Management of Subluxated Lenses

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Introduction

Subluxation of lens signifies partial displacement of crystalline lens or cataractous lens from its central position in the pupillary area.

Fig. 1. A subluxated clear lens

Etiology

Subluxation of lens may be congenital or acquired. Congenital subluxation of lens can occur as an isolated anomaly or associated with heritable disorders. The acquired subluxation of lens may occur due to hypermaturity of cataract, trauma, stretching, pull and degeneration of the zonule. 

Signs of Zonular damage should be sought in every patient especially if there is history of ocular trauma or pseudo-exfoliation (PXF).

- Systemic abnormalities associated with zonular weakness (Marfan’s, Weil Marchesani, Homocysteinuria) may not always be present. Hence a loose cataract may be overlooked with severe surgical consequences. Waneela MV Margress et al describe 5 subtle signs of zonular damage that should be looked for as a warning sign. These includes
  1. Visibility of the lens equator during eccentric gaze.
  2. Decentered nucleus in primary position
  3. Iridolenticular gap.
  5. Focal iridodonesis.

Obvious signs of subluxation are less easy to miss and includes phacodonesis, vitreous prolapse, iridodonesis, and lens subluxation.

Fig. 2. Anterior segment images demonstrating various degrees of subluxation of the crystalline lens

Even without development of cataract, subluxation of the crystalline lenses can induce significant visual symptoms such as large refractive errors, anisometropia, or amlyopia in a child undergoing visual development. Historically surgical treatment for subluxated lens was undertaken with great caution.
because of attendant complications and poor visual outcome.

Surgical management was limited to Surgical iridectomies, Laser iridectomy, discission, or ICCE

Patients who underwent ICCE were either left aphakic or prescribed aphakic correction, or CL or were advised Epikeratophakia. An ACIOL in an older patient was another option that was considered. These patients ended up with graft rejection, retinal detachment, glaucoma and gross visual loss.

Advances that have been made in the surgical management of patients with weak / missing zonules are SICS, Pars plana lensectomy and aphakic CL wear or 2nd ACIOL implantation, suturing PCIOL to ciliary sulcus / posterior aspect of iris, introduction of CTR (capsular tension rings), MCTRs (modified capsular tension rings), CTSs (capsular tension segments) and the possibility of small incision phacoemulsification with in the bag IOL implantation.

Management

Timing of surgery is critical and is governed by the amount of subluxation. Children who are in their visually formative years, need early surgery if they have a large amount of subluxation. Early surgery and visual rehabilitation prevents development of amblyopia or permits early initiation of amblyopia therapy. In children with minimal amounts of subluxation that is compatible with normal visual development can be followed up without surgical intervention.

An initial assessment of the BCVA for distance and near should be performed. Several attempts at refracting the child is necessary before deciding whether he is seeing best with the phakic or aphakic correction. If the subluxation is not large and there is no eminent danger of the lens dislocating posteriorly or anteriorly observation with institution of amlyopia therapy is all that is necessary.

If amplyopia cannot be effectively treated by conventional means such as glasses, contact lens, or patching, surgical treatment is advisable.

Surgical treatment is also advisable if there is
1. Progressive subluxation.
2. Lens bisects pupil
3. Threatened posterior or anterior dislocation
4. Any case of poor visual acuity in an older child or adult attributable to subluxated lens.

Preoperative evaluation

Comprehensive preoperative examination is necessary to increase the chances of surgical success.

a) The surgeon should make a note of the ‘area of zonular weakness’ by drawing it. He should also
i. Characterize the areas of zonular weakness in terms of degree of involvement.
ii. Location of the defect.
iii. Presence / absence of vitreous prolapse.
iv. Presence / absence of phacodonesis: Phacodonesis is most dramatic prior to pupillary dilation, as dilation often stabilizes the CB and iris, dampening any iris - lens movement.

A surgeon should be wary of inferiorly subluxated lens as such sublaxation is often indicative of 360° of very significant zonular damage combined with the effect of gravity. When the patient is made to lie down the lens will fall back posteriorly. In this situation it is unlikely that the surgeon will be able to remove the lens while preserving the capsular bag for PC IOL support. PPL should be considered in these cases.

b) Gonioscopy should be performed in older children to assess for angle recession, synchia etc if ACIOL implant is considered.

Patients should be counseled with regards to a sutured PCIOL / or a CTR.

c) The presence of comorbid conditions affecting visual outcome should be assessed.
d) Evaluation by internists to rule out systemic associations is also necessary.
e) Discontinuing oral anticoagulants as most of these patients have also cardiovascular diseases and may be on anticoagulant therapy.

