KERATOPLASTY FOR THE TREATMENT OF KERATOCONUS*

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FOREWORD

Keratoconus is a localized conus- or mound-shaped, non-inflammatory ectasia of the central portion of the cornea. The protruding area generally extends over the central half of the cornea and is always thinner than normal, but in some instances the conus may be excentric. Some cases, after beginning to develop this deformity, remain stationary without tendency to get worse; these cases are designated as abortive keratoconus. Others remain stationary after developing more advanced stages of the condition, but are still able to get fairly good vision with the aid of ordinary glasses, although this may require the use of high cylinders.

The conus may be reduced to one fifth of its normal corneal thickness and, in extreme cases, may become still thinner. Occasionally Descemet's membrane and its endothelial lining rupture, the aqueous passes into the parenchyma of the cornea with the subsequent development of corneal edema or hydrops. When this occurs, the protrusion increases. The apex or the whole conus becomes turbid and the condition is made worse until the endothelium regenerates and the acute ectasia clears up either spontaneously or with the aid of pressure dressings, often afterward remaining with permanent scarring of the apex of the conus. Other keratoconus cases show a gradual deterioration of the deformity, generally with progressive scarring of the apex, without the occurrence of acute edema. In some extreme cases, the cornea is so thin

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that, either spontaneously or as a result of minor trauma, it perforates, with the subsequent collapse of the anterior chamber.

Until the advent of the contact lens, cases of advanced keratoconus—where useful vision could no longer be obtained by the use of ordinary glasses— had to remain in a nearly blind state or submit to surgical procedures.

REVIEW OF THE LITERATURE

A review of the literature on the subject of surgical treatment of keratoconus reveals a formidable number and variety of surgical procedures advocated for the alleviation of this affection.

The first surgical procedure applied for the cure of keratoconus was that of Ware,[1] who in 1810 advocated paracentesis of the anterior chamber followed by moderate pressure to prevent the return of the projection. The same procedure was recommended by Dix[2] and Desmarres d'Evreaux[3] in 1847 and Bell[4] in 1785. Adams[5] in 1817 advocated needling of the lens in order to neutralize the increased refractive error produced by the deformity of the cornea.

Middlemore[6] in 1835 and Tyrell[7] in 1840 proposed moving the position of the pupil from behind the most altered portion of the cornea. The operation consisted of incarcerating the iris in a corneal opening made near the limbus. In 1839 Favio[8] resorted to the removal of a V-shaped flap at the apex of the cone, without the application of sutures.

In 1858 Critchett[9] modified the operation of Middlemore and Tyrell by tying a single knot in the prolapsed iris with a fine silk thread. The strangulated portion of the iris fell off in about 48 hours, and the iris remained incarcerated in the corneal cicatrix. The procedure, named "iridodesis" by the author, left the pupil in the desired position to obtain the most useful vision.

Bowman[10] in 1859 resorted to the practice of a double iridodesis. Having observed that vision in keratoconus im-

proved frequently by the use of a stenopeic slit, he incarcerated the pupillary borders twice, near the limbus, at opposite ends of the same diameter. The result was the formation of a slitlike pupil which could be placed in any desired direction across the cornea; Bowman, however, believed the vertically placed pupil was to be preferred.

In 1866 von Graefe[11] recommended the dissection of the superficial layers of the cone with a knife, followed by the application of a silver nitrate stick for the purpose of producing a flattened scar after the ulcer was healed. Meyer[12] in 1887 also advocated Graefe's operation, slightly modified.

Bowman[13] in 1869 and in 1873 resorted to the use of trephining to remove the superficial layers of the corneal cone. The center of the bulged area thus dissected was punctured and kept open with repeated paracentesis until the cone had flattened.

In 1872 Bader[14] claimed to have obtained favorable results by excision of an elliptical piece of full corneal thickness at the apex of the cone. To reduce the danger of iris prolapse in Bader's operation and to assist in early closure of the wound, Badal[15] in 1901 inserted 3 horsehair sutures vertically through the cornea, previous to removal of the apex. The sutures were quickly tied, following the excision of the elliptical piece of cornea.

Critchett[16] in 1895 also advocated the removal of a small elliptical piece of the cone at the apex. The incision was begun with a knife and completed with scissors. Wolfe[17] in 1882 first produced an opacity of the apex of the cone and then made a small artificial pupil behind a transparent cornea.

Grandclement[18] in 1891 advocated tattooing of the cone and optical iridectomy. In 1905 Stoewer[19] used a conjunctival flap to cover the cornea after the excision of the cone.

The cautery was used for the treatment of keratoconus as early as 1879 by Gayet,[20] and by Andrew[21] in 1884 and Critchett[22] in 1895. Since then, the number of contributions endorsing the use of the cautery has been immense. Among

the many authors recommending the cautery to burn the apex of the cone are Tweedy[23] and Sattler[24] in 1900, Swanzy[25] in 1903, Siegrist[26] in 1916 and Knapp[27] in 1929. Siegrist recommended cauterization combined with repeated paracentesis. Swanzy thought that cauterization should not perforate the cornea, whereas Tweedy and Knapp were supporters of perforation. Elschnig[28] in 1904 superficially cauterized the apex of the cone as well as an area of the same width connecting the apex with the nearest point of the conjunctiva at the limbus, the object being to produce vascularization with subsequent proliferation of the connective tissue and flattening of the cone.

A case of keratoconus was cured by Carpenter[29] in 1915 with the use of the high frequency spark. Iridectomy was used by von Graefe[30] in 1858, and advocated by Wells[31] in 1869. Corneoscleral fistulizing operations were recommended by Adams[32] and Tiffany[33] in 1914, by Green[34] in 1920, and by Wibo[35] and Rasquin[36] in 1934.

Fox[37] reported in 1925 that flattening of the cone may follow excision of a corneal segment adjacent to the ectatic portion.

Extraction of the lens, which was advocated by Adams[38] in 1817, was employed by Nicolato[39] in 1930, who recommended extraction of the lens in adults and repeated discissions in younger patients.

Sato[40] in 1941 advocated the splitting of Descemet's membrane in an effort to flatten the cornea.

The techniques just enumerated may be classified in 3 different groups:

1. Intra-ocular operations.
2. Excision of a portion of the cornea.
3. Flattening of the cornea by cauterization or other means.

The advocates of the first group, intra-ocular operations, such as paracentesis, lens extractions, deviation of the pupil, and fistulization, aim to neutralize the corneal deformity

with intra-ocular alterations; but they leave the corneal deformity, with the real cause of poor vision, unchanged.

Those of the second group, recommending corneal excision, approach the problem more sensibly because they try to cure or improve the condition by the removal of the deformity in order to obtain a more normal corneal curvature. This type of surgery, however, is too often disastrous to the eye. The operation frequently results in complications such as high astigmatism, adherent leukoma often extending to the pupillary area, and secondary glaucoma. Therefore, these techniques have been considered too unsatisfactory and hence have never become popular.

By far the most successful, and the least likely to cause serious intra-ocular complications, have been the procedures advocated by the third group for the purpose of flattening the conus by scar tissue formation; whether the scar is brought about by cauterization of the conus with chemicals, electrocautery, high frequency current or the splitting of Descemet's membrane. This last group of surgical procedures results in temporary or permanent improvement of vision, although this improvement seldom approaches vision within normal limits. Furthermore, the scarring designed to flatten the cornea not infrequently defeats the purpose of the operation by leaving the eye with no improvement or with worse vision than before, plus corneal opacities which are undesirable from the cosmetic standpoint.

