A Case of Marfan Syndrome

Patients with this syndrome have a higher risk of glaucoma and retinal detachment.

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Managing the subluxed lens of a patient with Marfan syndrome is one of the greatest challenges in contemporary lens-based surgery. Conventional approaches include intracapsular lens extraction or pars plana lensectomy with vitrectomy. After the lens is removed, glasses, contact lenses, iris- or angle-supported anterior chamber IOLs, or scleral sutured posterior chamber IOL can be used to correct the patient's aphakia.

Patients with Marfan syndrome have an increased risk of glaucoma and retinal detachment; up to 11% of patients with Marfan syndrome and 8% to 38% of those who have lens dislocation or undergone lens surgery experience a retinal detachment.[1] Anterior chamber IOLs are best avoided in these patients, and a surgical approach that preserves the capsular bag and avoids disruption of the vitreous will potentially reduce the risk of retinal detachment.

TAKE-HOME MESSAGE

- A 12-year-old with Marfan syndrome had a subluxated lens and reduced BCVA.
- Dr. Prasad and Ms. Armstrong recentered the capsular bag and implanted an in-the-bag IOL with the aid of a CTR and CTS.
- The patient's UCVA was 6/9 and 6/6 after 6 weeks postop.

CASE PRESENTATION

A pediatric ophthalmologist referred a 12-year-old boy with known Marfan syndrome to our practice after seeing him from the age of 2 years. The boy was managed reasonably with spectacle correction and maintained good vision in both eyes; however, more recently the subluxation in his right eye reduced his BCVA to 6/24. He was also on atenolol for aortic valve problems and had antenatal hydronephrosis; both were being managed by the appropriate specialists.

On the patient's first visit to our clinic, vision was 6/24 in his right eye (-24.50 +9.00 X 92.5) and 6/9+3 in his left (-3.00 +3.00 X 95). He also expressed that children at school were bullying him and stealing his glasses.

On exam, the edge of the lens was passing through the center of the pupil in his right eye; however, the subluxation was not as prominent in his left. The patient's intraocular pressure and peripheral retinal examination were normal, and there was no vitreous in the anterior chamber.

Ideally, I wanted to achieve the following parameters without disrupting the vitreous body: lens extraction through a continuous curvilinear capsulorrhexis, preserve and recenter the capsular bag, and place an in-the-bag IOL securely.

The anticipated challenges included: (1) biometry, which is crucial for a good refractive outcome, (2) achieving a central curvilinear capsulorrhexis, lens extraction, bag recentration, and IOL fixation without disrupting the anterior vitreous face, and (3) deploying techniques to reduce the risk of late complications, such as posterior capsular opacification and subluxation/dislocation of the IOL.

Achieving a central curvilinear capsulorrhexis in Marfan syndrome is difficult because the lens is unstable. The zonules' outward pull is uneven and the capsule is more elastic in younger patients, causing a tendency for the tear to run out. Even when a satisfactory capsulorrhexis is achieved, the threat of complications is not over. Also in a Marfan patient, the capsular bag may be smaller than in a normal eye, leading to an increased risk of tearing or ripping during implantation of devices to support and recenter it.[2]

HOW WOULD YOU PROCEED?

In any difficult surgical situation, experts often use different approaches to achieve the same result. For this patient, possible strategies follow one of two broad approaches: (1) sacrifice the capsular bag, clear vitreous from the anterior chamber, and implant an IOL or (2) preserve the capsular bag, recenter it, and implant a secure in-the-bag IOL.

Approach No. 1. If this strategy is chosen, lens extraction

Figure 1. Capsular Tension Segment.

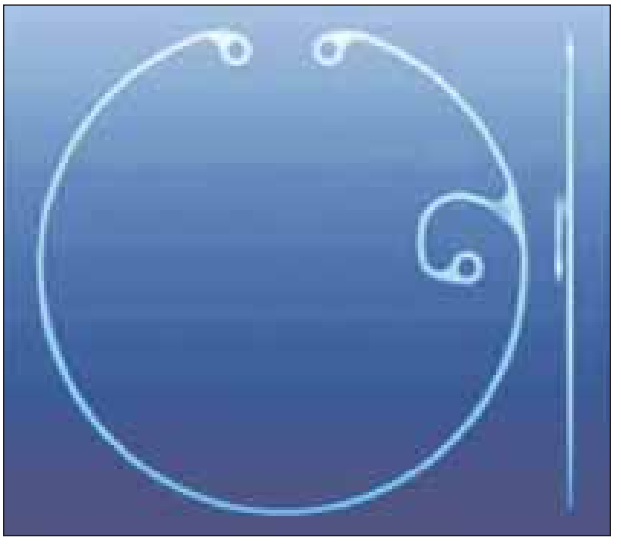

Figure 2. Modified Capsular Tension Ring.

