

Boston Keratoprosthesis Type I complicated by exposure secondary to Grave's Ophthalmopathy

Massachusetts



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Introduction

The Boston Keratoprosthesis Type 1 was initially introduced in 1974 by Dohlman et al. Since its FDA approval in 1992, it has been used as a full thickness corneal transplant in patients with multiple failed PKP and severe ocular surface disease secondary to SJS, OCP, aniridia, or chemical injury. The design of KPro Type 1 consists of a central PMMA optic/front plate connecting to a back plate. In between lies the donor corneal graft. The entire device is locked together with a titanium ring. The total length of the device is 3.7 mm, allowing for 60 degrees of visual field and is sutured into the recipient eye, similar to a PKP. Due to the irregular shape and material of the device, the donor graft is susceptible to desiccation, epithelial defect, thinning, and infection. A high Dk soft contact is used as a bandage over the device. Patients are put on long-term prenisolone acetate and vancomycin drops to prevent rejection and endophthalmitis, respectively. All of these patients require frequent follow up due to severe complications from the procedure, including, retinal detachment, RPM, infection, extrusion, and glaucoma.

History	
Demographics	73 yo Caucasian Female
Chief complaint	Progressively worsening vision OD beginning 5 days post-op for Boston Kpro I (7/28/17). Patient presented to the MEEI Cornea clinic on 9/11/17.
History of Present Illness	This patient has a longstanding history of Graves ophthalmopathy with progressive proptosis and exposure keratopathy. She presents with lateral tarsorrhaphy OU. She is reports having difficulty keeping her bandage contact lens in her right eye. Previously, the patient had been in so much pain OD that she had presented to her local ophthalmologist two Saturdays in a row and was ultimately given Oxycodone. The patient is s/p KPRO I OD 7/28/17 performed in CT. She has a h/o CME OD. She denies ever having a previous PKP OD. Ocular Medications: Genteal OU QID Durezol OD QID

Genteal gel OD qhs Patient Medical History

Hyperlipidemia, Grave's Disease S/p RAI 1994 S/p thyroidectomy 1994

Ofloxacin OD TID-QID

Social History

S/p orbital decompression OD 2014 Patient Ocular S/p lateral tarsorrhaphy OU 2015 History S/p CE/PCIOL OU 2016

Family Medical History

Unknown

Former Smoker (quit in 2000), denies alcohol or

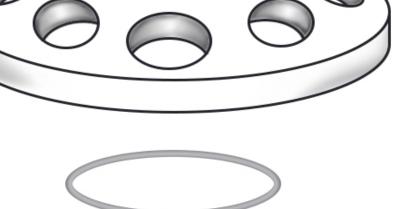
drug use

Front Part



Back Plate

Corneal Graft



Locking Ring

Above: Boston Keratoprosthesis Type I design

Exam \mathbf{OD} OS 20/500, PH 20/200 20/40, PH 20/20 Distance VA(sc) KPro Reactive, (-)APD Pupils SAFE **EOMs** SAFE

Visual Fields FCF FCF 10 (palpation) IOP 11 (tonopen) External evaluation **Proptosis Proptosis** Lateral tarsorrhaphy, 3 mm lagophthalmos

nasal, trichiasis, Kpro | Lateral tarsorrhaphy, I with 360 ring 3 mm lagophthalmos Anterior segment nasal, trichiasis, large infiltrate, area of epitheliopathy evaluation inflammatory and haze inferior ½ of membrane on cornea, PCIOL posterior optic with additional membrans

