Introduction:

Management of the subluxed lens in Marfan’s syndrome presents one of the most challenging situations in contemporary lens surgery. Conventional approaches have included intracapsular lens extraction or a pars plana lensectomy with vitrectomy. Once the lens was removed, aphakia was corrected by glasses, contact lenses or an iris or angle supported anterior chamber IOL or a scleral sutured posterior chamber IOL. Marfan’s syndrome patients are known to have an increased risk of glaucoma as well as retinal detachment (upto 11% of patients with Marfan syndrome, and 8–38% in those who have dislocation of the lens or have undergone lens surgery) [1]. Accordingly, anterior chamber implants are best avoided in this setting and a surgical approach which preserves the capsular bag and avoids disruption of the vitreous, will, at least potentially, reduce the risk of retinal detachment subsequently.

Considerations in the surgical approach:

Ideally one wants to achieve lens extraction through a continuous capsulorrhexis with preservation of the capsular bag, re-center the bag and secure it using a scleral fixation device and place an in-the-bag IOL, without disrupting the vitreous body.

The anticipated challenges in these cases include:

i) Systemic considerations: These patients need a full systemic workup, as they have associated features of Marfan’s which may increase the risk for an anaesthetic. They may have aortic arch or valve anamolies or indeed have had heart valve surgery already, in which case they may be on Warfarin.

ii) Biometry: Eyes with severely subluxed lenses need careful biometry to achieve a good refractive outcome. Remember that the refraction is influenced by the lens subluxation and may not correlate with the predicted IOL power.

iii) Achieving a central curvilinear capsulorrhexis: This is difficult because the lens is unstable, the outward pull provided by the zonules is uneven and the young patients capsule is much more elastic than the adult, leading to a tendency for the tear to ‘run out’.

iv) Preserving the capsular bag: Even when a satisfactory capsulorrhexis has been achieved, the game is not over. The bag of the Marfan’s patient may sometimes be smaller than that of the normal eye leading to an increased risk of tear or rip during implantation of devices to support and re-centre it [2].

v) Achieve lens extraction, bag re-centration and fixation and IOL implantation without disrupting the anterior vitreous face.

vi) Deploy techniques which reduce the risk of late complications: posterior capsular opacification and subluxation/dislocation of the IOL capsular bag complex.

(1) Possible surgical strategies

In any difficult surgical situation experts will often use different approaches to achieve the same end. For
Subluxed lenses in Marfan’s patients, possible strategies mainly follow one of two broad approaches.

One may choose to sacrifice the capsular bag, clear the anterior chamber of all vitreous and then implant an intraocular lens. Lens extraction can be done by an anterior or pars plana approach if this strategy is chosen. A pars plana approach has the advantage of allowing a thorough vitrectomy along with a full internal search of the peripheral retina. Whilst a complete vitrectomy is appealing in this setting, as it would eliminate any future vitreo-retinal traction, it is technically quite difficult to induce a posterior vitreous separation in a young patient, and attempts to achieve this may actually induce retinal tears during surgery. Because of this a deep anterior vitrectomy may be preferable. An IOL can then be implanted in the anterior chamber. Anterior chamber IOLs, open loop 2,3 or iris fixated (Artisan) 5 are simple and efficient techniques, but risks include iritis, pigment dispersion, corectopia, glaucoma, and endothelial loss. Because of these potential problems many surgeons choose to implant a posterior chamber lens in the ciliary sulcus, fixated by trans scleral sutures 6. To avoid the risk of future suture breakage leading to subluxation or dislocation of the IOL and to achieve better centration, the elegant technique of sutureless intrascleral posterior chamber fixation has been developed 7,8.

Techniques which preserve the capsular bag are intellectually more appealing, especially if the bag can be re-centred and secured with an in-the-bag IOL implanted. If all this can be achieved without disrupting the vitreous body, then (at least theoretically), the risk of complications like future retinal detachment can be reduced. A capsular tension ring (CTR) can be used to stabilise the capsular bag. However, CTR implantation in eyes with a subluxated lens does not correct capsular bag decentration 9. Lam and co authors overcame this obstacle by implanting a CTR and then suturing it and the capsular bag to the sclera to improve capsule centration 10. However passing sutures through the
capsular bag risks tearing it, and therefore may not be advisable. Cionni proposed the use of a modified (Fig.1) capsular tension ring (mCTR), as this device avoided the need to pass sutures through the capsular bag. Ahmed introduced the capsular tension segment (CTS), this smaller device is much easier to manoeuvre into the bag and position as required and avoids the risk of over stretching or tearing the unstable bag. (Fig.1)

(2) Preferred surgical technique:

For many surgeons today, the preferred approach is to go for a technique which preserves and re-centres the capsular bag, allowing in-the-bag implantation and fixation of the bag to the ciliary sulcus using a combination of a CTR and a CTS. (Fig.3 a - c)

The first thing to note is that the work up to surgery in such a case has to be meticulous, involving liaison with all the physicians involved in the patients care and the anaesthetist, so that a safe general anaesthetic can be administered.

