Management of Vitreoretinal Complications in Eyes With Permanent Keratoprosthesis

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Objectives: To evaluate the spectrum and treatment of posterior segment complications in eyes that had undergone successful keratoprosthesis (KPro) placement and to determine whether transeyelid vitrectomy techniques could be effectively used in eyes otherwise vulnerable to surface exposure.

Design: In the last 10 years, 110 patients received a Dohlman-Doane KPro at the Massachusetts Eye and Ear Infirmary, Boston. We evaluated 22 eyes in 18 patients that required subsequent vitreoretinal surgery to treat posterior segment complications. One surgeon using modified vitreoretinal techniques, as described below, performed all vitreoretinal procedures.

Results: The posterior segment complications included 6 cases of retro-KPro membranes, 13 cases of retinal detachments, and 5 cases of isolated vitreous opacities. All 6 retro-KPro membranes were effectively removed by vitrectomy without significant complication and 3 of these patients enjoyed improvement of visual acuity of at least 5 Snellen lines. Of 13 cases of retinal detachment, 6 patients had some improvement in visual acuity, 5 showed no appreciable change, and 2 had some decline in the final visual acuity. In all 5 cases of isolated vitreous opacity, the media was effectively cleared with pars plana vitrectomy. Three patients enjoyed improvement of visual acuity of at least 3 Snellen lines. Four cases of transeyelid vitrectomy were attempted and anatomic success was achieved in all 4 and vision improved in 3 of these patients. No special surgical complications were encountered in any of the 22 eyes as a result of these modified surgical techniques.

Main Outcome Measures: Best preoperative and postoperative visual acuity and anatomical success were evaluated in relation to the preoperative posterior segment complication.

Conclusions: Modified vitreoretinal surgical techniques can be effectively and safely used to treat posterior segment complications in patients with KPro devices. Retro-KPro membranes and other vitreous opacities were the most amenable to treatment. Retinal complications posed a special challenge. However, all of these cases highlight that modified vitrectomy techniques can be used in eyes with permanent KPro devices. These techniques can be performed without additional risk to the eye. Additionally, we demonstrated that transeyelid vitrectomy techniques could be used effectively to manage complications in eyes with severe ocular surface disease without undue exposure of vulnerable tissues.
PATIENTS AND METHODS

To date, 146 cases of KPro surgery were performed at the Massachusetts Eye and Ear Infirmary, Boston, with 110 cases performed in the last 10 years using modified Dohlman-Doane KPro designs. These cases included 13 patients with Stevens-Johnson syndrome, 27 with ocular cicatricial pemphigoid, 28 cases of alkali burns, and 42 other cases that include trauma, viral keratitis, and various corneal dystrophies.

All of the implanted KPro devices are of the original or modifications of the Dohlman-Doane design. Preoperative criteria and surgical placement of the KPro were previously described. The structural dimensions of type I and type II KPro devices are shown in Figure 1.

To identify the most common posterior segment complications, we retrospectively reviewed the medical records of all of the patients who underwent KPro surgery in the last 10 years who required pars plana vitreoretinal surgery. We analyzed all of the 22 eyes in the 18 patients who required vitreoretinal surgery. These included 12 patients with type I KPro devices and 6 with type II KPro devices. The pre-KPro diagnoses in these patients included 3 patients with ocular cicatricial pemphigoid, 2 having Stevens-Johnson syndrome, 3 having chronic uveitis, 4 having chemical burns, and 1 each having atrophy, trauma, Fuchs dystrophy, graft-vs-host disease, pseudophakic bullous keratopathy, and idiopathic corneal neovascularization. The posterior segment complications included 6 cases of retro-KPro membranes, 13 cases of retinal detachment, and 5 cases of isolated vitreous opacity. The preoperative diagnoses are listed in the Table. One of us (D.J.D.) performed all of the surgical procedures required in these patients. Informed consent, as determined by institutional requirements and regulations, was obtained from all patients prior to surgery.