in AC, 1+ cell

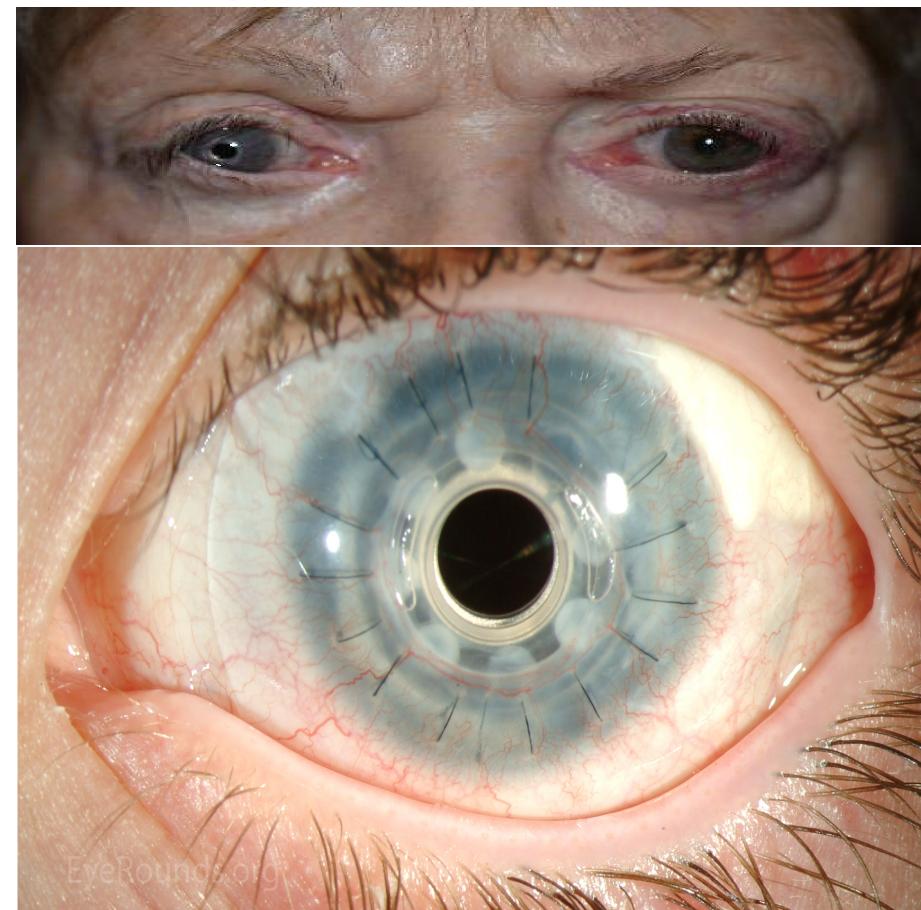
Hazy view with Posterior segment questionable blurred evaluation disc margins

