,
Department of Ophthalmology, Ground Floor, AVBRH building, DMIHER, Sawangi, Meghe, Wardha-442107 Maharashtra, India.
E-mail: suyashsingh9990@gmail.com

PLAGIARISM CHECKING METHODS: [Jain H et al.]
- Plagiarism X-checker: May 11, 2025
- Manual Googling: Dec 29, 2025
- iThenticate Software: Dec 31, 2025 (10%)

ETYMOLOGY: Author Origin

EMENDATIONS: 7

AUTHOR DECLARATION:
- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? No
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **May 10, 2025**
Date of Peer Review: **Aug 30, 2025**
Date of Acceptance: **Jan 02, 2026**
Date of Publishing: **Apr 01, 2026**