PREOPERATIVE EVALUATION

Preoperative evaluation typically required ultrasound evaluation, although in rare cases indirect ophthalmoscopy was sufficient to characterize the posterior segment abnormality. Doppler ultrasound of the optic nerve head for verification of integrity of the central retinal vessels was performed in patients with questionable light perception or suggestive of irreversible retinal damage. In the specific case of retro-KPro membranes, referral for pars plana vitrectomy (PPV) was made after either failure of the Nd:YAG capsulotomy to achieve the desired outcome or when it was determined that such laser treatment could induce further complications, as in the case of thick fibrovascular membranes. In cases of suspected endophthalmitis, initial treatment attempts were made with intravenous antibiotics following vitreous tap and injection. If these treatments failed to achieve vitreous clearing, they were referred for PPV with culturing of a vitreous sample.

Cases with vitreous hemorrhage were referred to the vitreoretinal service when an underlying retinal tear or detachment was suggested. Initial studies included B-scan ultrasonography when direct visualization was impaired. In most cases of retinal detachment, direct visualization of the retina identified the detachment. However, B-scan ultrasonography was used to identify the extent of the detachment, sites of possible tears, as well as points of vitreous adhesions. In most cases of retinal pathology, direct intraoperative visualization provided the most accurate and useful characterization.

SURGICAL TECHNIQUES

Because the anterior segment anatomy is frequently foreshortened in eyes undergoing KPro surgery, often with profound fibrosis and contracture in the area of the iris root and ciliary body, incisions for vitrectomy were located as anteriorly as possible in these cases. To place the incisions just beyond the KPro back plate, 9-mm marks spanning the KPro center were placed as entry sites for 3-port or 2-port PPV. Entry sites were made through the conjunctiva with a microvitreoretinal blade. Infusion was established using a variety of techniques. Although conventional sutured cannulas were used, it was more common to use a bent 21-gauge butterfly needle with the tubing taped to the eyelid to maintain position. This later technique had the advantage of maintaining a formed globe from the beginning of the surgery. Bottle height was maximized to maintain the chamber and facilitate intraocular hemostasis. Zeiger and bent microvitreoretinal blades were alternatively used to incise and free the retro-KPro membranes from the back plate, and the fragments were removed with the vitrector. Once adequate visualization is achieved, the remainder of the vitreous is removed as necessary. Patient 2 demonstrates a case of simultaneous retro-KPro membrane and endophthalmitis (Figure 2).

Three-port vitrectomy was used in all cases of suspected retinal pathology to allow for maximal visualization and flexibility in tissue manipulation. A Charles irrigating contact lens was initially used for visualization of the anterior vitreous. Further manipulations were performed using a contact wide-field viewing system with a 130° wide-angle lens to maximize peripheral visualization. When necessary, endolaser photocoagulation was performed with a straight-probe. A 5000-centistoke silicone oil was used at the end of the case for prolonged retinal tamponade in most cases, but long-acting gases were also used. The eye was filled to the edge of the KPro back plate.

Many patients who a KPro device have severe ocular surface disease necessitating the placement of a transeyelid type II KPro device. To avoid disturbing this interface, a microvitreoretinal blade was used to simultaneously penetrate full thickness through the eyelid, conjunctiva, and sclera into the vitreous space. Infusion was established with a 21-gauge butterfly-needle technique. The remainder of the techniques are similar to those described earlier for the transconjunctival approach. The ports were closed with deep 10-0 nylon skin sutures without the need for scleral sutures. Such a procedure was required in case 3 in which a dense retro-KPro membrane limited visual acuity and was refractory to Nd:YAG treatment. Figure 3 shows the surgical techniques used in the treatment of this patient.
RESULTS

Of the 110 cases of KPro placement in the last 10 years at the Massachusetts Eye and Ear Infirmary, there were a total of 22 cases in 18 patients with posterior segment complications. The most common posterior segment problems encountered were retro-KPro membranes, endophthalmitis, retinal detachments, epiretinal membranes, and vitreous hemorrhages. The most common posterior segment complication requiring surgical intervention was retinal detachment, which was found either in isolation or in combination with a vitreous opacity in 13 cases. The retinal detachments were seen in association with other retinal pathology including extensive cyclic membranes, epiretinal membranes, proliferative vitreoretinopathy, subretinal fibrosis, and other preretinal vitreous adhesions. There were 6 cases of retro-KPro membrane, 3 of which were in isolation. Two of the remaining 3 were seen in association with a retinal detachment, and the last was in the setting of concurrent endophthalmitis. Additionally, there were 5 cases of isolated vitreous opacity related to endophthalmitis in 4 cases and chronic uveitis in 1 case.