Biometry presents some unique challenges in patients with Marfan’s syndrome. One of the most important factors to consider when doing the biometry is which section the axial length measurement is taken through, i.e. the phakic or aphakic portion and then adjusting your instrument accordingly, to take into account the presence or absence of lens material. Depending on the extent of the subluxation you may need to consider dilating the patient in order to ensure you are definitely aiming the beam through the correct portion. If using the A-Scan ultrasound to measure axial length, you will be able to confirm the position of the probe by looking for the echo corresponding to the presence of the lens boundaries. Using a case of Marfan’s as an illustrative example: A 13 year old had bilateral subluxed lenses had an undilated refraction of R:-30.00/+6.00x115 L:-24.00/+5.00x60. When diluted and using his aphakic portion his refraction was R:+8.00/+3.50x120 L:+9.00/+2.50x80 achieving the same level of vision with both prescriptions. His phakic axial length was RE:26.49, LE:26.01 and aphakic axial length RE:26.36, LE:25.96. As you would expect the measurement should be comparable regardless of which portion it’s taken through, if the instrument is correctly programmed. The left eye was operated on using this measurement and has a deviated post-operative spherical equivalent of -0.68DS.

The lens chosen should be a 3 piece lens with a 360 degree square edge, This reduces the risk of post-operative posterior capsule opacification and the 3 piece designs allows for resturning into the scleral sulcus, should late breakage of the suture lead to IOL displacement in future years. I prefer a B&L (Bausch & Lomb) L161AO which is a 3 piece lens with a 360 degree square edge and an aspheric design. The Sofport injection system, used for injecting this lens allows it to come out into the capsular bag in a fairly flat disposition, minimising the potential for destabilising an already unstable bag during IOL implantation.

An illustrative case described below is used to demonstrate the steps in the technique. Surgery is usually done under a general anaesthetic.

(3) Case Presentation

A 12 year old boy with known Marfan’s syndrome was referred by a paediatric ophthalmologist who had been seeing him from the age of two years. He had managed reasonably with spectacle correction, maintaining good vision in both eyes. More recently the subluxation in the right eye had progressed to the point that his vision was reduced to 6/24 (BCVA). On presentation his vision was 6/24 in the right eye (-24.50 DS / +9.00 DC x 92 ½ and 6/9+3 in the left eye (-3.00 DS / +3.00 DC x 95). In addition he was being bullied at school and children kept stealing his glasses. Systemic workup included aortic valve problems for which he was on Atenolol and antenatal hydronephrosis both of which were being managed by appropriate specialists. On examination the right lens was subluxed markedly with edge of the lens passing through the centre of the pupil and the left lens subluxed to a lesser extent. Rest of the eye examination was unremarkable with normal intraocular pressures, no vitreous in the anterior chamber and normal peripheral retinal examination.

Axial length measurements through the phakic portion was RE: 25.50, LE:21.46 and through the aphakic portion RE:25.29, LE:21.25. Using the aphakic measurement we calculated a +18.00 Diopter B&L (Bausch & Lomb) L161AO IOL for emmetropia when implanted into the capsular bag.

Figure 4a shows the pre-operative situation, with a subluxed and decentred 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 towards the fornix. The scleral dissection begins at the limbus and extends peripherally for about 3mm. It is 4 mm wide (Fig 4b). Two paracentecis incisions are made on either side of this scleral flap and flexible ‘iris’ retractors are prepositioned to support the capsulorrhexis as it is later developed (Fig. 4c).

The capsulorrhexis is begun close to the iris edge at 12 o’clock as this is the centre of the lens (Fig 5a). Viscoat is used to fill the anterior chamber and tamponade the vitreous face. The capsulorrhexis is developed, taking care to keep a 3 mm distance from the lens equator (this facilitates later placement of CTR and CTS; an adequate anterior capsule is necessary to avoid the CTR or CTS flipping forward, out of the bag and into the anterior chamber later in the operation.). The tear is developed a few mm beyond the area where the iris retractor is poised to engage it, and once the tear is well clear, the ‘iris retractor is used to engage the edge of the rhesis and gently draw the lens equator out to the periphery thus helping to re-centre the capsular bag. The rhesis is developed further and the second iris hook engaged and drawn centripetally further re-centres the capsular bag allowing the superior part of the capsule to come into view (Fig 5b). The rhesis can then be completed. Gentle hydrodissection, with small slow waves of fluid is then done.

