

Surgical technique for the management of corneal perforation in brittle cornea

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Brittle cornea syndrome is among the few special scenarios in ophthalmology that are a nightmare not only for the operating surgeon but also for the patient. Here, the thin and fragile corneas are unable to maintain the shape and structural integrity of the globe and are more prone to minor traumatic or spontaneous corneal perforations. Suturing a brittle cornea and closure of the corneal perforation in a brittle cornea are very challenging requiring the utmost care and special precautions. If proper measures are not taken during the surgery, it may be difficult to salvage the eye. Hence, it is imperative to diagnose appropriately, suture effectively, taking necessary preventive measures in salvaging these corneas. This manuscript aims at providing tips for handling brittle corneal perforations. It will also discuss the problems encountered during surgery, highlight the suturing techniques that can be customized, and finally give an insight into postoperative care.

Key words: Blue sclera, brittle cornea syndrome, corneal laceration, corneal perforation, corneal tear, suturing technique

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Brittle cornea syndrome (BCS) is not only a diagnostic challenge^[1] for any ophthalmologist but also a difficult condition to manage.^[2] BCS, although underdiagnosed, is a rare, connective tissue disorder with characteristic ocular features like fragile cornea with extreme corneal thinning, blue sclera, high myopia, and retinal detachment.^[2] It can be associated with musculoskeletal abnormalities like developmental dysplasia of the hip, clinodactyly, hallux valgus, and hearing abnormalities.^[2]

As the name suggests, the cornea is very brittle in BCS that may spontaneously rupture under normal conditions. Repairing such perforations of extremely fragile corneas can be a nightmare, sometimes, requiring a penetrating keratoplasty.^[3-5] In fact, at times, it may become very difficult to salvage the eye^[2] and anything less than a perfect job would result in disfigurement with permanent loss of vision. Since suturing tears or lacerations in brittle corneas are technically difficult with no clear-cut guidelines or protocol on how to go about, this manuscript aims at describing the surgical steps with proper suturing technique to achieve the best surgical outcome.

Surgical Technique

Closure of corneal perforation in a brittle cornea (BC) requires utmost care and special precautions. Most of the times, these perforations can be very irregular in shape and

are, therefore, difficult to secure. The operating surgeon should be extremely patient during the entire procedure. Operating slowly as in a “slow motion” will keep the surgery entirely under ones’ control like “taming the tear” and avoids extension of the existing tear. If proper measures are not taken, it may not only be difficult to close the wound but may also result in new uncontrolled and larger lacerations making it difficult to seal the tear and more importantly salvage the eye.

First of all, it is important to study the cornea. If the cornea is thin and fragile with butter-like consistency (no resistance encountered from the corneal tissues), or the sclera is blue with or without the features of keratoglobus, and any attempt holding the cornea leads to cheese wiring, expect it to be a BC, and be gentle and meticulous while handling.

Type of anesthesia: In BCS, the corneal tear or laceration may not only be large and irregular but also can extend on the operating table. It may even spread to the sclera due to the poor scleral rigidity. Rarely, as previously reported,^[4] as the surgical plan may change from a simple repair of laceration to a corneal transplantation, hence better to operate these cases under general anesthesia (GA).

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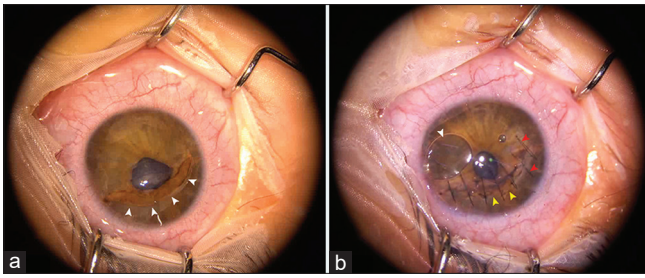


Figure 1: (a): Preoperative microscopic picture showing paracentral circumferential full thickness corneal tear (white arrowheads) with iris incarceration in the wound. (b): Postoperative microscopic picture showing well apposed corneal tear with *in situ* long sutures (red arrowheads) with air bubble (white arrowheads) in anterior chamber. Yellow arrowhead represents the overlapping zone of compression between the sutures