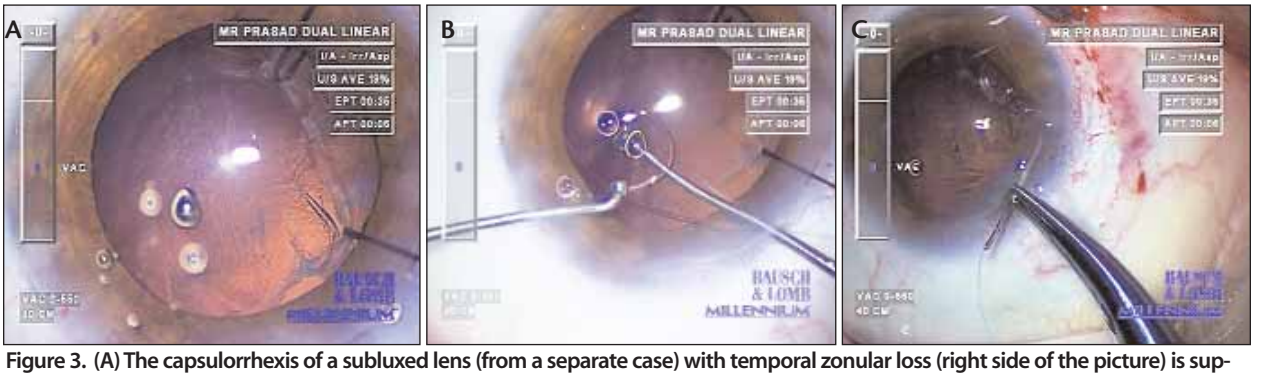

Figure 3. (A) The capsulorrhexis of a subluxed lens (from a separate case) with temporal zonular loss (right side of the picture) is supported by two iris retractors. The bag is thus stabilized, allowing completion of phacoemulsification. (B) The MCTR with a double armed 9-0 Prolene suture passed through its central islet is placed into the capsular bag, and the anteriorly displaced central segment is positioned in front of the rhexis with a dialing hook, while a second instrument is poised to guide the trailing end in. (C) The CIF-4 needles are passed behind the iris, but in front of the anterior capsule to emerge 1.5 mm behind the limbus under a previously prepared scleral flap. This secures the MCTR, and the Prolene knot is covered by the scleral flap. A secure in-the-bag IOL is then placed.

can be done with an anterior or pars plana approach. A pars plana approach allows a thorough vitrectomy and a full internal search of the peripheral retina. A complete vitrectomy is appealing in this case because it eliminates future vitreoretinal traction; however, it is difficult to induce a posterior vitreous separation in young patients, and attempting this may induce retinal tears.

Alternatively, a deep anterior vitrectomy may be preferable. An IOL may then be implanted in the anterior chamber; anterior chamber open loop[3,4] or iris fixated (Artisan; Ophtec, Groningen, Netherlands)[5] IOLs are simple and efficient techniques. This technique comes with its own set of risks, including iritis, pigment dispersion, corectopia, glaucoma, and endothelial cell loss. Therefore, many surgeons implant a posterior chamber IOL, fixated with transscleral sutures in the ciliary sulcus.[6] To avoid future suture breakage—leading to subluxation or dislocation of the IOL—and achieve better centration, the elegant technique of sutureless intrascleral posterior chamber fixation has been developed.[7]

Approach No. 2. Techniques that preserve the capsular bag are intellectually more appealing, especially if the bag can be recentered and an in-the-bag IOL implanted securely. If we do not disrupt the vitreous body, then at least theoretically the risk of complications, such as future retinal detachment, is reduced. A capsular tension ring (CTR) can be used to stabilize the capsular bag; however, if a CTR is used in an eye with a subluxated lens, capsular bag decentration is not corrected.[8] Lam and colleagues described suturing the CTR and the capsular bag to the sclera, thus improving capsule centration.[9] Passing sutures through the capsular bag risks tearing it and therefore may not be advisable. Ahmed intro-

duced the Capsular Tension Segment (CTS; Morcher GmbH; Figure 1)[11] to avoid over-stretching or tearing the unstable bag. Cionni proposed a Modified Capsular Tension Ring (MCTR; Morcher GmbH, Stuttgart, Germany; Figure 2) to avoid passing the sutures through the capsular bag.[10] For an example of how to implant the MCTR, see Figure 3.

If this were your patient, how would you proceed?

HOW WE PROCEEDED

I chose an approach that preserved and recentered the capsular bag, allowing in-the-bag IOL implantation and fixation of the bag to the ciliary sulcus. I used a combination of a CTR and CTS.The surgical workup in such a case has to be meticulous and should include all physicians involved in the patient's care.