A thorough ocular examination including a cycloplegic refraction, slit-lamp examination and detailed fundus evaluation should be done to assess the extent of subluxation and to plan the treatment approach. The
presence of iridodonesis, phacodonesis, lens edge and visible zonules, and relatively deep or an irregularly deep anterior chamber should be noted. If the lens is clear, look for irregular red reflex, displacement of “Y” sutures, and high refractive error. Intraocular pressure recording (IOP) and gonioscopy must be carried out.

Systemic examination is important. A thorough family history, complete cardiovascular and musculoskeletal evaluation may be needed in Marfan’s syndrome. In doubtful cases sodium nitroprusside test for homocystinuria must be done before subjecting the patient for general anesthesia.

Management of Clear Subluxated Lens

Conservative Approach

A minimally subluxated crystalline lens requires only observation and periodic follow-up. If the subluxation causes visual disturbances due to induced astigmatism or myopia, the management includes a cycloplegic refraction and subjective verification with prescription of full correction. Examination of undilated, aphakic and phakic portions of the pupil should be done to ascertain whether the patient has got unilateral diplopia or confusion. Appropriate spectacle correction with aphakic glasses, contact lenses or prisms is provided which gives better visual improvement than phakic correction. Argon or Nd:YAG laser iridoplasty can be tried to enlarge the aphakic portion of pupil.

Management of Subluxated Cataract

Lens extraction via a small incision PE and PCIOL implantation should be attempted in every case and the basic surgical principles are described below.

Surgical principles to be understood include

1. Incision should be placed away from area of zonular weakness to help reduce stress on the existing zonules during PE. Unfortunately majority of the patients have generalized zonular weakness. In this situation the surgeon should place the incision in the gradient opposite to the zone of maximum zonular weakness. However, the surgeon should not jeopardize his surgical ability by operating in a meridian he is uncomfortable with.
2. Surgeon should work through the smallest incision possible without compromising the ability to perform necessary maneuvers. This will minimize fluid egress through the incision and prevent anterior chamber collapse. The initial AC entry should be just large enough to introduce a visco cannula.
3. A generous amount of highly retentive viscoelastic is placed over the area of zonular dialysis to help tamponade the vitreous and to maintain a deep non collapsing AC.
4. The capsulorhexis is started in an area remote from the dialysis to help utilize the counter acting forces of the remaining healthy zonules.
5. A second instrument is used for counter traction or to push the lens into view if it is significantly decentered under the iris.
6. When there is extensive zonular loss or weakness it may be a good strategy to start the rhexis by cutting the anterior capsule with a sharp tipped blade.
7. A rhexis of 5.5 mm – 6mm will facilitate all manipulations of the nucleus.
8. Hydrodissection: should be performed carefully yet thoroughly to maximally free the nucleus thereby decreasing zonular stress while manipulating the nucleus.
9. A soft nucleus can be completely prolapsed into the anterior chamber to simplify removal and virtually eliminate all zonular stress.
10. Phacoemulsification should be performed using low vacuum and aspiration settings in order to keep the bottle height at a minimum, a technique known as ‘slow-motion Phaco developed by Robert Osher.
11. Bottle height: it is important to keep the bottle at an optimum height, neither too high nor too low.
   a. Very high bottle height can in turn force fluid through weak areas of the zonules hydrating the vitreous resulting in positive pressure, anterior chamber shallowing and vitreous prolapse.
   b. Too low bottle height can result in an out flow, which is greater than inflow again resulting in
shallowing of anterior chamber, a negative pressure in AC and further vitreous prolapse as the anterior segment is less pressurised than the posterior segment.

12. Divide and or chop technique are preferred in eyes with zonular weakness. This technique minimises zonular stress during phacoemulsification if surgeon is careful to apply equal forces in opposing directions to avoid displacing the nucleus.

13. “Visco dissect” nuclear halves / quadrants in areas of zonular weakness. The viscoelastic should be injected below the nuclear fragment and the capsular bag-lifting the nuclear fragment as well as expanding and stabilizing the capsular bag. Additional cortical removal by visco dissection will limit stress on the remaining zonules during aspiration of cortex.

14. Automated Irrigation and Aspiration device is not preferred for cortex removal as it can hydrate vitreous and increase vit prolapse. Manually aspirate with a 24/27 G canula striping cortex in a tangential manner instead of radially to limit stress on zonules. A J’ cannula can be used for sub incisional cortex. Ensure removal of all vitreous from the anterior chamber if it is present. Use ‘Dry vitrectomy’ with automated vitrector after filling anterior chamber with viscoelastics. For significant vitreous loss a bimanual vitrectomy should be performed.