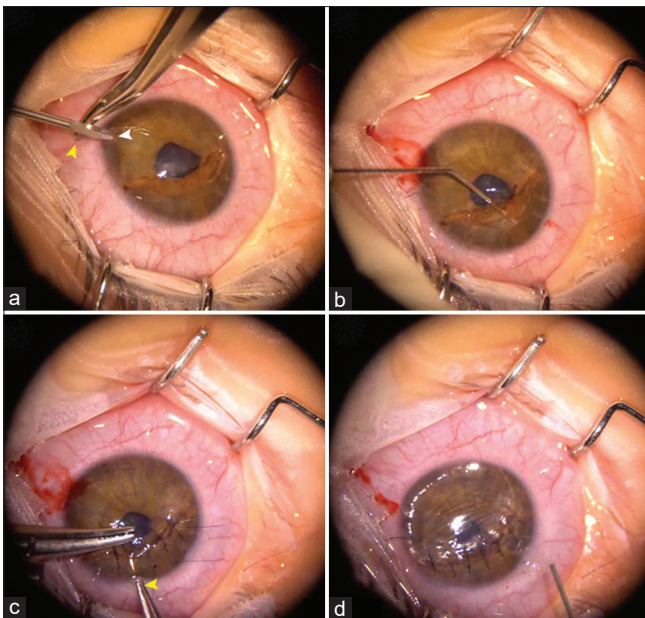


Figure 2: (a): Intraoperative microscopic picture showing guarded vertical entry into anterior chamber (white arrowhead) with support from same side of limbus (yellow arrowhead). (b): Intraoperative microscopic picture showing gentle release of iris incarceration from the wound using saline cannula. (c): Intraoperative microscopic picture showing needle passage through the lacerated wound with support from limbus (yellow arrowhead). (d): Postoperative microscopic picture showing well apposed corneal tear with *in situ* long sutures with well-formed anterior chamber containing air bubble

In BCS, the principles of suturing a normal corneal laceration need to be modified with the following considerations: We have described the principles of suturing a brittle corneal laceration in a five year old child, who presented to the Cornea and Anterior segment services as an emergency with a corneal tear in her right eye [Fig. 1a] following eye rubbing.

A guarded speculum should be avoided to prevent undue pressure on the globe. Entry into the anterior chamber (AC) [Video Clip 1] has to be guarded and slow. While making the entry, support should be taken from the same side of limbus and a vertical entry should be planned [Fig. 2a]. As the globe is hypotonus, this entry will result in

a regular corneal tunnel type wound as shown in the video. Size of this side port must be adequate and not too tight as it is not uncommon to have an irregular extension of side port following repeated handling and manipulation through a tight wound. In case of iris incarceration into the wound, the iris tissue should be released very gently from the wound using, either saline [Fig. 2b], viscoelastic substance or an iris retractor [Video Clip 2]. Remember to avoid holding the cornea while releasing the iris as it may result in the extension of the tear. In addition, the entire iris tissue should not be released in one go but part by part as the suturing is continued along the wound. Injecting of viscoelastic [Video Clip 3] during the suturing should be done in a controlled manner. Injecting too much viscoelastic substance can result in sudden increase in the intraocular pressure, thereby, propagating the extent of tear.

While suturing the cornea, suture bites should be longer and deeper [Video Clip 4]. The needle should be passed in a single go and avoid multiple entries [Fig. 2c]. Grasping the cornea during its passage should be avoided. Needle should be pulled out by applying counter pressure without holding the edge of the wound. Tension on the suture should be adequate enough to oppose the wound rather than compressing it too much. Tight sutures should be avoided; instead, multiple sutures should be taken. Sutures should be longer to avoid weak wound areas and closely placed together maintaining a significant overlap of the compression zone such that tight suturing is prevented, which in turn can lead to cheese wiring. This is unlike the suturing in normal cornea wherein smaller bites are preferred towards the visual axis with slight overlap of compression zone. The burial of knots is an equally important step [Video Clip 5] which has to be done carefully as it may not only result in suture track leakage but also in the tear extension in the worst case. The longer suture bites ensure that knots can be safely buried away from corneal tear wound. The knot needs to be small and compact and can be crushed to further reduce its profile before burial. Coating the knot with the viscoelastic substance facilitates easy passage during knot burial. Anterior chamber (AC) formation [Video Clip 6] is the final as well as an important procedure [Fig. 2d]. Air is preferred along with saline in forming the AC. Many times the saline may leak out if the wound is not very water tight and repeated filling of the AC with saline may increase the chances of side port tear. In addition, air tamponade can act as an adjunct in the application of fibrin glue that can be used, if the wound is not water tight. Note that after formation of the AC, there is a possibility of a rise in the intraocular pressure, which may again result in cheese wiring of the cornea if sutures are placed too tightly. It is, therefore, important to fill the AC adequately enough with air to ensure proper opposition of the corneal lips without any ocular hypertension or hypotony. Fig. 1b is the final appearance of the eye after suturing.