When doing biometry on a patient with Marfan syndrome, you must carefully choose what section of the axial length measurement to take (ie, phakic or aphakic portion) and adjust your instrument for the presence or absence of lens material. Depending on the extent of the subluxation, you may need to dilate the pupil to ensure proper alignment of the beam. If using A-scan ultrasound, you can confirm the position of the probe by looking for the echo corresponding to the presence of the lens boundaries.

The patient's phakic axial length measurements were 25.50 in the right eye and 21.46 in the left. The aphakic axial length in his right eye was 25.29 and 21.25 in his left. Using the aphakic measurement, we decided to use an 18.00 D IOL (SofPort AO; Bausch & Lomb, Rochester, New York) for emmetropia. This lens has a 360º square edge, and the SofPort injection system (Bausch & Lomb) delivers it into the capsular bag in a fairly flat position, minimizing the potential for destabilizing an already unstable bag.

Surgery was performed under general anesthetic. Figure 4A shows the patient's subluxed and decentered lens. A fornix-based conjunctival flap is made in the zone of maximum subluxation, followed by the creation of a partial thickness scleral flap with its base toward the fornix. The 4 mm wide scleral dissection extends approximately 3 mm

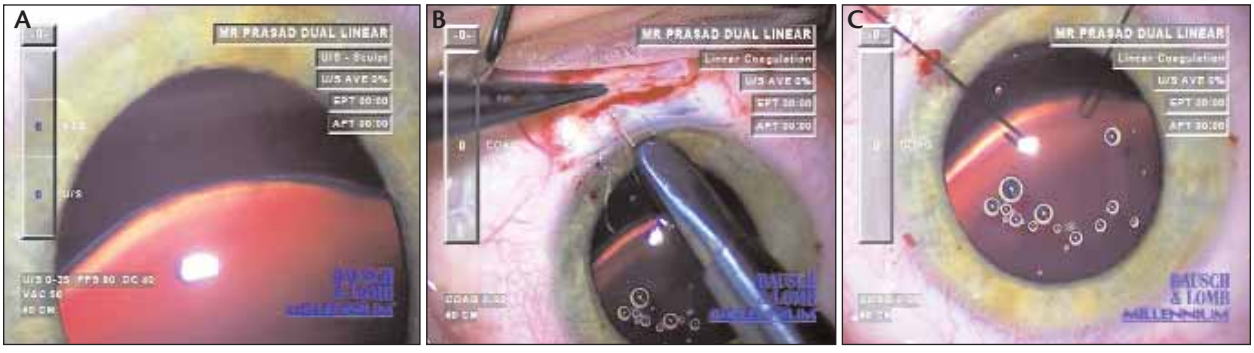

Figure 4. (A) Preoperative findings of a subluxed and decentered lens. (B) A scleral flap is placed after a fornix-based conjunctival flap is made in the area of maximal lens decenteration. (C) Flexible iris retractors are positioned through two paracentasis incisions to support the capsulorrhexis as it develops.

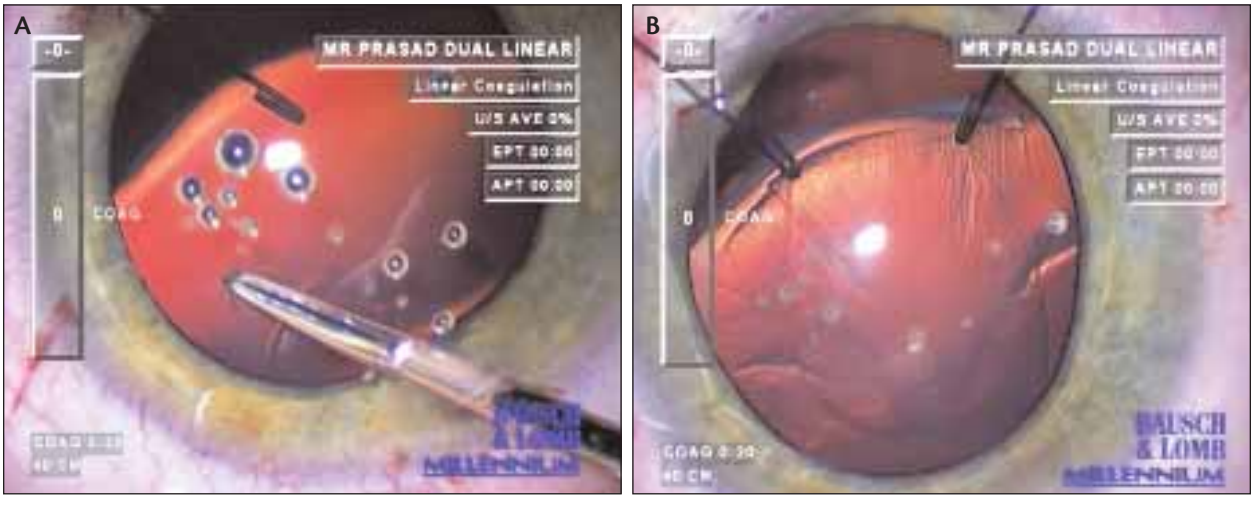

Figure 5. (A) The capsulorrhexis starts in the center of the lens, which is lying close to the edge of the iris due to the decentration lens. (B) The capsulorrhexis is developed and the preplaced iris retractors engage the capsule edge and draw the lens to a central position.

