

# AlphaCor Implantation in Patient with Ocular Cicatricial Pemphigoid

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## Abstract

**Purpose:** To report a case of successful treatment of a patient with ocular cicatricial pemphigoid with AlphaCor keratoprosthesis implantation and subsequent cataract extraction.

**Methods:** We report a case of successful AlphaCor keratoprosthesis implantation in patient with OCP. A careful preparation (repeated electrolysis of trichiasis and fornix and lid reconstruction surgery) was done before AlphaCor surgery. Uneventful phacoemulsification with posterior chamber intraocular lens implantation was performed 18 months after stage II AlphaCor surgery.

**Results:** Long-term (6 years) retention of keratoprosthesis was achieved with improvement of visual functions.

**Conclusions:** Ocular cicatricial pemphigoid is considered as a relative contraindication for AlphaCor implantation. We have achieved very good long-term outcomes with optimization of patient's condition before AlphaCor surgery.

**Keywords:** Ocular cicatricial pemphigoid; AlphaCor; Surgical treatment; Visual functions

## Introduction

Ocular cicatricial pemphigoid (OCP) is a chronic cicatrizing conjunctivitis. Although it is a chronic vesiculobullous disease primarily involving the conjunctiva, it frequently affects other mucous membranes, including the mouth and oropharynx, genitalia, and anus as well as the skin [1]. Patients frequently present with recurrent attacks of mild and nonspecific conjunctival inflammation with an occasional mucopurulent discharge. In its early phases, this insidious disease may present with conjunctival hyperemia, edema, ulceration, and tear dysfunction. Oral mucosal lesions are common and may be a clue leading to early diagnosis. Although the clinical course is variable, progressive deterioration with common remissions and exacerbations usually occurs. Destructions of goblet cells, keratinization of conjunctiva, shortening of the fornices, and symblepharon formation are the main ocular findings. With progression of scarring, entropion and trichiasis develop, leading to abrasions, ulceration, corneal vascularisation and scarring. The treatment of OCP includes local and systemic drugs. Systemic treatment should be continued until all signs of active disease have resolved. Corticosteroids are used as rescue medication, for curtailing acute exacerbations. However, because of their well known long-term adverse effects, corticosteroids must be combined with immunosuppressive and/or anti-inflammatory agents [2]. Other measures, such as surgical correction of eyelid deformities and intraocular surgery, are most successful when disease activity has been under control for an extended period of time. Cryotherapy of the eyelids or electrolysis and other measures aimed at control of trichiasis are essential. Donor graft penetrating keratoplasty (PK) or keratoprosthesis implantation have been performed with limited success.

AlphaCor is an artificial cornea for patients whose corneal blindness is unlikely to be managed successfully by means of standard penetrating keratoplasty (PK) with donor corneal tissue. This device is intended to provide permanent (as defined by FDA) rather than temporary, corneal replacement. In appropriately selected patients, AlphaCor allows replacement of a diseased and opaque host cornea

and allows transmission and refraction of light, with an acceptable field of view, restoring as much as possible of the patient's full potential visual acuity. It's design provides an acceptable cosmetic outcome [3].

We would like to report a case of successful treatment of a patient with ocular cicatricial pemphigoid with AlphaCor keratoprosthesis implantation and subsequent cataract extraction with posterior chamber intraocular lens implantation.

## Case Presentation

A 75-year-old man with long term history of repeated bilateral cicatrizing conjunctivitis was referred to our department in April 2006. He was evaluated and treated in a number of ophthalmology departments previously and his documentation and medical records were incomplete. Diagnosis of OCP was made based on clinical features and histological analysis more than 10 years ago. He was treated sporadically with topical and systemic immunosuppressive agents and repeated (4 times) unsuccessful donor graft penetrating keratoplasties (PK) were performed in his right eye. Over the past year his condition was stable only on topical medication.

Best corrected visual acuity was hand movement in both eyes at presentation. Slit lamp evaluation revealed partial tarsoraphy in his right eye, bilateral conjunctival scarring with symblephara and trichiasis in the left eye. There were bilateral corneal leukoma with deep vascularisation in 4 quadrants. Ultrasound examination showed normal posterior segment configuration. An attempt to optimize conditions

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in his left eye was made. Repeated electrolysis of trichiasis and lids and fornix reconstruction surgery were performed. The patient was educated about the necessity of long-term regular eye and lids hygiene, and lubrication drops were administered. Because the conditions had improved and remained stable, first stage of AlphaCor implantation surgery was performed in December 2006 in his left eye. After surgery antibiotic (chloramphenicol 0.5%) and steroid (dexamethasone 0.1%) eye drops were administered. Biointegration of keratopresthesis was sufficient (Figure 1) and second stage surgery was performed on April 2007 (Figure 2). Best corrected visual acuity (BCVA) improved to 0.2 for distance, and Jaeger no.3 for near vision. We have not observed any complication. Postoperative topical medication included antibiotic and steroid drug, Doxycycline orally (100 mg twice a day) was administered to reduce the enzymatic response (production of gelatinase and collagenase) of the host tissue after AlphaCor implantation.

During next 18 months slow cataract progression was observed (Figure 3). Visual acuity has worsened to 0.05. An uneventful phacoemulsification with posterior chamber intraocular lens (PC IOL) implantation was performed with very good postoperative outcomes (Figure 4). Best corrected visual acuity improved to 0.3 for distance, and Jaeger no.3 for near vision. The survival period of AlphaCor is now more than 6 years and the patient is very satisfied.

