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## Case Report

# Surgical Management of Keratoconus in a Young Adult with Cognitive and Behavioral Disability

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### ABSTRACT

**Case Presentation:** A 25-year-old man with cognitive impairment (CI) and behavioral issues presented for corneal collagen crosslinking (CXL) surgical management of progressive keratoconus.

**Results:** The conventional CXL technique was modified to avoid complications due to potential post-operative behavioral issues.

**Conclusions:** Significant post-operative pain and aggressive behaviour can occur following epi-on CXL. Aggressive behaviour can result in significant complications in relatively low-risk eye surgery. Pre-operatively, it is appropriate to devise a multifaceted surgical, medical and behavioural treatment plan including psychotropic if needed in patients with history of cognitive impairment and aggressive behaviour.

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### Introduction

Keratoconus (KC) is a noninflammatory progressive corneal degeneration that causes loss of stromal collagen fibers and reduced structural integrity. Pathologic changes to corneal architecture associated with KC lead to varying degrees of visual loss and pain. Eye rubbing may hasten the progression of KC. Corneal collagen crosslinking (CXL) is an effective procedure that slows the progression of KC. CXL promotes the formation of bonds between adjacent corneal collagen fibers that increase tissue strength and lessen pathologic alterations of stromal architecture. The conventional CXL procedure (i.e. epi-off CXL) is performed by delaminating the corneal epithelium, saturating the underlying stroma with riboflavin 0.1% solution and exposing the tissue to 370 nm ultraviolet light for 30 minutes [1].

The corneal epithelium typically heals within 5 days. A variation of the conventional CXL procedure is epi-on CXL which preserves intact corneal epithelium in order to reduce post-operative pain, inflammation, potential for scarring and risk of infection [2]. Some have reported satisfactory results using epi-on CXL whereas others suggest that epi-on is less effective due to differences in penetration of riboflavin into the

stroma [3-5]. Both epi-off and epi-on CXL techniques are commonly performed with topical anesthesia in an office setting. Cognitive impairment (CI) with or without behavioral issues may make the surgical management of KC more challenging. The purpose of the present case report is to describe CXL treatment using a multispecialty approach in managing the eye disease and behavioral issues in a young adult with KC.

### Case Report

This study was approved by the Institutional Review Board of Cincinnati Children's Hospital Medical Center and conformed to the requirements of the United States Health Insurance Portability and Accountability Act and the principles of the Declaration of Helsinki. A 25-year-old male with KC, CI, attention deficit hyperactivity disorder (ADHD), periventricular leukomalacia, obesity, Pierre-Robin sequence and obstructive sleep apnea was referred to our outpatient ophthalmology clinic for surgical CXL treatment. On examination, the patient had ophthalmic features of KC including Munson's sign, mild central corneal scarring, high astigmatism and inferior scissoring of the retinoscopic light reflex, bilaterally. Irregular astigmatism was 3.00 and

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4.00 diopters in the right and left eyes, respectively. Reliable digital corneal imaging was not possible due to limited patient cooperation.

The patient's history included episodes of aggressive or violent behavior during stressful events. Although the patient's scheduled medications included oxcarbazepine and lisdexamfetamine, exacerbations of increased agitation were typically managed with emotional reassurance and cognitive redirection by the patient's mother or other caregiver. The patient had a history of episodes of paradoxical increased agitation after receiving with lorazepam. The patient's mother indicated her strong aversion to the use of additional psychotropic medication in the management of her son's behavior.

The patient lived with his parents, ambulated well but had little speech capability and required assistance with daily hygiene and self-care. Bilateral corneal crosslinking was recommended to reduce potential KC progression and subsequent adverse effects on quality of life. The patient was a poor candidate for penetrating keratoplasty (i.e. corneal transplantation) due to his history of eye rubbing and behavioral issues.