Figure 6. (A) A CTR is placed in the capsular bag. (B) A CTS with a Prolene suture already passed through the central islet is then positioned into the capsular bag. (C)The IOL is then implanted into the capsular bag.

peripherally from the limbus (Figure 4B). Two paracentecis incisions are located on the sides of the scleral flap, through which flexible iris retractors support the capsulorrhexis as it develops (Figure 4C).

The capsulorrhexis is started close to the iris edge at the 12-o'clock position (ie, center of the lens; Figure 5A). Viscoat (Alcon Laboratories, Inc., Fort Worth, Texas) is used to fill the anterior chamber and tamponade the vitreous face. The capsulorrhexis is developed, taking care to keep 3 mm from the lens equator to facilitate later placement of the CTR and CTS. An adequate anterior capsule is necessary to avoid the CTR/CTS flipping forward, out of the bag and into the anterior chamber later in the operation.

The tear is developed a few millimeters beyond the area where the iris retractor is poised to engage. Once the tear is clear, the iris retractor, engaged near the edge of the rhexis, gently draws the lens equator to the periphery to recenter the capsular bag. The rhexis is developed further, and the second iris hook is engaged and drawn centripetally, further recentering the capsular bag and allowing the superior capsule to come into view (Figure 5B). The rhexis can then be completed, followed by gentle hydrodissection using small, slow aliquots of fluid.

A CTR is placed to stretch the capsular bag (Figure 6A); phacoemulsification may then be performed without threat of the bag flopping onto the probe. Once the lens matter is removed, the CTS is positioned in the bag. A double-armed 9-0 Prolene suture on CIF-4 needles is passed through the central islet of the CTS before it is guided into the capsular bag (Figure 6B).

The IOL is then implanted into the capsular bag (Figure 6C) and the CIF-4 needles are passed in front of the anterior capsule (but behind the iris) and emerge approximately 1.5 mm away, both behind the limbus and under the previously designed scleral flap. The iris retractors are then released, allowing the surgeon to better judge the position of the capsular bag. The suture is tightened (Figure 7), and the incisions are sutured after removing viscoelastic. Intracameral cefuroxime injected at the end for endophthalmitis prophylaxis.

FOLLOW-UP

Two weeks postoperatively, the boy's UCVA recovered to 6/18 and 6/12 with pinhole. At 6 weeks, his vision was 6/6 with minimal refraction (-1.00 +1.25 X 180), and the IOL was well centered. He requested surgery for his other eye.

The only concern was that one end of the 9-0 Prolene suture used to secure the CTS was visible under conjunctiva, as the scleral flap had slightly contracted.

WHAT HAVE WE LEARNED FROM THIS CASE?

I learned three main lessons from this case and other similar cases. First, instability of the anterior chamber must be avoided at all costs—mainly to avoid the vitreous coming forward. Initially, I deployed a ophthalmic viscosurgical device (OVD) exchange technique to avoid the capsular bag diaphragm flopping forward and backward. Every time the phaco probe was removed from the eye, I injected OVD through a sideport incision, thus maintaining chamber support. While injecting the OVD, I would gradually ease off the irrigation and safely withdraw the probe, maintaining a deep anterior chamber and preventing the capsular bag from coming forward.

Second, I learned how to avoid contraction of the scleral flap. Instead of conjunctival dissection and a scleral flap, I make a 600-µm limbal incision with a guarded diamond blade in the region of maximal lens edge decentration. A crescent blade is then used to dissect a 4 X 3 mm (from limbus to periphery) partial thickness scleral pocket. This allows the Prolene knot to be buried under the sclera without disturbing the conjunctiva. As the sides of the pocket are attached, there is no real risk of scleral flap contraction.

Third, using bimanual irrigation and aspiration (no phaco) to remove the lens matter through two paracentesis incisions (1 mm each) allows greater stability and reduces the risk of vitreous disturbance. This maneuver should be only conducted when the lens matter is soft (as in this young patient). Once all lens matter has been removed, it is safe to make a 3-mm incision for CTS and subsequent IOL placement.

Figure 7. Once the CTS has been secured by two passes of the Prolene suture, the bag is nicely centered with a well-placed IOL.

CONCLUSION

I have presented evolving techniques for lens surgery in Marfan syndrome. Although some aspects are debatable, I believe that the combination of a CTR and a CTS fixated transclerally under a scleral pocket allows secure in-the-bag IOL placement and potentially reduces the risk of future complications. ■

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