Appelbaum[41] in 1936 published an excellent paper discussing in detail the etiology, pathologic characteristics, symptomatology, objective signs, and the medical, optical and surgical treatment of keratoconus. Concerning the surgical treatment of this disease, Appelbaum states that "surgical intervention aims to produce flattening of the cornea in order to improve eyesight. When no degree of useful vision is obtained with the use of contact glasses, operative intervention may be considered—but no sooner. Only in cases of advanced or nearly hopeless conditions should the patient undergo opera-

tion. Most ophthalmologists agree with this. Too much cannot be expected of surgical treatment. At best, it gives a result far from ideal and none too lasting. The unsightliness which inevitably follows must be anticipated, and the appearance of the eye is always marred to some extent." Afterward, summarizing his discussion on the surgical treatment of keratoconus, Appelbaum says: "Finally, in stating an opinion, condensing the merits of the operations proposed by an equal number of competent surgeons, one could not improve even today on the statement of Wells[42] in 1873, that 'All these methods of treatment of conical cornea are still upon trial, and nothing decisive can as yet be said as to their relative advantages and disadvantages.' "

Entirely agreeing with the statements of Appelbaum and Wells, I was convinced that the choice of a surgical method for the treatment of advanced keratoconus could not be made from the methods previously described. An impartial evaluation of their efficacy proved that more often than not they brought about a deterioration of the eye instead of the improvement they were supposed to accomplish. I also felt that only cases considered hopeless—where vision could not be improved by the use of ordinary lenses, contact lenses or when contact lenses could not be tolerated—should be submitted to surgery.

The advent of contact lenses enabled many sufferers from keratoconus, whose vision could not be improved with ordinary glasses, to obtain useful vision; but there are still many cases of keratoconus in which vision, on account of dense scarring at the apex, cannot be improved by the use of contact lenses. There are other cases in which the aid of contact lenses succeeds in restoring fairly good vision, but due to ever-increasing irritation, contact lenses are not well tolerated. In still another group of patients, contact lenses are not satisfactory because of their tendency to cause cloudiness of the cornea or because the buffer solution becomes turbid too rapidly. All these cases affected with advanced kerato-

conus, with very precarious vision — less than 20/200 (industrial blindness)—which do not improve with ordinary or contact lenses or where the contact lenses are not well tolerated, are cases in which surgical intervention is justified.

RULES GOVERNING THE SURGICAL TREATMENT OF KERATOCONUS

The successful treatment of keratoconus must be subjected to the following essential rules:

1. Since poor vision seems to be caused by the corneal deformity alone, and not by other intra-ocular pathology, surgery must be limited to the cornea.
2. In order to obtain the best possible vision, the whole corneal protrusion must be removed and replaced with corneal tissue of normal curvature and thickness. The pupillary area must be free of scarring.

The only surgical procedure which fulfills these 2 essential requirements is keratoplasty of the partial penetrating type. Therefore, keratoplasty seems to me the logical procedure to be applied in advanced cases of keratoconus, when useful vision can no longer be obtained by the use of ordinary or contact lenses, or when contact lenses are not well tolerated.

In 1936 I[43] performed the first keratoplasty in an advanced case of keratoconus, which resulted in marked improvement of vision and seemingly lasting cure. Encouraged by the first trials, I proceeded to treat other advanced cases of keratoconus with corneal transplantation. The results have been most encouraging. A high percentage of cases remain with clear grafts, marked improvement of vision and, in so far as my 12 years' experience indicates, are permanently cured. I have already operated upon more than 200 advanced cases of keratoconus and in more than 80% the results can be considered most successful. The average improvement of vision that can be expected is from 20/200 or less to 20/50 or better (Figures 1 and 2).

I have singled out keratoconus for this discussion on keratoplasty, because keratoconus has special features which distinguish it from other corneal opacities or deformities. Failure to observe certain rules in the treatment of keratoconus by keratoplasty may defeat the success of the operation. The rules concerning these special features have to be taken into consideration during the preoperative study, at the time of the operation and during postoperative recovery.

PREOPERATIVE STUDY

During the preoperative study, thickness of the cornea and extent of the keratoconus must be carefully studied with the slit lamp and corneal microscope. It is important to know how extensive the keratoconus is, because the size of the graft and its location depend upon the extent and location of the conus. The graft must be large enough to substitute for all or as much as possible of the conus. If the transplant is smaller than the conus, the deformity is likely to advance. The graft will not develop keratoconus, but the pre-existing thin cornea remaining will continue to protrude; or if it does not advance after the operation, the graft, although clear, remains set in a protruding area of cornea which results in very high myopia, astigmatism or both, thus defeating the success of the operation for visual purposes.

The patient depicted in Figure 3 serves to illustrate this point. He was affected with advanced keratoconus in both eyes; vision in both eyes was fingers at 1 foot and, on account of the pronounced conus, he could not be fitted with comfortable contact lenses. A corneal transplantation was performed in the right eye, using a 4½ mm. graft (Fig. 3a). The transplant remained clear, but was set in a protruding area which resulted in very high myopia and astigmatism. Vision improved only slightly after the operation because of the remaining protrusion. In the fellow eye, a keratoplasty was performed afterward, using a transplant of 5½ mm. (Fig. 3b). Again this transplant was found to be too small; thin cornea

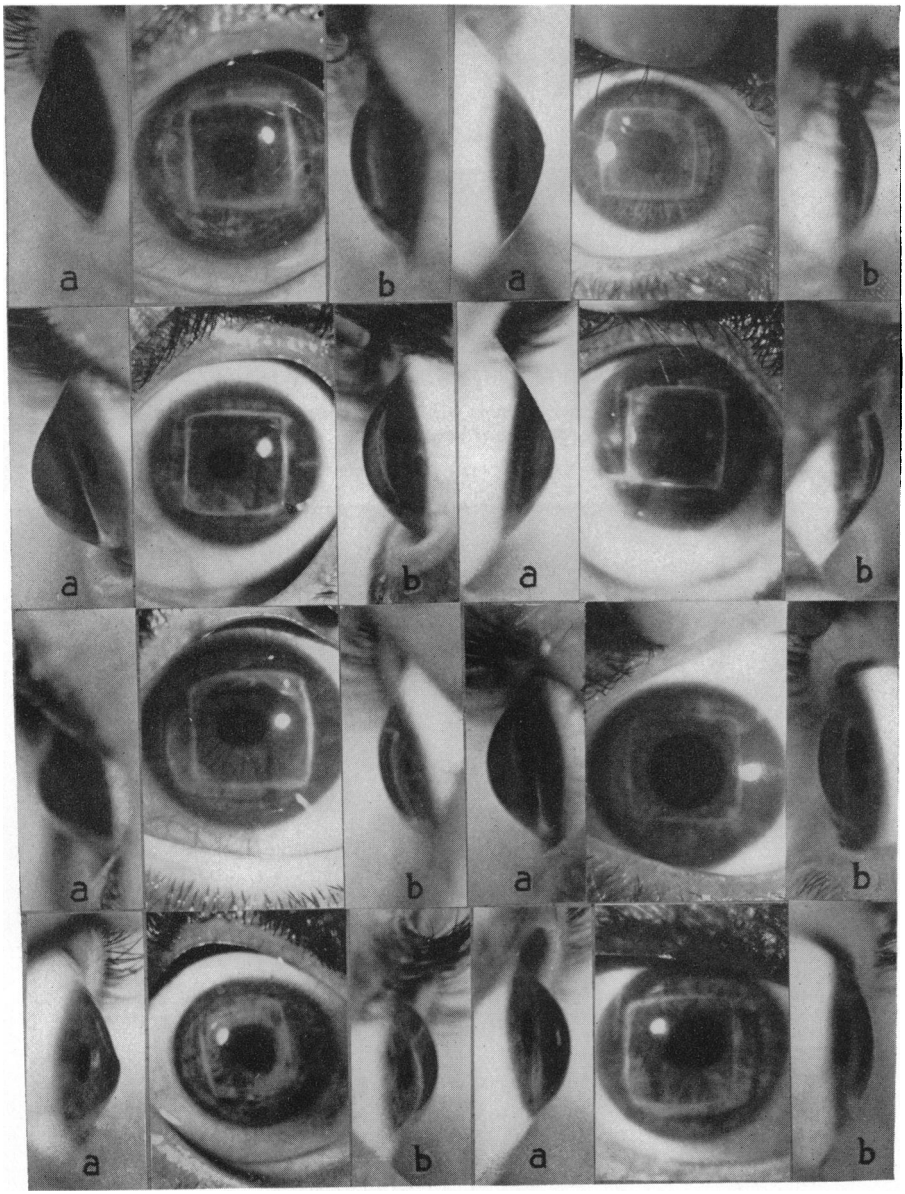

Fig. 1.—Photographs of clear square corneal transplants in eyes affected with advanced keratoconus. Profiles show: (a) before operation, (b) after operation.