Prior to surgery, a detailed multi-specialty treatment plan was developed utilizing input from the hospital-based general eye surgeon, fellowship-trained corneal surgeon, clinical research coordinator, research pharmacist, psychiatrist, psychologist, primary care physician, anesthesiologist and inpatient primary care hospitalist. Bilateral simultaneous epi-on CXL was performed under general anesthesia. Under an Investigational New Drug Exemption (IND) obtained for this study from the United States Food and Drug Administration (FDA), the riboflavin dosage was increased by 50% for the procedure to enhance penetration into the corneal stroma. After the CXL procedure, the patient emerged from general anesthesia with agitation and eye rubbing and developed a painful corneal abrasion of the right eye. The patient was hospitalized for 3 days for post-operative for pain and behavior management. The day after discharge, the patient returned to the emergency department with increased pain, eye rubbing and agitation. The patient's mother then agreed to one dose of olanzapine which provided good de-escalation of the patient's agitation. He was readmitted to the hospital and an examination under anesthesia revealed a persistent corneal abrasion without evidence of infection. He remained in the hospital for another 3 days for pain control and intervention by multiple services including ophthalmology, hospital medicine and behavioral medicine services.

On post-operative day 7 another examination under anesthesia was performed and persistent right corneal abrasion was present. A suture tarsorrhaphy was performed to promote healing of the cornea. However, the tarsorrhaphy led to escalation in the patient's agitation and violent behavior. Under the supervision of the behavioral medicine and hospital medicine services, 4-point physical restraints were temporarily required to manage the patient's violent behavior. Scheduled olanzapine was initiated despite the mother's initial refusal. Ziprasidone was also ordered for breakthrough agitation but was never needed after the initiation of scheduled olanzapine. One week later, the cornea was healed and the tarsorrhaphy removed at the bedside without difficulty. Olanzapine was continued long term resulting in fewer episodes of agitation and eye rubbing at home.

Progression of KC was detected in the right eye 1 year following surgery. Although epi-on CXL was repeated on the right eye, the KC continued to progress. Ultimately, the patient underwent epi-off CXL and tarsorrhaphy of the right eye after maximizing his dosage of oxcarbazepine and the addition of fluvoxamine. Although epi-off is typically considered more painful than epi-on CXL, the patient did not have agitation after epi-off CXL while complying with his enhanced psychotropic medication regimen. The patient was able to recover at home and demonstrated improved cooperation during subsequent post-operative office examinations. KC of both eyes remained stable 6 months after the last CXL procedure.

## Discussion

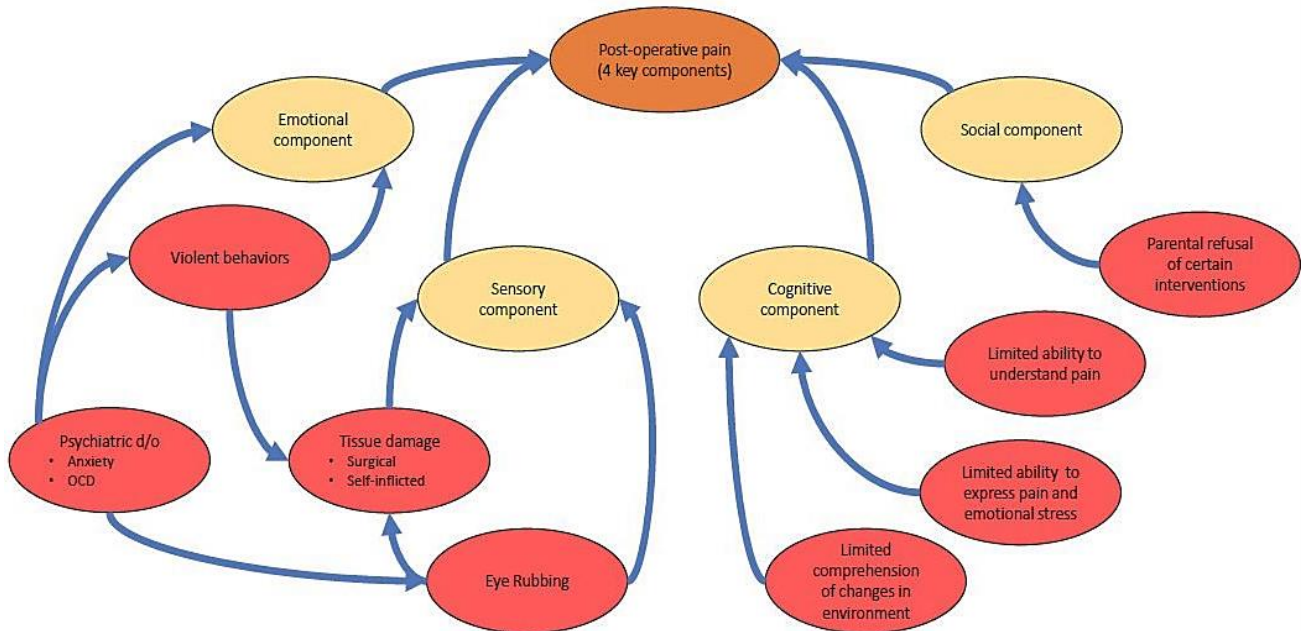
Corneal crosslinking has been shown to preserve vision and reduce the episodes of pain, inflammation, scarring and opacification caused by KC [1-5]. Providing surgical care including pre- and post-operative care can be more challenging in patients with CI and behavioral issues. We anticipated potential post-operative behavioral issues and took several steps to avoid complications. First, epi-on CXL was performed instead of epi-off in order to reduce the likelihood of post-operative pain, behavioral agitation and improper healing due to eye rubbing. Second, CXL was performed under general anesthesia instead of only topical anesthesia due to the patient's limited ability to cooperate with verbal instructions. Third, since intact corneal epithelium limits penetration of riboflavin, we increased the dose of riboflavin by 50%. Although one eye remained stable for 2 years following treatment, it is difficult to comment on efficacy of this modification based on this one case. Fourth, an inpatient treatment plan was devised pre-operatively with the relevant surgical, medical and behavioral health providers.