**IOL placement options**

1. The surgeon should decide if it is safe to use an ACIOL or PCIOL.

2. If an ACIOL is used the remnants of the capsular bag should be removed to prevent contraction and opacification.

3. If the surgeon uses a PCIOL it should be either
   a. Sutured to the scleral wall or
   b. Placed in the capsular bag

Ciliary sulcus placement of PCIOL without suture fixation in an eye with significant zonular compromise is not recommended.

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**Placement of PCIOL into the capsular bag**

1. Placement of PCIOL into the capsular bag is challenging when there is significant zonular weakness as one must achieve IOL centration, and maximize long term stability.

2. Use of 6 mm optic diameter IOL decreases the chances of undesirable edge-glare symptoms should lens decentration occur post operatively. Haptic configuration designed for broad contact with equatorial capsular bag increases the chances of long term centration. Use of silicone plate haptic IOL should be avoided in the presence of zonular dialysis as there is greater chance of capsular contraction and decentration.

3. Insertion of CTR to provide 360° capsular bag expansion and greater stabilization.

4. If the ZD is located at the incision site, lens placement is more difficult.
   a. One Option is to first place the entire lens into the AC. Then using a two handed technique, the superior haptic is inserted into the capsular bag followed by a similar maneuver for the inferior haptic.

5. Orientation of the IOL: There are 2 schools of thought.
   a. Orienting the IOL in a plane parallel to the zonular dialysis (ZD) in order to take advantage of the remaining intact zonules. This orientation will provide optimum support but may induce ovaling of the capsular bag and an increased risk of postoperative decentration.
   b. Placing one haptic in area of ZD will ensure stretching of the bag and decrease ovaling. However it should be borne in mind that only one haptic is adequately supported.

It is recommended to orient the haptics in whichever axis that provides the best centration intraoperatively. This is accomplished by careful rotation of PCIOL.

**Capsular Tension Ring (CTRs)**

- Drs Witschel and Legler (1993) from Germany demonstrated that CTRs could provide both intraoperative and postoperative stabilization of
the capsular bag. Produced by Morcher GmbH, in Stuttgart, Germany and made of polymethyl methacylate (PMMA), this ring can be inserted into capsular bag at any point after a continuous curvilinear capsulorhexis has been completed.

- Use of CTR is contra indicated if a CCC is not attained or if a posterior capsular rent occurs.
- In eyes with profound zonular compromise or lens subluxation may not achieve adequate stabilization or centration despite CTR placement.

Long term stability even in presence of CTR is doubtful in eyes with progressive zonular weakness such as Marfans, PXF etc. Phacoemulsification with the proper use of endocapsular device can give excellent results in patients with subluxated cataracts

![CTR Image](image1.png)

**Step 1:** Injection of VE under the surface of the residual anterior capsular ring to create a path for the CTR. Viscoelastic agents dissect peripheral cortex away from the capsular fornices and make up cortex entrapment by CTR less likely.

**Step 2:**
- a) CTR is introduced either using a smooth forceps through the main wound or a paracentesis or by
- b) Using a CTR injector / CTR shooter (Geuder)

**Step 3:** In eyes where CTR is introduced prior to phacoemulsification a safety suture is looped through the leading eyelet and is allowed to trial out of the eye at the main incision during PE and cortical aspiration. This guiding suture is used to retrieve the CTR from the eye should a posterior capsular rent occur during phacoemulsification or cortex removal. If the procedure is uneventful the suture is cut and removed.

**Standard capsular tension ring**

Credit goes to Hara and Yamada\textsuperscript{12} for introducing endocapsular ring in the year 1991 for maintenance of the circular diameter of capsular bag. Later the device was further refined and modified for managing severe degree of subluxated lenses\textsuperscript{12-16}.

The standard CTR in made of polymethyl methacrylate (PMMA) material and has an oval shaped cross section with eyelets at both free ends (Fig.5). It is a compressible circular ring with two smooth edged end terminals.

CTR is manufactured by Morcher GmbH (Struttgarf, Germany) and Ophtek (Groningen, The Netherlands)
Fig. 6. Morcher CTRs

and is US-FDA approved. The Morcher ring, also known as Reform ring, comes in three different sizes based on the uncompressed diameter (Fig.6).

**Capsular ring size**

Selection of CTR size is based on capsular bag dimensions. A large capsular bag usually requires a larger ring; 13mm ring is being most commonly used. White to white corneal measurement and axial length can be used as a rough guide in the selection of CTR. It would be appropriate to use a larger CTR in adults with highly myopic eyes.

Step 4: Placement of IOL in the bag and has the CTR is no different than routine cases, infact it may be even easier as the bag is better supported.

Step 5: Suturing the CTR to Scleral wall for support.

a) Robert Osher has desired a technique of suturing the CTR to the scleral wall by straddling the CTR with 10° prolene double suture. This technique

although it works well involves risk of rupturing the capsular bag since it is under stretch due to presence of CTR.