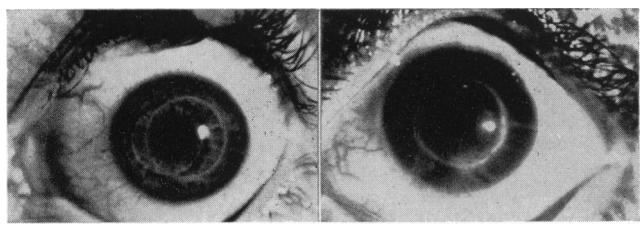

Fig. 2.—Photographs of clear circular corneal transplants in eyes affected with advanced keratoconus.

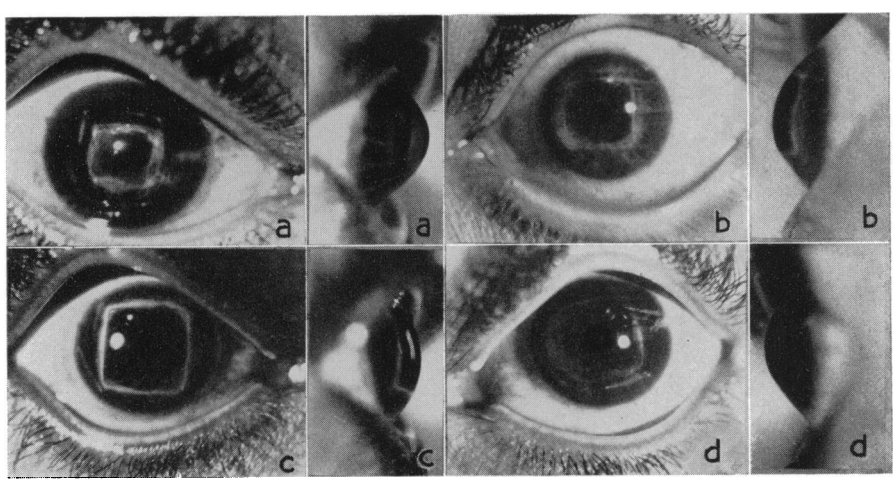

Fig. 3.—Photographs of 4 corneal transplants performed in the 2 eyes of the same patient. (a) 4½ mm. graft, (b) 5½ mm. graft, (c) 7 mm. graft, (d) 6 mm. graft.

remained with subsequent protrusion of the graft and vision only slightly improved. A third transplant of 6 mm. was then performed (Fig. 3d), the second on the left eye, but still was not large enough to excise the whole conus, resulting in moderate protrusion, high myopia of −20 which gave corrected vision of 20/50. Finally, the right eye was again operated upon and, with the experience of 3 previous operations, a larger transplant of 7 mm. (Fig. 3c) was used, by means of which the whole conus was excised. Fairly good corneal curvature was obtained and vision was 20/25−3 with a correction of $-4.00 \supset -1.75 \times 120°$.

The minimum size of graft to be used in keratoconus should be 6 mm., with the exception of some cases with an extremely small conus. In such instances, grafts of 5½ mm. or even smaller can be used. In more pronounced keratoconus, grafts of 6½ mm. or as large as 7 mm. will be necessary.

It is always desirable to place the graft in the center of the cornea in order that the center of the graft will coincide with the center of the pupil. In keratoconus, however, the location of the graft has to be controlled to a certain extent by the location of the conus. When the conus is off-center, the graft should be displaced accordingly, in order to remove as much as possible of the protruding area. In this case, if the graft is small (less than 6 mm.), its edge may fall in or near the pupillary area, interfering with vision. This interference is, of course, greater if the cicatrix is made more pronounced during the healing process, thus reducing the useful area of the graft falling within the pupillary area (Figure 4B). If the graft is large (6 mm. or more), even in case of abundant scarring at the edges, there is always enough useful area of the graft in the pupillary area.

This useful area for the same size graft is larger in the square graft than in the circular one if the corners are placed as illustrated in Figure 4A. This fact should make the square graft more desirable than the circular one when dealing with

large keratoconus. In addition, the corners in the square graft extend farther toward the limbus, and the scarring which

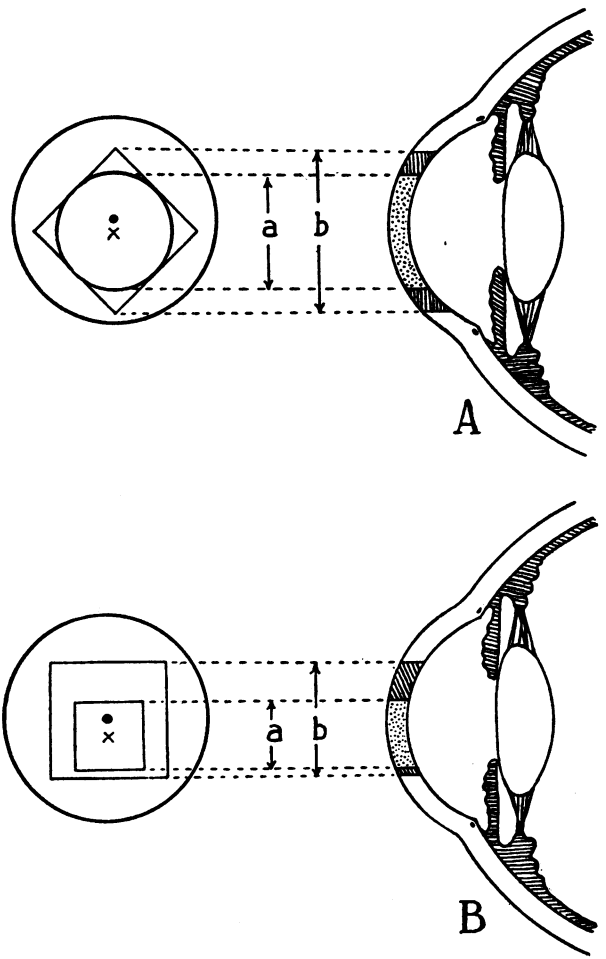

Fig. 4.—Diagrams showing: (A) the position of the edges of excentrically placed round (a) and square (b) grafts in relation to the pupillary area (• shows the center of the cornea, while x shows the center of the graft). (B) the position of the edges of the excentrically placed small graft (a) and larger graft (b).

takes place at the incision has a tendency to greater flattening of the graft and the cornea of the host, resulting in better

curvature than when a round graft is used. In order to accomplish the same effect with a round graft, it would have to be much larger, corresponding to the distance—not from opposite sides of the square graft, but the diagonal distance from corner to corner (Figure 5).