SURGICAL OUTCOMES

All 6 retro-KPro membranes were effectively removed by vitrectomy without significant hemorrhage and with complete clearing of the visual axis. The 3 patients with isolated retro-KPro membrane (cases 1, 3, and 4) enjoyed improvement of visual acuity of at least 5 lines of Snellen. The 1 patient associated with endophthalmitis (case 2) saw an improvement from light perception to 20/200. The 2 remaining patients (cases 20 and 22) were in the setting of a concurrent retinal detachment and saw minimal to no improvement in vision, although the membrane was effectively cleared from the optical axis.

Of the 13 cases of retinal detachment, 6 patients had some improvement in visual acuity (cases 9, 11, 15, 18, 19, and 20), 5 showed no appreciable change (cases 10, 12, 14, 21, and 22), and 2 had some decline in final visual acuity (cases 16 and 17). Anatomical success could not be achieved in 4 patients despite aggressive surgical intervention. Of the 6 patients who showed some visual improvement, 4 had some other vitreous opacity that was simultaneously cleared at the time of detachment repair; 2 with vitreous hemorrhage (cases 11 and 15) and 1 each with uveitis (case 18) and retro-KPro membrane (case 20). Of the remaining 7 patients who saw either no improvement or a decline in vision, 3 patients demonstrated extensive proliferative vitreoretinopathy (cases 16, 17, and 21). In all but 4 patients (cases 14, 19, 20, and 21), the retina was successfully anatomically reached postoperatively. The 4 patients who were determined to be inoperable were secondary to proliferative vitreoretinopathy and chronic scarring.

In all 5 cases of isolated vitreous opacity (cases 5 through 8 and 13), the media was effectively cleared with a PPV approach. Three of these patients enjoyed improvement of visual acuity of at least 3 Snellen lines (cases 5 through 7). One patient (case 8) had a chronic wound leak around the KPro stem that progressed to device extrusion. One patient with chronic uveitis (case 13) developed a retinal detachment postoperatively.

Four cases of transeyelid vitrectomy were performed (cases 3, 18, 19, and 22). In all 4 cases anatomical success was achieved and vision improved in 3 of these patients (cases 3, 18, and 19), with patient 3 enjoying dramatic improvement from hand motions to 20/20 visual acuity after removal of a thick fibrovascular retro-KPro membrane. No surgical complications were seen in any of these patients that could be associated with the transeyelid approach.

In summary, in all 22 cases of vitreoretinal surgery performed for posterior segment complications in KPro eyes, no special surgical complications were induced by these modified techniques. No cases of uncontrolled vitreous hemorrhage, retinal detachment, giant retinal tears, choroidal detachment, endophthalmitis, or KPro device damage were noted in any of the aforementioned cases.

COMMENT

The increase in long-term stability of KPro devices resulted in a shift in the types of complications encountered. There have been 173 cumulative postoperative years with vision of 20/200 or better starting from preoperative visions of counting fingers or worse over the period 1990 through 2000. The factors that have improved surgical outcome include glaucoma shunts, changes in KPro design, prophylactic antibiotics, and immunomodulators. Traditional complications such as corneal melt, implant extrusion, and glaucoma still figure prominently. However, the most common causes for failure are encountered in the posterior segment. Leading among these is uncontrollable uveitis as well as other causes of posterior segment pathology such as retro-KPro membrane, endophthalmitis, and retinal detachment. Management of these complications requires modified vitreoretinal techniques.