A CTR is then inserted into the bag to stretch it (Fig 6a), and thus allow phacoemulsification without the bag flopping onto the probe, as is possible in a largely unsupported bag. Once lens matter has been removed a CTS with a 9-0 prolene on a double armed Ethicon CIF-4 needle passed through the central islet is guided into the capsular bag (Fig 6b). The IOL is then implanted into the capsular bag (Fig 6c) and the needles are passed in front of the anterior capsule but behind the iris to emerge about 1.5 mm behind the limbus under the previously designed scleral flap. The iris retractors are then released allowing the surgeon to judge the position of the capsular bag better. The suture is tightened (Fig 7) and incisions sutured after removing viscoelastic. Intracameral Cefuroxime was injected at the end for endophthalmitis prophylaxis.

Two weeks post operatively vision had recovered to 6/18 unaided, 6/12 with a pinhole and at six weeks
vision was 6/6 with minimal refraction (-1.00 DS / +1.25 DC x 180). The IOL was well centred. He requested surgery for his other eye.

(4) Surgical pearls

i) Unstability of the anterior chamber must be avoided at all cost, mainly to avoid vitreous coming forward. Meticulous fluid – viscoelastic exchange technique avoids the capsular bag diaphragm flopping forward and backward. This means that every time the phaco probe has to be removed from the eye, this is done by stopping aspiration, but with irrigation continuing, viscoelastic is injected through a side port to support the anterior chamber. Whilst viscoelastic is being injected, one gradually eases off the irrigation, and then the probe can be safely withdrawn whilst maintaining a deep anterior chamber, and preventing the capsular bag from coming forward.

ii) Instead of conjunctival dissection and a scleral flap, a 600 micron limbal incision is made with a guarded diamond blade in the region of maximal lens edge decentration. This incision is similar to a limbal relaxing incision. A crescent blade is then used to dissect a 4 mm (wide) x 3 mm deep (from limbus to periphery), partial thickness scleral pocket. This allows the prolene knot to be buried under sclera, with conjunctiva undisturbed. Moreover as the sides of the 'pocket' are attached, there is no real risk of scleral 'flap' contraction in the future.

iii) When the lens matter is soft (as in a young patient), most of the surgery is done through two paracentesis incisions (about 1 mm each), with all the lens matter removed using bimanual irrigation-aspiration (no phacoemulsification). This allows for greater stability and reduces risk of vitreous disturbance. Only when all lens matter has been removed a 3 mm incision is made to enable placement of a CTS and then the IOL.

Conclusion

I have here presented the evolving techniques for lens surgery in Marfan’s syndrome. Whilst 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. Meticulous surgical technique is essential to avoid disrupting the capsular bag and disturbing the anterior vitreous face. When this is done excellent visual and anatomic outcomes are attained. At least potentially, this should reduce the risk of long term complications, time will tell if this potential is achieved. In a personal series of eleven consecutive Marfan’s eyes operated on over the last two years, in the bag placement with a well centred bag and secure fixation of a capsular tension segment using a prolene suture was achieved in all cases with good visual outcomes.

Reference:

In a lighter vein

MEMORY

RRV

I had previously written in this column that the most essential quality for a medical practitioner is patience. It is true, alright. But equally important is another one - good memory for names and faces. At least some of your patients expect you to remember them when they come back to you - six years after their previous visit. And are very gratified if you do. Some of them even feel insulted if you don't.

In my school days we had our family doctor who used to manage anything from allergy to angina and pruritis to pregnancy. Every time one of the family members went to him, he used to ask about the others; their marriages; children's educational problems etc. And he never made a mistake. I always used to wonder how he managed to remember each one of his patients with their myriads of profiles and problems. Of course he had a good memory. But when I myself became a doctor, I happened to ask him as to his prodigious reminiscental capabilities he laughed and said: "Oh! I make small notations on the edge of the prescription as to the job or class or wife's name etc."

I tried to emulate him when I started practicing. But my memory was never as good; and I couldn't remember what my notations stood for.

The problem is worst when you think you remember the face but can't put a name to it. And the patient talks on familiarly and has not brought the old prescription. When the time comes to write the prescription you use various techniques. One is to ask him/her what his/her FULL name is. If it is such small and simple name like "Unni", you are in trouble. Or you ask him/her what the OFFICIAL name is. If the answer is 'It is the same one', again you are in trouble. May be you can ask what his/her initials are. Most of them will say the name along with the initials. Again it won't help if he/she answers simply "P.K." or "C.T." With ladies I try another ruse. I ask what their husband's name is and prefix a 'Mrs.'; but you are in a fix if she demurely say "Oh, doctor, but I am not married." So now a days I take the bull by the horns and say: "Sorry, but I can't remember your name". With young boys or girls I may add, "How you have grown up! I couldn't recognise you". Do not try this with older women Instead, say: "You appear younger, I couldn't recognise you." Fortunately in my experience, men do not mind being asked the name visit after visit as much as the fairer sex.

A couple of days back I asked a female patient her name and heard the familiar 'how can you forget me' thing. But considering that she was clad in a 'burkha' with a cloth mesh in front of her eyes, I felt vindicated.