Another factor which has to be considered before the operation is the dilatation of the pupil. When large grafts are used, the pupil should be widely dilated to prevent the formation of anterior synechia, which is one of the most frequent complications in keratoconus. The pupil generally

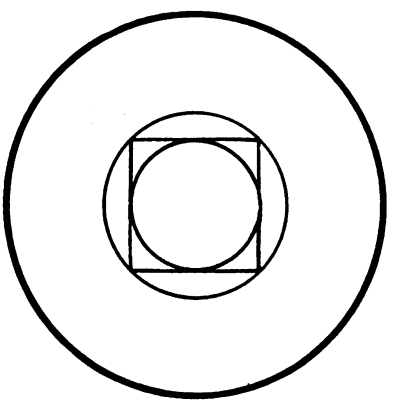

Fig. 5.—Diagram showing the relation of useful vision for the same size graft in the square and round transplant.

dilates widely with repeated instillations of 3% solution of atropine sulphate and 10% emulsion of neosynephrine hydrochloride. If the pupil should fail to dilate beyond the limits of the area where the graft will be located in the cornea, it will be preferable to use a smaller graft or to prepare the eye with a large iridectomy rather than risk the development of anterior synechia, with subsequent danger of opacification of the graft and secondary glaucoma, or both.

In addition to these local factors in the eye, the general condition of the patient should be carefully studied, eliminating all pathologic conditions which are likely to influence the

normal postoperative course of the transplant, such as foci of infection, malnutrition and other systemic conditions. Infections in the nose and throat are particularly likely to cause postoperative uveitis and it would be advisable not to perform a corneal transplantation in the presence of infected tonsils and nasal pathology, including sinus involvement, delaying the operation until these conditions have been cleared up by medical or surgical treatment.

The 2 types of operation performed have been the author's square and circular partial penetrating keratoplasties, illustrated in Figures 6 and 7. A detailed description of the operation has been made in another article.[44]

DURING THE OPERATION

It has already been mentioned that the conus is sometimes reduced to one fifth of the normal corneal thickness and, in extreme cases, may be still thinner. On account of this thinness, while outlining the incisions or inserting the sutures, the cornea may perforate. If the perforation occurs while outlining the incisions—whether with the double-bladed knife or with the trephine—before the sutures have been inserted, it may be difficult to continue the operation. However, the surgeon well trained in keratoplasty will be able, although with more difficulty, to continue with the operation and place the sutures after the cornea has been perforated, particularly if the suture material is good. Of course, in this event the remaining uncut portion of the incision has to be completed with the aid of scissors, both in the square as well as in the circular graft. For the surgeon less skilled in this type of surgery, it would be preferable to postpone the operation for about a week to provide time for the cornea to close the opening firmly and the anterior chamber to re-form, before proceeding with the operation.

If the perforation of the cornea takes place while inserting the sutures because the anterior chamber has been entered with the needle, the aqueous humor will drain slowly, render-

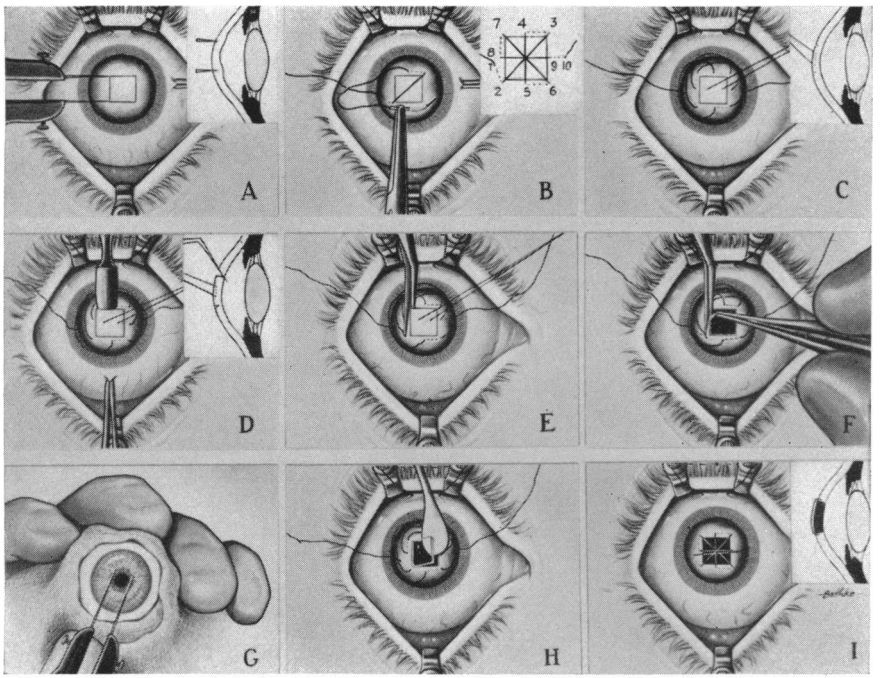

Fig. 6.—Successive steps in author's square partial penetrating keratoplasty.

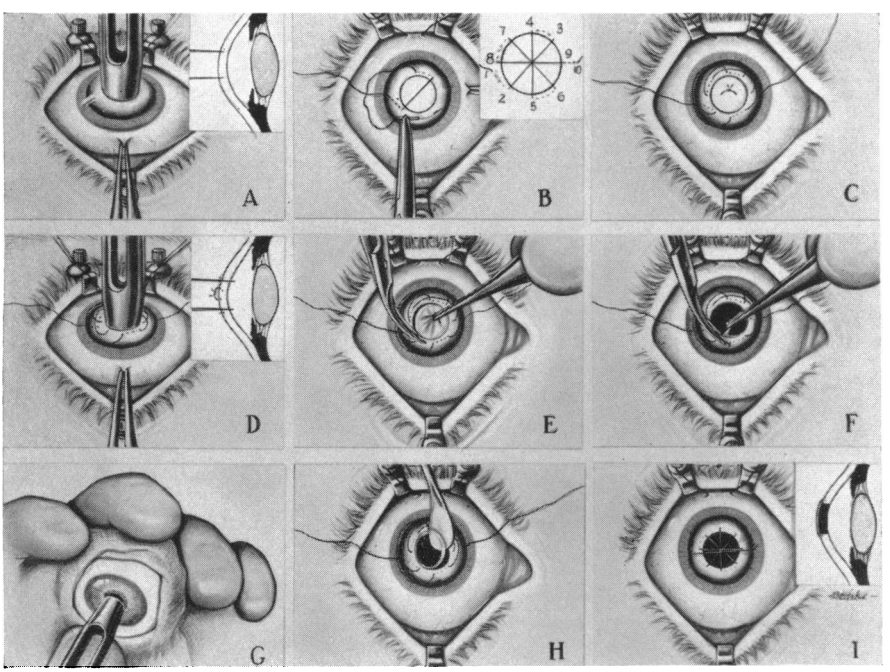

Fig. 7.—Successive steps in author's circular partial penetrating keratoplasty.

ing the eye hypotonic. If this complication takes place in eyes with normal corneal thickness, it is feasible—and not difficult —to withdraw the needle and pass it through more superficial layers of the cornea; otherwise a fistula may form during the postoperative recovery with drainage of aqueous humor, flattening of the anterior chamber and likelihood of intra-ocular infection, development of anterior synechia and secondary glaucoma.

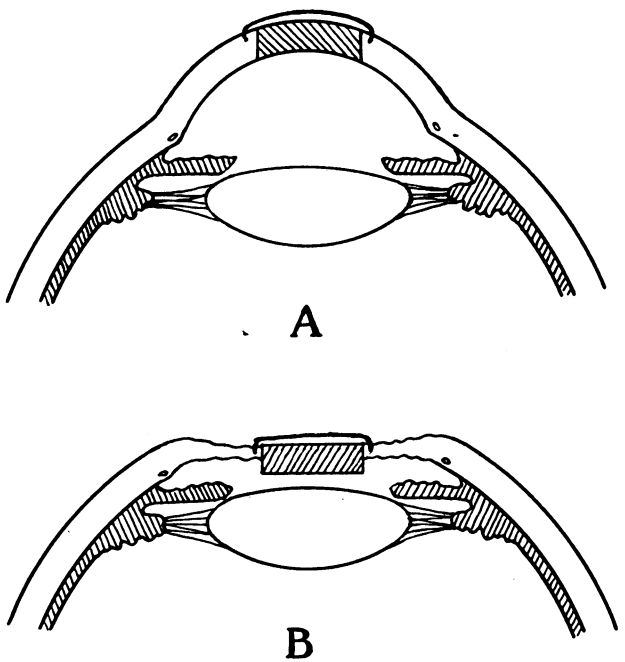

Fig. 8.—Diagrams showing: (A) resilient cornea holding the graft in position, (B) collapsed and wrinkled thin cornea held in position around the graft by sutures placed close to its borders.