RETRO-KPro MEMBRANES

Approximately 30% of eyes will eventually develop retro-KPro membranes. Most are amenable to Nd:YAG laser treat-
<table>
<thead>
<tr>
<th>Patient No./Age, y</th>
<th>Pre-KPro Diagnosis</th>
<th>Pre-KPro Visual Acuity</th>
<th>Type of KPro Device†</th>
<th>Best KPro Visual Acuity</th>
<th>Posterior Segment Complication</th>
<th>Visual Acuity Complication</th>
</tr>
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<tbody>
<tr>
<td>1/32</td>
<td>Graft-vs-host disease</td>
<td>LP</td>
<td>I</td>
<td>20/25</td>
<td>Retro-KPro membrane and anterior vitreous membranes</td>
<td>20/400</td>
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<td>2/68</td>
<td>Atopy</td>
<td>HM</td>
<td>I</td>
<td>20/20</td>
<td>Endophthalmitis, vitreous debris, and retro-KPro membranes</td>
<td>LP</td>
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<tr>
<td>3/34</td>
<td>SJS</td>
<td>HM</td>
<td>II</td>
<td>HM</td>
<td>Retro-KPro membrane</td>
<td>HM</td>
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<td>4/61</td>
<td>OCP</td>
<td>HM</td>
<td>I</td>
<td>20/25</td>
<td>Retro-KPro membrane</td>
<td>HM</td>
</tr>
<tr>
<td>5/74</td>
<td>Pseudophakic bullous keratopathy</td>
<td>HM</td>
<td>I</td>
<td>20/25</td>
<td>Sterile endophthalmitis</td>
<td>HM</td>
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<tr>
<td>6/72</td>
<td>Trauma after cataract extraction</td>
<td>CF 1 ft</td>
<td>I</td>
<td>20/40</td>
<td>Endophthalmitis</td>
<td>HM</td>
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<tr>
<td>7/26</td>
<td>Corneal NV of unclear origin</td>
<td>CF 1 ft</td>
<td>I</td>
<td>20/25</td>
<td>Endophthalmitis and anterior vitreous membranes</td>
<td>LP</td>
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<tr>
<td>8/39</td>
<td>Burn patient</td>
<td>HM</td>
<td>I</td>
<td>20/25</td>
<td>Endophthalmitis and leak</td>
<td>LP</td>
</tr>
<tr>
<td>9/30</td>
<td>Chemical burn after failed grafts</td>
<td>LP</td>
<td>II and Vit†</td>
<td>†</td>
<td>Macula-off RD</td>
<td>LP</td>
</tr>
<tr>
<td>10/58</td>
<td>OCP</td>
<td>LP</td>
<td>I</td>
<td>...</td>
<td>VH, preretinal clot, and peripheral traction RD</td>
<td>HM</td>
</tr>
<tr>
<td>11/49</td>
<td>JRA</td>
<td>HM</td>
<td>I and Vit†</td>
<td>†</td>
<td>Vitreous debris and elevated macula</td>
<td>HM</td>
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<tr>
<td>12/58</td>
<td>Syphilis, uveitis, and hypotony</td>
<td>HM</td>
<td>I and Vit†</td>
<td>†</td>
<td>Uveitis</td>
<td>HM</td>
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<td>12/78</td>
<td>OCP</td>
<td>CF 4 ft</td>
<td>II and Vit</td>
<td>LP</td>
<td>VH and RD</td>
<td>LP</td>
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<td>14/42</td>
<td>Alkalai burn</td>
<td>HM</td>
<td>I and Vit</td>
<td>20/60</td>
<td>RD and PVR, and soft eye</td>
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<tr>
<td>15/55</td>
<td>SJS</td>
<td>CF 4 ft</td>
<td>II and Vit</td>
<td>20/25</td>
<td>Uveitis, dislocated IOL, and RD with identified break</td>
<td>HM</td>
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<td>16/46</td>
<td>Congenital rubella after graft failures</td>
<td>HM</td>
<td>I</td>
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<td>Retro-KPro membrane, cyclic membrane, and macula-on RD</td>
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<td>Alkalai burn after failed grafts</td>
<td>LP</td>
<td>II</td>
<td>20/25</td>
<td>Leak, VH, total RD adherent to KPro, and giant retinal tear and PVR</td>
<td>LP</td>
</tr>
<tr>
<td>... / ...</td>
<td>Fuch dystrophy with failed grafts</td>
<td>...</td>
<td>I</td>
<td>20/200</td>
<td>Retro-KPro membrane and RD</td>
<td>HM</td>
</tr>
</tbody>
</table>

*KPro indicates keratoprosthesis; LP, light perception; PPV, pars plana vitrectomy; ellipses, not applicable; HM, hand motions; SJS, Stevens-Johnson syndrome; OCP, ocular cicatrical pemphigoid; CF, counting fingers; NV, neovascularization; Vit, anterior or open-sky vitrectomy; RD, retinal detachment; AFX, air-fluid exchange; SO, silicone oil; JRA, juvenile rheumatoid arthritis; VH, vitreous hemorrhage; PRV, proliferative vitreoretinopathy; ERM, epiretinal membrane; soft eye, an eye in which there is no detectable intraocular pressure; and IOL, intraocular lens.