Discussion

BCS is a rare multisystem connective tissue disorder characterized by generalized corneal thinning resulting in a fragile cornea, which is unable to maintain the structural integrity and shape under the normal biomechanical stress thus increasing the probability of ocular ruptures either spontaneously or after a minor trauma. Ocular features associated with BCS include high myopia, keratoconus, keratoglobus, and blue sclera. The associated extra-ocular

features include deafness, red hair, hyper elasticity of the skin without excessive fragility, and hyper mobility of the joints. It is an autosomal recessive condition that results from missense mutations in two genes, ZNF469 and PRDM5.^[6-8]

Previous studies in the literature reports have shown that the suturing of a brittle corneal laceration or tear is challenging and difficult.^[4-7,9] A case, reported by Izquierdo *et al.*,^[9] documented an intraoperative extension of the existing corneal tear in a case of brittle cornea during the burial of the knot warranting a scleral patch graft to seal the rupture site. Another case^[7] reported cheese wiring of the suture through a brittle cornea resulting in track leaks needing re-intervention in the immediate postoperative day. A similar case of a ruptured brittle cornea showed the inability to approximate the edges of the thin cornea and a tectonic sclera-corneal grafting was performed to restore anatomical integrity to salvage the eye.^[4] Thus, in short, suturing a brittle cornea is a nightmare.

Till now, there have been no clear-cut surgical guidelines to suture a brittle corneal tear or laceration. As the cornea is fragile and thinned out, the conventional corneal suturing technique described does not really help out in closure of these perforations as evidenced above.^[4,7,9]

Hence, while suturing the corneal ruptures in BC, modify your suturing by taking longer bites that are deeper than usual, support the wound rather than holding its edge while taking bite, take multiple sutures very close to each other, avoid tight sutures, and form the AC with air. In an event of wound leak at the end of surgery, one can always use cyanoacrylate or fibrin glue as an adjunct to ensure wound closure.

Postoperative care is equally important in these patients. Patient needs to be closely followed up and needs to wear lifelong protective polycarbonate glasses. Patients should be advised to avoid hand-eye contact, eye rubbing, and contact sports. Suture removal should be delayed as wound may not have healed and as there is a high chance of tear re-opening, suture removal has to be done in operation theater only. Be cautious and aware of fresh perforation that may be encountered during or after suture removal requiring closure.^[10]

As most BC would be associated with axial myopia, keratoconus, or keratoglobus and present at an early age,^[2,5] it is imperative to visually rehabilitate these patients early by providing spectacle prescription, thereby preventing the chances of amblyopia. Ensure that they are prescribed polycarbonate glasses to protect the eyes.

As trivial trauma can rupture the cornea in case of BCS, many times patients present with poor vision in the other eye. In such scenarios, the surgeon might be dealing with the only salvageable eye requiring the best efforts. The above-mentioned tips can help surgeons to save the day and achieve the best possible anatomical and visual outcome.

To the best of our knowledge and search in PubMed (brittle cornea syndrome, repair, surgery, technique), ours is the first publication providing clear-cut guidelines about surgical management of BC perforation. Thus, tackling the BC with the above described surgical technique can provide confidence to the surgeon handling such difficult cases. This article may benefit novice cornea surgeon and even inexperienced general ophthalmologists (who has not seen BC case before) to tackle such challenging special cases with ease.

Conclusion

Tackling a BC is challenging. If we correctly handle BCS rupture with patience, we will be able to achieve a well opposed corneal tear or a lacerated wound, thereby salvaging the patient's eye and eventually achieving good visual outcome.

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Conflicts of interest

There are no conflicts of interest.

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