In keratoconus, it is possible to carry out the same maneuver of withdrawing the needle and passing it through more superficial layers of the cornea; but on account of the extreme thinness of the cornea in some cases and because the cornea lacks rigidity and resiliency, the cornea collapses and wrinkles (Figures 8A and 8B), so that the reinsertion of the

needle may be found extremely difficult. Under these circumstances also, the surgeon well trained in keratoplasty may proceed with the operation to a successful conclusion; for the less skilled surgeon, however, it would be preferable to postpone the surgical procedure rather than risk failure in the operation.

After the removal of the conus, particularly in eyes where a large transplant of 6½ mm. or more is used, the pupil may be found to be without sufficient dilatation, and the size of the pupil to be smaller than the opening in the cornea. This instance can be prevented before the operation, as has already

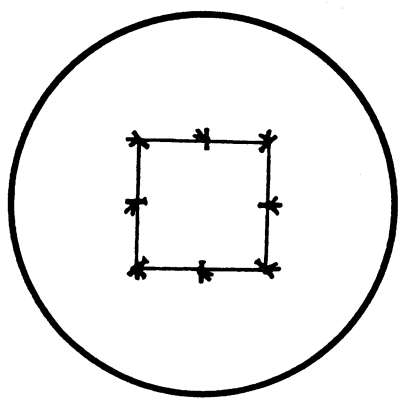

Fig. 9.—Diagram showing border-to-border suturing in a square transplant.

been mentioned by repeated instillations of 3% atropine and 10% emulsion of neosynephrine hydrochloride, and at the time of the operation by injection of cocaine 2% and adrenalin 1:1,000 under the conjunctiva. If, in spite of this preparation before placing the graft in position, the pupil is smaller than the opening in the cornea, direct instillation of cocaine 4% and adrenalin 1:1,000 or even 1:100 into the corneal opening may dilate the pupil wider than the corneal window. If, in spite of the instillations, the iris shows within the opening, it is advisable to excise the involved portion of iris and place sutures border to border, as illustrated in Figure 9,

which, although injuring the graft somewhat, give a better coaptation of the borders, promoting an early restoration of the anterior chamber and tending to avoid the development of anterior synechia. The performance of an iridectomy in these cases will, of course, be contested by those who perform keratoplasty with contracted pupils. Statistics will show in the future which method gives the greater number of anterior synechiae, the one using the contracted pupil or the one using the dilated pupil. Both my experimental work in animals and clinical experience in humans have proved to me that when the pupil is left contracted at the time of placing the graft, it is more likely to develop anterior synechia.

Due to the lack of resiliency of the cornea and the large grafts generally used in advanced cases of keratoconus, it is easier to injure the lens while perforating the cornea than while operating upon eyes with normal corneal thickness and resiliency where smaller grafts are usually used. Therefore, in making the corneal perforation, particularly with the trephine, a great deal of care must be exercised to withdraw the trephine quickly, immediately after penetration into the anterior chamber has taken place, because the cornea collapses and the lens capsule comes in contact with the edge of the trephine almost instantly. On account of the large area of lens exposed when the grafts are large, it is easier to injure the lens while trimming the border of the incision if that should be necessary. Therefore, the extreme care that must be exercised during these manipulations to avoid injury of the lens cannot be emphasized too strongly.

I should like to mention one more complication that may occur in advanced cases of keratoconus. In one instance, I used a square transplant of 7 mm. to substitute completely for the extensive keratoconus with extremely thin cornea. After the conus was excised without injury to the lens, the lens mushroomed through the corneal opening and spontaneously extruded itself intact in its capsule. In this case, in order to maintain the graft firmly in position, it was necessary

to suture the border of the graft to the border of the cornea of the host, using 8 sutures, in the manner illustrated in Figure 9.

If the lens is injured by the keratome or trephine or scissors while completing the incision, it will be advisable to remove as much as possible of the injured lens, including the capsule. In this complication, the vitreous has a tendency to prolapse during the operation or postoperative recovery, thus dislocating the graft. Therefore, it is advisable in these cases, in order to maintain the graft firmly in position, to suture the graft to the cornea of the host, border to border, as illustrated in Figure 9.

In the case of large grafts, it is advisable to have the graft ready before the excision of the cornea of the host, in order not to delay closing the corneal opening as soon as the dissection of the conus has been concluded.

In keratoconus, contrary to what occurs in corneas with normal thickness and resiliency, there is a tendency for the cornea to collapse. This often results in a host opening smaller than the graft, although the graft had been made the same size as the corneal opening. In this case, as opposed to what happens with corneas of normal thickness and normal resiliency where the cornea of the host holds the graft, it is the graft that holds the thin cornea of the host around it. Under these circumstances, the displacement of the whole graft or part of the edge of the graft is not infrequent. In order to prevent this displacement of the graft, 2 essential points must be observed in placing the sutures:

1. The sutures must be placed near the edge of the incision.
2. The distance between the sutures must be shorter than the diameter of the graft.

In spite of these precautions to prevent displacement of the graft, in cases of large grafts, it is sometimes difficult to hold the graft in position. In such instances, direct suturing of the border of the cornea of the host with that of the donor is to be recommended. At least 4 sutures placed in the corners of the

square graft, or at the ends of 2 perpendicular diameters in the circular grafts, are advisable (Figure 10).

One more essential point which must be observed in keratoconus concerns the type of donor material to be used. Since in keratoconus with thin cornea it is not the cornea of the host which holds the graft in place but the graft that holds the cornea of the host around it, it is preferable that the graft be obtained from adult eyes. Younger material from fetus, still-born or infants, does not have enough resiliency to hold the cornea of the host around the borders of the graft.

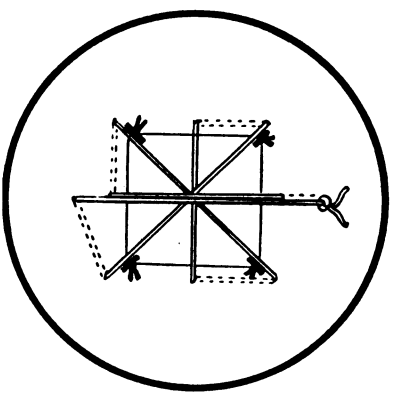

Fig. 10.—Diagram showing author's continuous suture to hold the graft in position, reinforced by 4 border-to-border sutures, placed in the corners of the graft to prevent its displacement.

In cases of excentric keratoconus extending toward the limbus, the graft has to be displaced off-center to coincide with the position of the deformity. In these cases, the graft must be large enough to be able to enclose the pupillary area within the graft, otherwise the scarring at the edges of the graft will interfere with vision. Of the circular and square grafts, the square graft placed with the corners as illustrated in Figure 4A is preferable because it not only includes the displaced conus, but also the central area of cornea, most desirable for useful vision.