Hazy view with normal disc margins

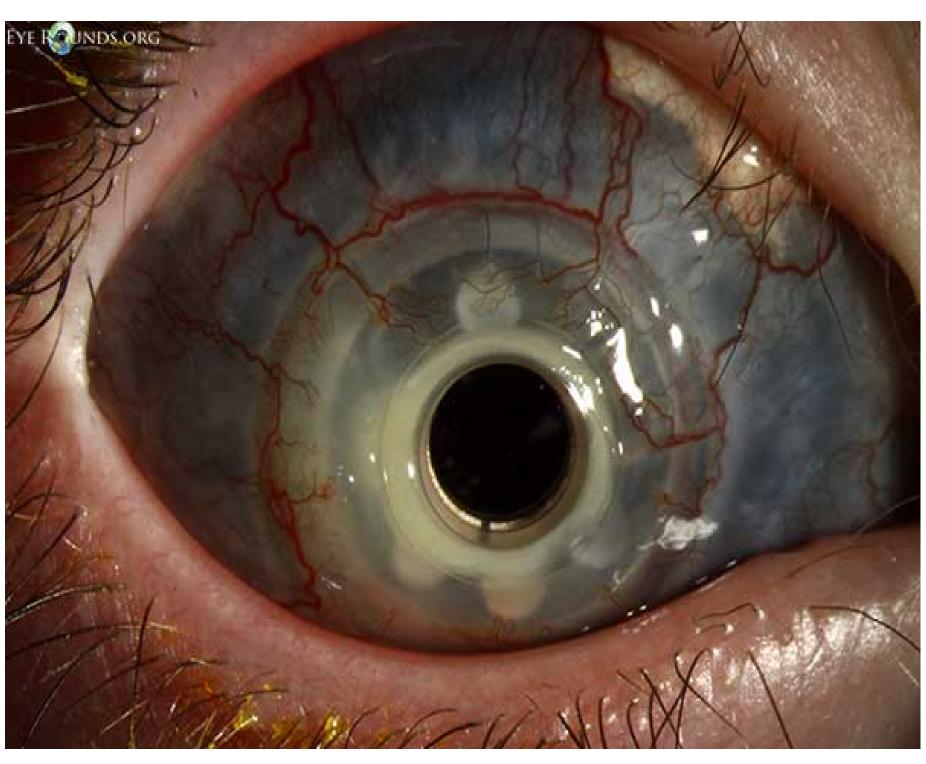
Differential Diagnoses

Bacterial keratitis Fungal keratitis Endophthalmitis Retroprosthetic membrane

Below: External photograph of our patient demonstrating KPro I OD, lateral tarsorrhaphy OU, and trichiasis OU



Above: Photo of a normal post-operative Boston Kpro I



Above: Microbial infection with 360 degree ring infiltrate around

Additional Studies

Cultures (9/11/17)