†A type I Dohlman-Doane KPro has a 7-mm-diameter front plate with a central 3.5-mm, optically clear zone and a 7-mm–base diameter back plate. This device is used in patients with adequate tear film stability. A type II Dohlman-Doane KPro has a 7-mm-diameter front plate and a central 3.5-mm, optically clear zone along with a 2-mm anterior hub that protrudes through eyelid tissues, and 8.5-mm–base diameter back plate. This device is used in patients with severe ocular surface disease.

‡The follow-up period ranged from 3 to 6 months after the last surgical procedure.
As with any intraocular surgery, one of the most debilitating complications is endophthalmitis. In KPro surgery the rate of sterile endophthalmitis is higher than in other intraocular procedures. This may be secondary to immune reaction to antigens released during tissue melt and necrosis. The clinical picture of sterile and infectious endophthalmitis can be varying and confounding, but in general, sterile endophthalmitis is more insidious in onset and occurs in the absence of substantial ocular pain or injection. Vitreous, as well as intravitreal and intravenous antibiotics are administered in cases suggestive of endophthalmitis. The initial antibiotics of choice are vancomycin hydrochloride and ceftazidime, though these are tailored once the culture results become available. A similar pathologic analysis of one such removed membrane (Figure 4) revealed dense, relatively avascular fibrous tissue with numerous fibroblasts, some of which are pigmented. A dense acellular zone of collagen, with an occasional foreign body giant cell surrounds this tissue. This most likely represents corneal downgrowth around the stem of the KPro. A similar pathology is often seen in failed cases of corneal grafts or cyclitic membranes. The visual improvement seen postoperatively in patients initially seen with isolated retro-KPro membranes highlights the relative efficacy and safety of PPV in eyes with KPro devices with minimal risk to the eye.

ENDOPHTHALMITIS

Treatment of retinal detachment in KPro-treated eyes is challenging both in visualization as well as surgical access to involved retinal tissues. Retinal detachments may be secondary to retinal breaks, but may also be due to vitreoretinal adherions and proliferative vitreoretinopathy that accompany chronic inflammation. In such cases, removal of the instigating traction is paramount to successful results. Another visually significant finding in chronic inflammation is epiretinal membrane formation, as demonstrated in patient 17. While basic surgery...
cal principles are obeyed in KPro-treated eyes, the surgical approaches must be modified to achieve good visual outcome. Scleral buckling procedures are often of limited value owing to the compromised scleral and anterior segment tissues seen in these disease states. One of the great limitations lies in the lack of a suitable expansible substance to achieve adequate retinal tamponade. Although we are able to achieve anatomical success in many of these cases, and in many cases stabilize the vision, the prognosis for visual restoration remains poor. Perhaps earlier intervention or prophylactic treatment in high-risk cases may increase the success rate.

GENERAL PRINCIPLES

Despite the exact nature of the surgical procedure, several common obstacles emerge in KPro-treated eyes. These include obtaining adequate surgical exposure, visualization, and hemostasis. Though exposure and visualization are limited in a closed eye system, we prefer this to the intraoperative complications encountered in open-sky techniques. During PPV, a wide-angle lens system may be used with the illumination of a light pipe to obtain more posterior and peripheral views. Two-port vitrectomy is often adequate in simple cases of vitreous washout, membrane peel, or obtaining a biopsy specimen. However, in more complicated scenarios, standard 3-port vitrectomy is required. During instrument manipulation, care must be taken to avoid damage to the back plate, as iatrogenic opacities in the visual axis will limit visual prognosis. Finally, as blood in the vitreous is an active stimulant, special care must be paid to limiting hemorrhage, both intraocular as well as spillover from external ports. Beginning the surgery with an intraocular penetration by a 21-gauge needle for irrigation limits hemorrhage, permits additional instrument introductions, and prevents the transient inadvertent collapses seen with sutured cannula techniques. The maintenance of a closed, firm eye throughout the procedure is perhaps the single most important aspect in achieving successful visual outcomes in these eyes. We advocate a limited vitreous washout at procedure completion if significant red blood cells are present. Postoperative management includes topical antibiotics, 1% of atropine sulfate, and steroids which may be administered periocularly.