POSTOPERATIVE RECOVERY

During the postoperative recovery, there is more danger of displacement of the graft in eyes operated upon for keratoconus than in eyes operated upon for other corneal conditions with normal corneal thickness, and where smaller grafts are used. For this reason, the patient should have both eyes bandaged and remain quiet in bed with fluid and semifluid diet, for about 7 days. During this period the eye should not be examined. Examination of the eye previous to the seventh postoperative day will increase the number of complications, particularly displacement of the graft, with development of anterior synechiae. On the seventh postoperative day, the eye is examined for the first time and penicillin ointment and atropine 1% ointment applied to dilate the pupil. The reactions of the patient are tested at this dressing. If the patient is quiet, not tense, and gives good co-operation, permitting examination of the eye without blepharospasm, the sutures can be removed the following day under local anesthesia. But should the patient be tense, of the high-strung type, with a great deal of blepharospasm—a "squeezer"—the sutures should be removed the following day under general anesthesia, preferably sodium pentothal. The purpose of applying atropine is to obtain dilatation of the pupil because, in keratoconus with thin cornea, at the time when the corneal sutures are removed there may be a collapse of the anterior chamber with loss of aqueous humor, due to slight dehiscence of the incision. Very seldom, however, is this opening extensive, and the transplant almost always cicatrizes well in spite of the collapse of the anterior chamber, and without complications such as development of anterior synechiae. Anterior synechiae are less likely to occur if the pupil has been widely dilated previous to the removal of sutures by the use of atropine.

The development of anterior synechiae is one of the most frequent complications in keratoplasty for the treatment of keratoconus. If the synechia is smaller than 2 mm. occa-

sionally the use of atropine or eserine loosens it. If the synechia is more extensive, it will necessitate surgery in order to prevent opacification, vascularization of the graft and often secondary glaucoma. The surgical treatment of the synechia must not be carried out before the fourteenth postoperative day because, until then, the union of the graft with the cornea of the host is not firm and premature surgery might lead to dislocation of the graft. The operation, however, should not be delayed beyond 3 weeks, otherwise the graft may already have become opaque on account of the synechia. Therefore, the best time to carry out this surgery is in the third postoperative week and preferably under general anesthesia.

SURGICAL TECHNIQUES FOR THE TREATMENT OF ANTERIOR SYNECHIAE

If the synechia extends from ⅕ to ½ of the circumference of the graft, the operation of choice is as follows:

Depending upon the location of the synechia, the incision in the sclera is made in the quadrant nearest the synechia; the lower temporal for synechiae in the lower half, the upper temporal for synechiae in the upper half of the transplant. A routine preparation of the eye, about 1 hour before the operation, is made with instillations of atropine 3% and neosynephrine 10%, in order to obtain as wide a dilatation of the pupil as possible. At the time of the operation, after routine preparation of the eye, argyrol 25%, a 4% solution of cocaine, and a solution of adrenalin 1:1,000 are instilled into the eye. A few drops of cocaine and adrenalin 1:1,000 are injected under the bulbar conjunctiva (Figure 11A), for the purpose of obtaining ischemia, wider dilatation of the pupil and to counteract a tendency to contraction after the anterior chamber has been opened.

An incision is made through the conjunctiva and Tenon's capsule, about 7 mm. from the limbus and parallel to it (Figure 11B). Two 00 silk sutures are inserted in the sclera 4

and 6 mm. from the limbus and parallel to it. These sutures are used for fixation; one of them is held by the assistant and the other by the surgeon. The area of the sclera between the 2 sutures is treated by superficial electrocoagulation or by a hot probe to obtain a bloodless operative field (Figure 11B). With the knife, an incision is made in the sclera similar to that performed for cyclodialysis (Figure 11C). When the sclera has been perforated and the choroid is reached, a long spatula similar to that used for cyclodialysis but 15 mm. in length is introduced through the scleral opening and then between the sclera and choroid into the anterior chamber. The adherent iris is separated from the cornea with the aid of the spatula (Figure 11D). The spatula is then withdrawn and the tip of a Randolph perforated cannula with the shape of a cyclodialysis spatula attached to a syringe (Figure 11E) is introduced in the same manner as the spatula into the anterior chamber and air is injected to push the iris away from the cornea. The cannula is then withdrawn and the conjunctival incision closed with a continuous 000000 silk suture. This cyclodialysis incision several millimeters from the limbus is preferable to incisions in the limbus because, with the latter, it is not always possible to keep the air within the anterior chamber long enough to prevent the recurrence of the synechia.

When the synechia is extensive, the operation which I found satisfactory is as follows:

The operation should be carried out, preferably, under general anesthesia. With the aid of scissors and conjunctival forceps, a conjunctival and episcleral flap of 2 to 3 mm. between 10 and 2 o'clock is made around the limbus (Figure 12A). The conjunctival flap is reflected down over the cornea and three 000000 silk sutures mounted in atraumatic needles are inserted, 1 at 12 o'clock and the other 2 equidistant from the central one and the ends of the conjunctival incision. The sutures are passed first through the conjunctiva and then through the sclera, entering the sclera about 2 mm. from

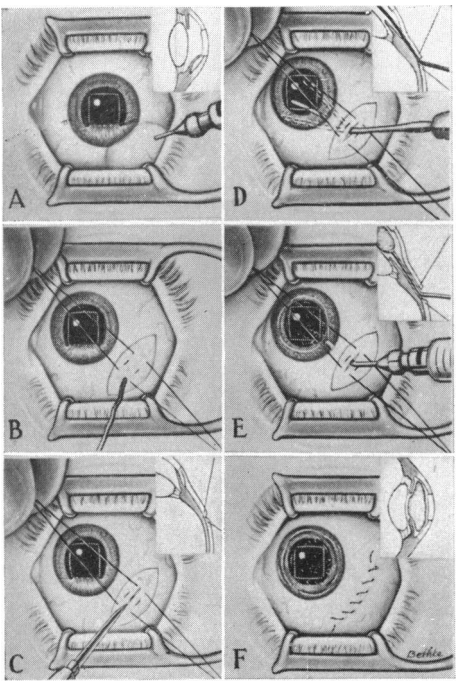

Fig. 11.—Successive steps in author's technique for the treatment of moderate anterior synechiae.

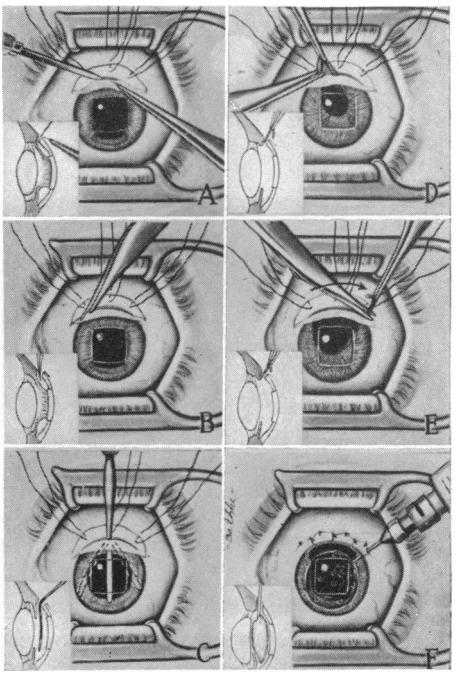

Fig. 12.—Successive steps in author's technique for the treatment of extensive anterior synechiae.

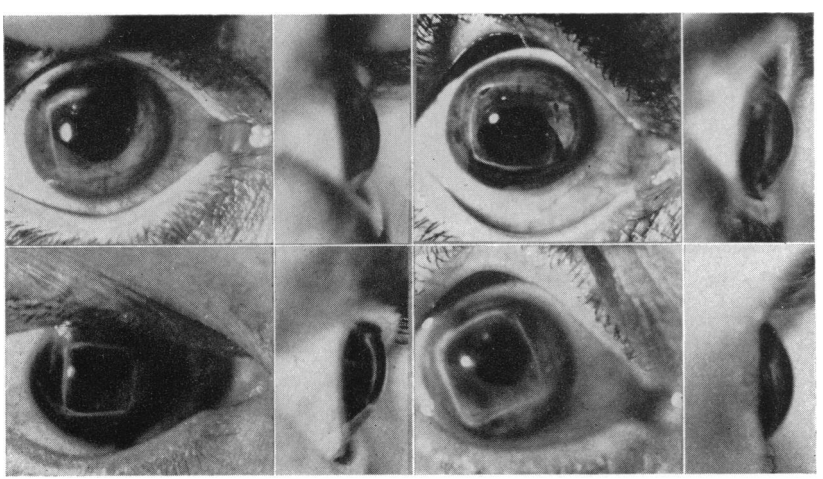

Fig. 13.—Photographs of clear transplants after operations for extensive synechia.