Vladimic Pfizer’s technique involves needle passage thro a small peripheral capsulorrhexis. However the integrity of the capsular bag is violated and the risk of rupture is present.

In more severe subluxation, modified CTR, like Cionni’s M-CTR with one or two eyelets attached to the central ring is used and a posterior chamber IOL (PCIOL) placed in-the-bag.

Robert Cionni’s Modified CTR (MCTR) \(^8,9,10\) has an unique fixation hooklet designed for scleral fixation without violating the integrity of capsular bag. The fixation hook courses anteriorly and centrally in a second plane, wraps around the capsulorrhexis edge and rests on the residual anterior capsular rim.

**Modified capsular tension ring (M-CTR)**

The standard CTR is unable to provide adequate intraoperative support and centration of the bag in

**Modified capsular tension ring (M-CTR)**

The standard CTR is unable to provide adequate intraoperative support and centration of the bag in
grossly subluxated cataracts or lenses. Cionni\textsuperscript{5} developed the modified CTR (M-CTR) (Morcher – GmbH) called Cionni ring in the year 1998. This ring provides a solution to extensive zonular deficiency or damage or progressive zonular damage by allowing the surgeon to anchor the capsular bag to the sclera.

3 modules are currently available.

Model 1 L : Single fixation hook distant from insertion end of the ring.

Model 2 C: Single fixation hook near insertion end of the ring.

Model 2 L: has 2 fixation hooks which is very useful in patients with significant zonular weakness and may be the ring of choice in patients with progressive zonular weakness as in Marfan’s Syndrome.

The Cionni’s modified ring has an open ring design with one (model I-L or I-R) or two (model 2-L) fixation eyelets attached to the central ring (Fig 7). The eyelet allows the ring to be sutured to the sclera. It protrudes 0.25mm forward from the body of the ring and then sits anterior to the anterior capsular rim and allows maintenance of capsular bag integrity on suturing to the sclera. A 9.0 prolene is preferred over 10.0 prolene as the incidence of breakage is less with the former. An adequately sized rhexis is essential to prevent iris chaffing, pigment dispersion and chronic uveitis.

An eccentric rhexis has to be performed in order to be certain that after the bag is recentred, the capsulorhexis opening is recentered as well.

![Capsular tension segment](image)

**Capsular tension segment**

Capsular tension segment (CTS) is a partial ring of 90-120 circumference, and is made of PMMA (Fig. 8 & 9). It has a radius of 5mm and an anteriorly positioned fixation eyelet like M-CTR. This was designed by Ahmed \textsuperscript{17}, and was manufactured by Morcher GmbH. CTS is useful for cases with profound zonular insufficiency. The CTS provides support in the transverse plane, when sutured to the scleral wall. When circumferential support is needed, a CTR may be implanted in conjunction with an already positioned CTS. The CTS in available in 3 sizes having radius of curvature of 4.5 mm, 5 mm and 5.5 mm.

The choice of endocapsular support device depends mainly on the nature of zonular weakness, degree of zonular loss, and the extent of zonular instability. CTRs are indicated in cases of mild, generalized zonular weakness or in small localized zonular dialysis of less than 3-4 clock hours. In more advanced or progressive

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**Table 1: Indications of capsular tension ring, modified-capsular tension ring and capsular tension segment**

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<thead>
<tr>
<th>Requirement</th>
<th>CTR</th>
<th>M-CTR</th>
<th>CTS</th>
</tr>
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<tbody>
<tr>
<td>Requires continuous curvilinear capsulorhexis</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
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<tr>
<td>May be placed prior to lens removal</td>
<td>With difficulty</td>
<td>With difficulty</td>
<td>Yes</td>
</tr>
<tr>
<td>Use with anterior capsule tear</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>Use with posterior capsule rent</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
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<tr>
<td>Use with large zonular dialysis (more than 8 clock hours)</td>
<td>No</td>
<td>Yes</td>
<td>Yes (multiple segments)</td>
</tr>
<tr>
<td>Use in progressive zonulysis</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Allow for suture fixation to sclera</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>May be easily removed from eye if needed</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>Cortical removal difficulty</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
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CTR: capsular tension ring, M-CTR: modified-capsular tension ring, CTS: capsular tension segment
cases of zonular instability, the Cionni M-CTR or the CTS is indicated. Indications of CTR, M-CTR and CTS are given in Table 1.

Closed chamber endocapsular phacoemulsification combined with ECR, in patients with mild to moderate subluxation of lens not associated with complications such as secondary glaucoma and retinal detachment, often gives encouraging visual results. The implantation of ECR has provided safety and efficiency during phacoemulsification and IOL implantation, significantly reduced the rate of complications and overall improved visual results.

References