Figure 2. Case 2. Endophthalmitis and retro-keratoprosthesis (KPro) membrane. A, Preoperative view of patient 2 demonstrating mild conjunctival chemosis, retro-KPro membrane, and anterior vitreous debris. B, Intraoperative view demonstrating secured infusion cannula inferiorly and bent microvitreoretinal blade behind the KPro device stripping the retro-KPro membrane. C, Vitrector in the anterior vitreous indicating some of the extensive vitreous veils that were seen throughout the vitreous in this patient. D, Intraoperative view showing numerous preretinal clumps of white blood cells and debris. After a complete vitrectomy was performed, these were removed using a flute needle and performing aspiration with the vitrector.
to obtain surgical access without disrupting the protective surface barriers. Vitrectomy in type II KPro devices poses a unique challenge in all 3 categories of exposure, visualization, and hemostasis. Full-thickness vitrectomy requires a modified infusion device, for which we used a 1.5-in, 21-gauge, butterfly-infusion needle that we inserted as the very first maneuver. This was followed by transeyelid penetration with a microvitreoretinal blade for additional instrument access across the eyelids. With experience, it became relatively straightforward to reintroduce various instruments across these transeyelid incisions into the eye. It is possible that the use of cannulated vitrectomy incisions would be beneficial in these cases to facilitate navigating the instruments across the eyelid and sclera.

**Figure 3.** Case 3. Retro-keratoprosthesis (KPro) membrane in a type II keratoprosthesis. A, Preoperative view demonstrating a type II KPro device with a thick retro-KPro membrane. B, Calipers are set on 9 mm to avoid hitting the KPro back plate. View shows microvitreoretinal blade entering the vitreous space with one single passage through eyelid skin, underlying ocular surface, and sclera. Skin hemostasis is achieved with a 20-gauge needle cautery to avoid inadvertent spillover into the vitreous space. C, Two-port vitrectomy using butterfly-infusion needle superiorly and vitrector inferiorly. The third instrument seen in the bottom-left corner is a 0.12-forceps to stabilize the eye. After the leathery membrane was initially incised using an microvitreoretinal blade, the fragments are gently removed with the cutting action of a vitrector. Care is taken to avoid extensive traction on the membrane as it may be firmly adherent to ciliary body or even peripheral retina. D, After the membrane was removed a clear view of the anterior vitreous is possible. The tip of the butterfly-infusion needle is visible. On postoperative day 1 the patient had 20/20 visual acuity.

**CONCLUSIONS**

The previous cases demonstrate that modified surgical techniques may be successfully used to treat visually devastating complications in KPro-treated eyes. Though general vitreoretinal surgical principles are obeyed, special attention is paid to exposure, visualization, and hemostasis. Keratoprosthesis surgery is itself an evolving field, and management of posterior segment complications will...
similarly require continual modification. There are several limitations, which still await technological and surgical advancements. The vision rescued by heroic anterior segment surgery in end-stage ocular disease can be preserved by continued diligence and innovation in the management of associated vitreoretinal complications.

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REFERENCES


Ophthalmological Numismatics

Carl Ferdinand von Graefe (1787-1840) at age 24 years was the youngest appointed professor of surgery and director of the surgical-ophthalmologic clinics at the University of Berlin. He is a founder of the field of modern plastic surgery and devised operations for blepharoplasty, rhinoplasty, and cleft palate. He was also the father of Albrecht von Graefe. The medal shown was engraved by C. K. Pfeuffer and struck by Loos for von Graefe’s 42nd birthday in 1829. The obverse (Figure 1) depicts his bust facing left surrounded by an inscription. The reverse (Figure 2) depicts an inscription surrounded by an oak wreath.

Courtesy of: Jay M. Galst, MD, 30 E 60th St, New York, NY 10022.