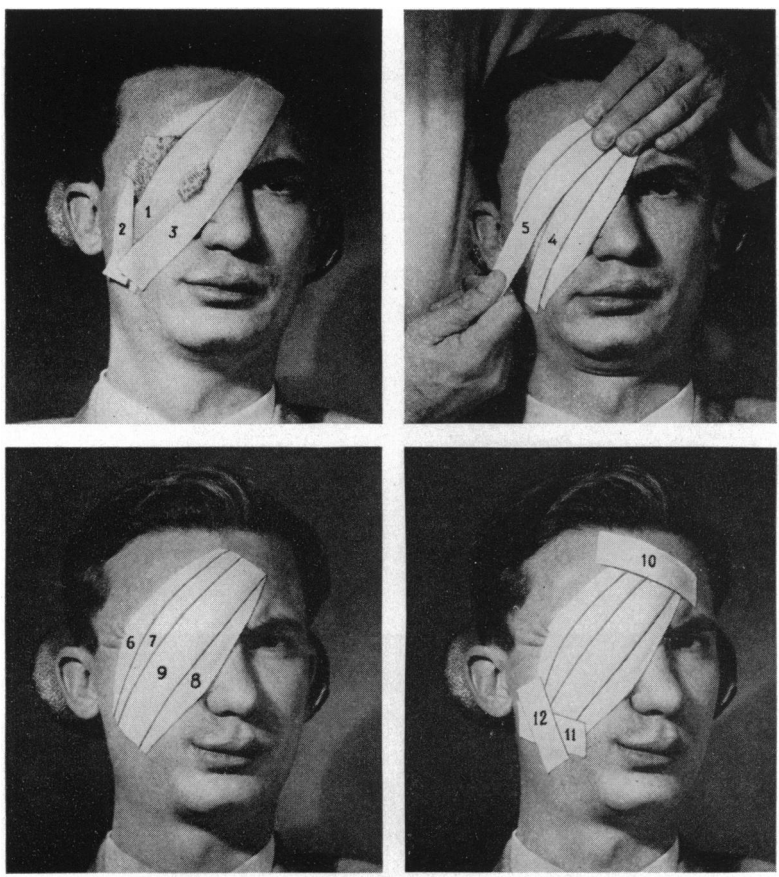

Fig. 14.—Photographs showing successive steps in the application of pressure dressing.

the limbus and taking a bite of about 1 mm. (Figure 12A). The sutures are now drawn out of the way in order to make the incision with a minimum of interference. With the knife, an ab-externo incision is made at the limbus around 12 o'clock (Figure 12A). When the anterior chamber is entered, the incision is enlarged temporally and nasally with the aid of scissors (Figure 12B). With the cyclodialysis spatula, the iris is then separated from the cornea (Figure 12C).

A large iridectomy from 10 to 2 o'clock is then performed (Figures 12D and E). The sutures are passed through the corneoconjunctival junction, including a small bite of corneal tissue, and tied. Additional conjunctival sutures are inserted and air is injected into the anterior chamber (Figure 12F) in an effort to push the iris away from the cornea, thus preventing the recurrence of the synechia.

After this procedure, if an anterior synechia develops in the lower half of the graft, the first operation described can be carried out. I have had several cases, treated thus successively with the 2 operations, in which the eye remained without synechiae, with a clear transplant and good vision (Figure 13).

In these 2 operations, bacteria leading to endophthalmitis or panophthalmitis are often introduced into the eye. I have had several such instances; therefore, the need for a very aseptic technique and great care in performing these operations should be stressed. Prophylactic treatment with systemic application of penicillin should be carried out, beginning the day previous to the operation and continuing until all the signs of inflammation in the eye have disappeared. In addition, intravenous injections of typhoid vaccine should be instituted.

In keratoconus, the transplant often has a tendency to protrude during the postoperative course. This must not be considered a complication, but almost the normal cicatrization of grafts in keratoconus.

In order to obtain a more normal curvature, it is necessary to maintain a pressure dressing which is started the fifteenth

postoperative day and is continued until the fifth or sixth postoperative week. The first week of pressure dressing, the pressure must not be too strong; otherwise the incision may reopen, leading to development of anterior synechiae. However, in the following weeks the pressure can be substantially increased until as much pressure as possible can be applied by the fourth week. In order to apply adequate pressure, it is not necessary to use bandages of the roll type around the head, which sometimes make the patient very uncomfortable. I have found that the dressing illustrated in Figure 14 gives adequate pressure for about 2 days. For this pressure dressing to be effective, it should be carried out in the following fashion:

The ocular area within the rim of the orbit should be well packed with gauze and 2 rows of strips of adhesive applied, pressure being exerted from the first strip of adhesive, with the second row reinforcing the pressure obtained in the first row. The cheek and front ends of the adhesive should be held tightly in position by other strips of adhesive in order to prevent curling of the edges and loosening of the bandage.

With this pressure dressing, a good curvature of the cornea may already have been obtained by the fourth postoperative week. In such cases, the pressure dressing has to be applied thereafter with care, so as not to produce too much pressure and to avoid excessive flattening of the cornea, which might result in high hyperopia, astigmatism or even contact of the cornea with the iris and development of anterior synechiae.

The same type of pressure bandage described above has been found useful in the treatment of acute ectasia or hydrops, which so frequently occurs in keratoconus. The bandage is continued until the ectasia has entirely disappeared and the eye has become quiet (Figure 15).

Sometimes in keratoconus, with more frequency than in all other corneal deformities treated by corneal transplantation, the transplant remains protruding for over 4 weeks in spite of the pressure dressing, apparently due to the thinness of the

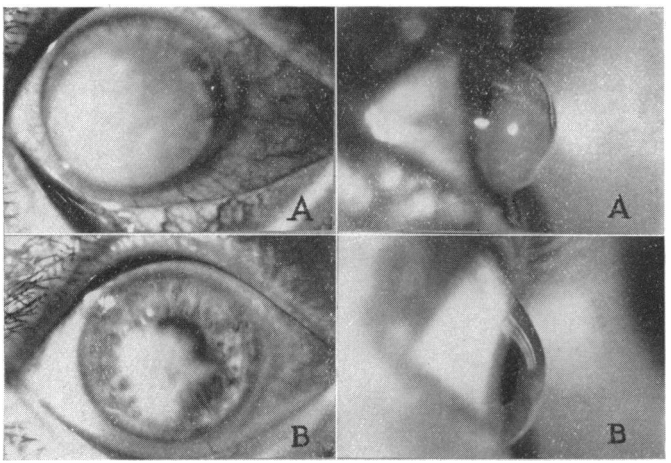

Fig. 15.—Photographs of acute ectasia before (A) and after (B) application of pressure dressing.

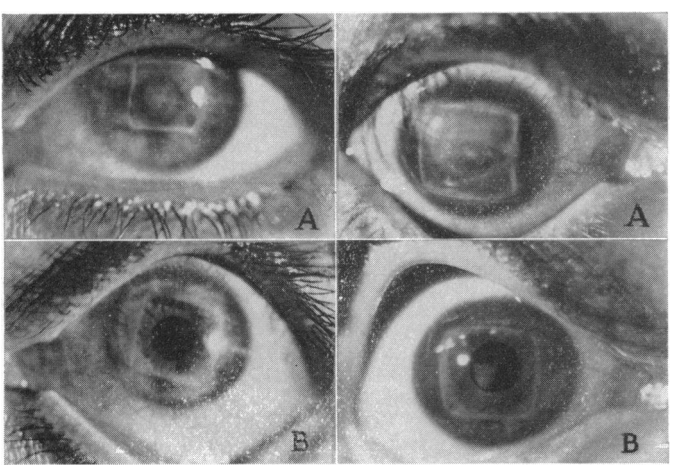

Fig. 16.—Photographs of (A) cloudy transplants, (B) clear retransplants.