Our patient developed a corneal abrasion with the first epi-on procedure. Corneal abrasion causes varying degrees of pain which contributed to behavioral issues in this case. Pain can be defined as "a distressing experience associated with actual or potential tissue damage with sensory, emotional, cognitive, and social components [6]." Control of post-operative pain requires pain assessment and if indicated, intervention followed by reassessment and additional intervention if indicated. The most common pain assessment is the patient's self-reporting of pain which may not be reliable or possible in patients with CI. The pain assessment tool utilized by our institution for children and adults with CI is the Face-Legs-Activity-Cry-Consolability (FLACC) scale which includes direct visual observation of patient motor behaviors to determine pain level [7]. Another assessment tool, called the Non-Communicating Adult Pain Checklist (NCAPC), is a validated pain scale designed for adults with CI that assesses vocal reaction, emotional reaction, facial expression, body language, protective reaction, and physiological reaction to determine an overall pain score [8].

Using Brandow's model of pain management, one may delineate contributing pain factors observed in this case (Figure 1) [9]. Research is limited regarding pharmacological treatment of pain in individuals with intellectual disability [10]. However, the pain management medication choices should be dictated by the individual's symptoms and based on the same principles applied to those without intellectual disability [10]. Our multispecialty pain control approach involves surgeons, pain management service providers (anesthesiology), psychiatrists, psychologists and medical hospitalists to determine the

regimen of opioids and non-opioids for analgesia as well as other psychotropic medications and non-pharmacologic behavioral interventions that mitigate pain. Surgical and self-inflicted tissue damage (Figure 1) was managed with topical ocular antibiotic and steroid and later with tarsorrhaphy. Psychiatric disorders were managed initially

with the patient's usual oxcarbazepine (anti-epileptic with mood stabilizing properties) and lisdexamfetamine (stimulant for ADHD). Olanzapine (anti-psychotic) and fluvoxamine (selective serotonin reuptake inhibitor) were added later to further stabilize mood and reduce obsessive compulsive behaviors, respectively.



**Figure 1:** Post-operative pain model for adult with cognitive impairment and behavioral issues.

One alternative method has been described by Sisk *et al.* for the post-operative management process utilized for intraocular surgery patients with known history of severe self-inflicted ocular trauma [11]. Sisk *et al.* admitted patients after intraocular surgery to the intensive care unit to maintain deep pharmacologically sedation and intubated for 24 hours. The method provided proper head positioning, avoided early self-inflicted recurrent eye trauma and allowed for a complete funduscopic examination one day after surgery. Although our patient had a history of pre-operative eye rubbing, we did not employ this method since CXL is considered lower risk ocular surgery, does not require head positioning and does not require complete post-operative fundus examination.

## Conclusions

Epi-on CXL may cause significant pain that can be difficult to manage with patients with underlying behavioral issues. Behavioral issues can contribute to the development of complications. Pain is a multifaceted issue that may require analgesics and pharmacological management of behavior. A multispecialty approach is desirable in the pre-operative planning and post-operative management of surgical patients with behavioral issues. Although epi-off is typically believed to be more painful than epi-on, this adult patient with CI tolerated epi-off better than epi-on with less need for opioid pain medication after optimization of his psychotropic medications.

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