## Discussion

AlphaCor is a biocompatible, flexible, one piece device made from poly (2-hydroxyethyl methacrylate) (PHEMA). This material was chosen because it is hydrophilic character and therefore permits



Figure 1: Left eye after stage I AlphaCor surgery.



Figure 2: Left eye after stage II AlphaCor surgery.



Figure 3: Left eye before cataract surgery.



Figure 4: Left eye after cataract surgery.

penetration of biological fluids from the host tissue in order to facilitate bio-colonization. The manufacture of AlphaCor utilizes the different physical forms of PHEMA obtained by varying the water concentration during manufacture. The outer rim, or skirt, of the concentric core-and-skirt device comprises opaque high water content PHEMA sponge, with porous structure suitable for biointegration by cellular ingrowth and collagen deposition. The central core of the device is a transparent PHEMA gel, which provides the transparent optic with the required refractive power. The two concentric regions are joined by means of an interpenetration of polymers across a junctional zone known as the interpenetrating polymer network (IPN) [3-5]. The entire device has a diameter of 7.0 mm, a thickness of 0.6 mm and surface curvatures that result in appropriate refractive power when implanted. The device is presently available in two powers: AlphaCor-A (for aphakic patients) and AlphaCor-P (for phakic or pseudophakic patients). It is placed within a lamellar corneal pocket with tissue posterior to the optic being removed at the time of implantation (stage I surgery) and the tissue anterior to the optic being removed secondarily (stage II surgery) [3,4,6]. The porous skirt remains enclosed within corneal stromal tissue, with which it biointegrates due to cellular colonization and collagen deposition. Alphacor design features include its flexibility and form (analogous to a small donor corneal graft) that allows a relatively non-invasive implantation procedure. The lack of any rigid components minimizes mechanical stresses and makes estimations of intraocular pressure (IOP) possible. The design of the optic, when associated with an opening in the covering tissues after completion of stage II surgery, produces an acceptable visual field and allows intraocular examination. The IPN between the core and skirt creates a permanent and very strong junction preventing aqueous leakage.

The appropriate selection of patients for AlphaCor surgery is crucial for success. There should be severe, debilitating corneal disease causing blindness, with a poor chance of success from primary or repeated donor PK. AlphaCor performs best in a reasonable normal ocular environment. This includes relative eyelid health, a sufficient tear film and an absence of active inflammation. Generally, three broad classes of potential AlphaCor recipient could be identified: 1) those with poor prognosis from donor PK, but with good prognosis for AlphaCor implantation; 2) those with a poor prognosis from donor PK and also a relatively poor or uncertain prognosis, in terms of final vision, with artificial cornea, due to previous glaucomatous damage or macular disease, but with a good prognosis for an anatomically satisfactory outcome without significant complications; 3) those with a greater risk of significant complications affecting not only final vision but also reducing the chance of successful long-term device retention. However those categories are not strictly divided and the indications have been evolving with experience. History of herpes simplex virus infection (HSV) was previously considered as an exclusion factor for AlphaCor surgery, but new data proved that HSV is not a risk factor for melting [3,7]. Ocular cicatricial pemphigoid is considered as a relative contraindication for AlphaCor implantation (category 3).

Ocular cicatricial pemphigoid is an acquired sight-threatening autoimmune disease. It ranks in heterogeneous group of autoimmune blistering diseases - Mucous membrane pemphigoid (MMP). The immunopathological mechanism is represented by a type II hypersensitivity reaction, in which the antigen-antibody-complement interaction, takes place at the level of the conjunctival epithelium basement membrane. This condition can produce severe conjunctival damage. Clinically, OCP is a bilateral disease that is characterized by acute inflammation of the conjunctiva, with redness, blisters, and ulceration of the conjunctiva [8]. Most importantly, however, up to 42% of patients continued to demonstrate progressive conjunctival scarring in the absence of clinically detectable inflammation [1]. Chronic inflammation is associated with subepithelial scarring that leads to fornix shortening, leading to symblepharon, ankyloblepharon, xerophthalmia, keratoconjunctivitis sicca, blepharitis, keratinization of the lid margin, entropion, and trichiasis formation. These result in ocular surface disfigurement with corneal ulceration, infection, and ultimately blindness. Pharmacological treatment includes local and systemic immunosuppressive and/or anti inflammatory drug. Surgical correction of eyelid deformities, cryotherapy of the eyelids or electrolysis and other measures aimed at control of trichiasis are essential. Donor graft penetrating keratoplasty (PK) or keratoprosthesis implantation have been used with limited success. Our case showed that the patient with OCP has achieved very good long-term outcomes after AlphaCor implantation. Optimization of patient's condition prior to AlphaCor surgery (repeated electrolysis of trichiasis, and fornix and lid reconstruction surgery) was a key factor for success. Even uneventful and successful phacoemulsification with PC IOL implantation was performed 18 months after stage II AlphaCor surgery. Phacoemulsification in patients with AlphaCor is relatively challenging surgery but good results could be achieved [9,10].

In conclusion, care of the patients with OCP is challenging and

time consuming, ongoing vigilance in follow-up is essential and each case requires individual assessment and management.

#### Conflict of Interest

None of the authors has a financial or proprietary interest in any product mentioned.

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