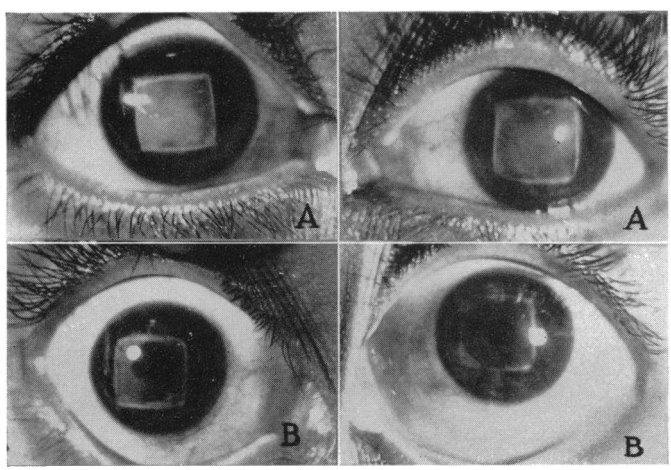

Fig. 17.—Photographs showing cloudy transplants (A), after uveitis, which finally cleared up (B).

edge in the recipient cornea. Under these circumstances, the nutrition of the graft is impaired, often leading to cloudiness of the graft and occasionally to the development of corneal edema with bullous degeneration of the epithelium. This corneal edema is generally improved by applications of sodium chloride hypertonic 15% jelly, from 3 to 6 times daily. If the bullous degeneration of the epithelium fails to clear up with this conservative treatment, it will be advisable to scrape the epithelium and treat the scraped area and about 1 or 2 mm. of the surrounding cornea by light superficial electro-dessication. With this treatment, the bullous degeneration of the epithelium will disappear, but at the expense of permanent superficial cloudiness of the graft, which will require further surgery if there is enough cloudiness to greatly impair vision.

If, in spite of the applications of pressure dressing, the transplant should cicatrize with protrusion or uneven curvature, resulting in pronounced myopia and astigmatism, although preserving permanent transparency, the success of the operation will be defeated for visual purposes. Contact lenses may occasionally help this situation; if not, a second transplant, preferably larger than the previous one, will be necessary.

I have mentioned here only those complications most likely to occur in cases operated on for keratoconus. Other complications observed in corneal transplantation have been discussed in another article.[45]

RETRANSPLANTS

If the transplant should become cloudy, either on account of protrusion during the postoperative recovery, anterior synechiae successfully treated by surgery, or because of postoperative uveitis, the eye will still be favorable for another corneal transplantation if the surrounding cornea has not developed complications such as opacity or vascularization. When the eye is still favorable for further surgery, at least

6 months but preferably a year should elapse before the second transplant to provide time for the eye to become entirely quiet. Figure 16 shows transplants which remained clear after a second operation, where the previous one had resulted in a cloudy graft.

CLOUDINESS OF THE GRAFT

In cases of delayed cicatrization due to protrusion or following uveitis, the transplant may become cloudy to the point where the patient can see only to count fingers or distinguish hand motion at 1 foot, but sometimes these transplants clear up in a few months to such an extent that it is no longer possible to detect any cloudiness of the graft, with vision improving sometimes to 20/30 and even 20/20 (Figure 17). This clearing-up process can be expected to take place within a year after the operation. Any opacity remaining after the first year may be considered as permanent, requiring further surgery if vision remains poor.

In some cases of extremely pronounced keratoconus, approaching keratoglobus in appearance, I have tried to flatten the cornea by superficial electro-dessication in order to obtain more normal curvature and render the eye more favorable for keratoplasty. So far, however, all the cases thus treated have resulted in failure.

COMMENT

I am convinced that the partial penetrating type of keratoplasty is the treatment of choice for advanced keratoconus. When a suitable technique is used, the percentage of permanently, greatly improved vision should be high, from 75% to 90%. The percentage of success will, of course, be higher the more experienced and skilled the surgeon. The surgeon less experienced in keratoplasty, given the proper selection of cases and technique, and taking into consideration the observations here presented, may also expect a high percentage of satisfactory results.

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DISCUSSION

DR. RALPH O. RYCHENER, Memphis, Tenn.: Dr. Castroviejo mentioned that in cases of acute hydrops pressure bandages are very effective in reducing the hydrops. One cannot draw many conclusions from a few scattered cases, but it so happens that I have had 3 such cases to care for, in all of whom I did repeated daily paracenteses along with pressure bandages, which apparently lessened the amount of corneal opacification that they received as a final result, because in all 3 of those cases, one of whom had previously worn contact lenses, we were able to fit them satisfactorily with ordinary lenses so that their vision was acceptable. We obtained from 20/70 to 20/40 and good reading vision. I believe repeated paracenteses were an important part of the treatment.

DR. CONRAD BERENS, New York City: I want to ask only one question. I performed a square transplant in one eye of a patient with keratoconus, and he had a good deal of anterior protrusion of the graft. In this particular case we used the Castroviejo technic but with 2 criss-cross sutures, and it took about 6 months for the graft to clear. He had a complication—a very serious coliform infection which we were unable to control until we gave him streptomycin. That seemed to cure the conjunctival and associated nasal infection. In the second eye we thought we would try a round graft, and that we would sew the graft in with 4 sutures. We did this and had an absolutely flat cornea immediately. No complications developed for 2 weeks and we expected an excellent result because the donor cornea was better and the anterior surface of the graft did not protrude. Unfortunately, the entire posterior half of the graft gradually became opaque, and the density of the opacity increased for 4 months when it began to clear at the periphery. It is

now 7 months after operation and the center of the graft has not cleared. The question from these 2 experiences is whether it is not better in thin corneas for the graft to extend anteriorly and posteriorly. Also from this question, whether we should sew grafts in thin corneas so firmly that the anterior surface of the graft is flush with the anterior surface of the cornea or whether we should not put the sutures in rather loosely so that the cornea may be permitted to protrude forward and backward. I hope Dr. Castroviejo will be able to answer this question.

DR. RAMÓN CASTROVIEJO, closing: Dr. Rychener advocates paracentesis for the treatment of cases of keratoconus which have developed acute hydrops. I have successfully treated this type of case by the application of pressure dressing alone. It seems logical to accept the most conservative of the two methods accomplishing the same end. However, the best treatment for acute hydrops will be established when sufficient statistical data are available to compare both the surgical and the nonsurgical methods.

Dr. Berens has had a case of square transplant with a tendency to protrusion at the corners. This tendency to protrusion can be observed both in the square as well as in the round transplant. For the purpose of comparison, I operated on two brothers and two sisters affected with corneal dystrophy, using a round transplant on one brother and one sister and a square one in the other brother and other sister. Of these four cases, one square and one round transplant protruded, necessitating prolonged pressure dressing before flattening of the protruding edges of the grafts was obtained. During the operation, when I find that the transplant has a tendency to become displaced, I use direct suturing of the borders of the cornea of the host with those of the donor, placing at least four sutures either in the corners of the square graft or at the ends of two perpendicular diameters in the circular grafts. I do not like the use of this border-to-border suturing as a routine procedure because of the added trauma to the graft which may lead to infection. This point has been discussed in the article, but I have not been able to develop it in this presentation because of limitation of time. As long as there is protrusion of the graft, its nutrition is impaired with subsequent tendency to nebulosity or opacification. When they are finally well-rooted and at the same level as the cornea of the host, some of these nebulous grafts eventually clear up, requiring sometimes between six months and a year until completion of this clearing-up process.