Positive for Methicillin Resistant Staph Aureus

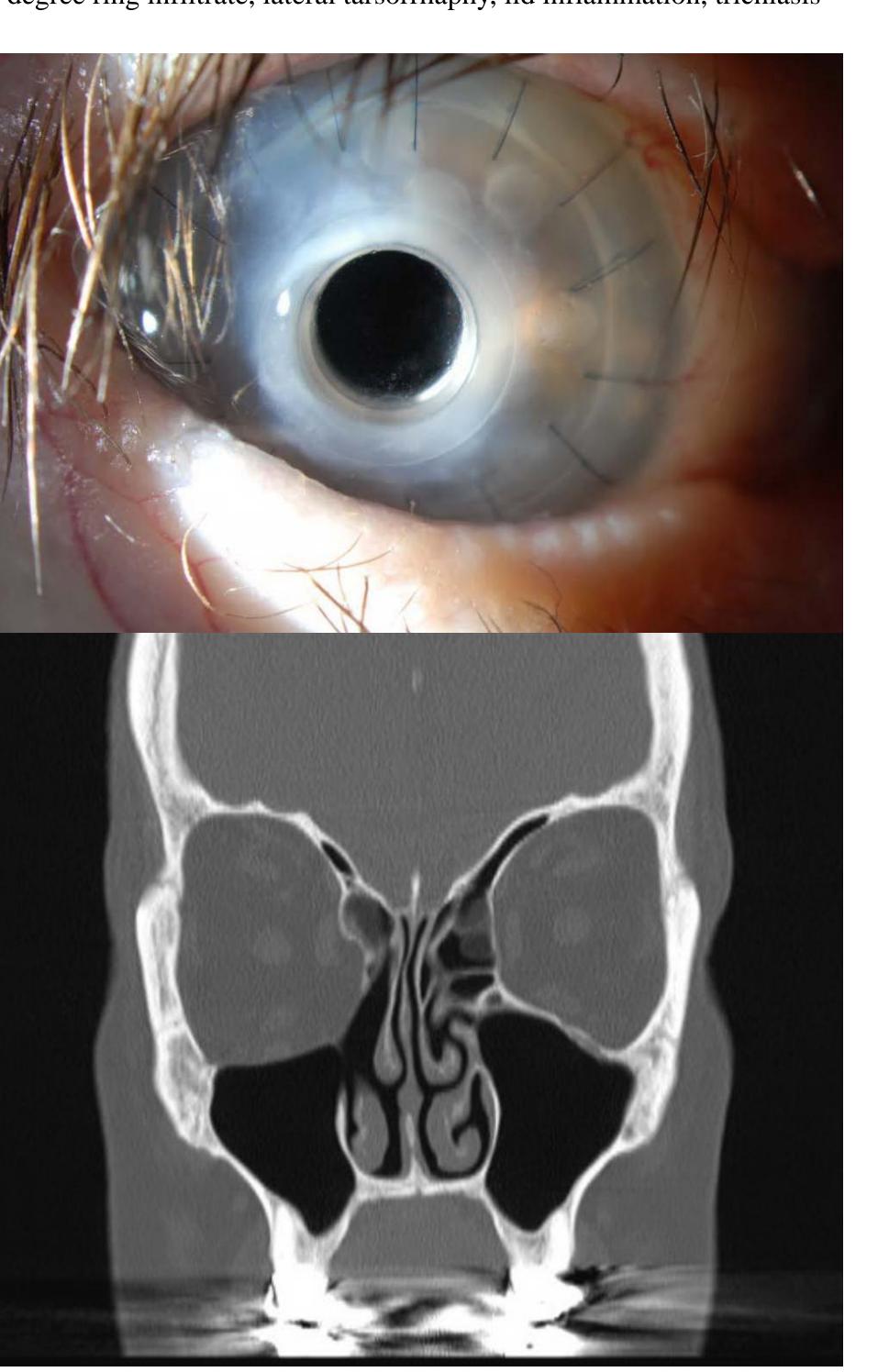
CT Orbit

Status post partial resection of the medial wall and floor of the right orbit as well as a prior right lateral orbitotomy with left rightsided proptosis compared to the left. Low density in the inferior rectus muscle is also compatible with thyroid eye disease.

Last known TFTs (3/12/17)

TSH: .06 Free T3: 3.1

Below: Slit lamp photo of the right eye showing KPro with 360 degree ring infiltrate, lateral tarsorrhaphy, lid inflammation, trichiasis



Above: Coronal CT orbit demonstrating evidence of prior orbital decompression OD

Below: Slit lamp photo of the patient's left eye demonstrating diffuse epitheliopathy secondary to trichiasis, exposure, and meibomian gland dysfunction



Assessment

OD: exposure keratopathy secondary to proptosis, lagophthalmos, and trichiasis; not corrected by lateral tarsorraphy. Inability to keep bandage CL is contributing to above findings.

KPro I with likely microbial infection at stem

OS: Exposure keratopathy secondary to proptosis, lagophthalmos and trichiasis; not corrected by lateral tarsorraphy



Above: External photograph showing significant lagophthalmos OU. This photo further illustrates the importance of bandage contact lens in the setting of KPro in patient at risk for exposure

Treatment

Cultures obtained in office Refer to Oculoplastics for tarsorraphy revision and electroepilation Fortified Vancomycin q1h Fortified Tobramycin q1h

Consider fortified antifungals pending cultures

Conclusion

This case demonstrates a patient with a complex ocular history who was treated with a Boston Keratoprosthesis Type I for an unconventional diagnosis. Typically this procedure is designated as a last resort effort to restore/maintain vision in patients with multiple failed PKP or severe ocular surface disease from SJS, OCP, chemical injury, or aniridia. This patient had a history of several ocular findings that predispose her to complications with a KPro. It is not common practice to treat exposure keratopathy from uncontrolled exophthalmos with a KPro. It is unusual that the patient's underlying proptosis was not managed as the primary cause of her corneal disease. The patient's previous history of lateral tarsorraphy presented an issue with keeping the bandage CL in place. This is a significant problem with KPro I as the graft is nearly guaranteed to have complications due to desiccation and exposure. The cultures that were attained on initial presentation came back positive for Methicillin Resistant Staph Aureus. The patient was continued on fortified antibiotics until follow up. She was also seen by oculoplastics and is scheduled to have LLL and LUL recession with posterior spacer graft from the upper lid tarsus. There are several factors contributing to this complicated clinical picture, which highlight the necessity for a multi disciplinary treatment approach. The Boston Keratoprosthesis Type 1 can be an effective way to preserve vision in complex corneal disease when appropriately indicated.