RETINITIS PIGMENTOSA* **A NEW THERAPEUTIC APPROACH**

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THE aetiology of various types of retinal and macular degeneration is unsatisfactory and controversial, and little treatment can be given. Many authors have attributed these changes to disturbance of the choriocapillaris and local angiosclerosis of the choroid, and the retinal lesions have been regarded as secondary to these changes (Verhoeff and Grossman, 1937; Brown, 1940). Certain areas, *i.e.* the macular, equatorial, and peripheral regions, seem to be more prone to degeneration.

An analysis of the vascular supply of these areas reveals that the macular region is supplied primarily from the choroid, and that the retinal vessels do not play an important role in its nourishment. Dartnall and Thomson (1949) showed histologically and subjectively that the macula does not have retinal blood vessels but that this is compensated for by the abundance of choriocapillaries. In the equatorial region there is an anastomosis between the posterior and anterior ciliary circulation and this is probably the least resistant part of the vascular system of the choroid. The peripheral region is poorly supplied by retinal blood vessels. The peripheral margin of the retinal vascular system is formed by wide-calibre capillary arches joining the termination of the arteries of the veins. The frequent occurrence of cystoid degeneration in this region is probably due to its relatively poor blood supply which leads to atrophic retinal changes. The many conditions have been attributed primarily to choroidal sclerosis, disciform degeneration of the macula, senile macular degeneration, myopic degeneration of the fundus, retinitis pigmentosa, and circinate retinopathy.

The management of these conditions is often difficult. Vasodilators have been recommended but have given equivocal results. Filatov and Verbitska (1946) recommended tissue therapy for retinal pigmentosa. Campos (1956) recommended the implantation of strips of lateral and/or medial rectus close to the choroid after a lamellar scleral resection, aiming at the revascularization of the choroid. Burnside (1956) demonstrated experimentally on monkeys that a simple advancement of inferior oblique muscle strip over the macular area improved the vascular supply of the macula by establishing a collateral circulation with the choroid. Burnside (1958) advocated this method for the treatment of senile macular degeneration. Tamesis (1960) modified the technique of Burnside and implanted the muscle strip in the supra-choroidal space over the macula; he claimed good results in cases of

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retinitis pigmentosa, choroidal degeneration, high myopia, and angiospastic retinopathies. These developments have stimulated our interest in this subject and we have attempted to give some relief to patients suffering from retinitis pigmentosa by passing a slip from the lateral rectus through a 4-mm. long tunnel under the sclera and re-attaching it at its insertion. This allows the muscle to come into direct contact with the choroid in the suprachoroidal space.

Material and Methods

Fifteen eyes in an advanced stage of degeneration were selected for this operation. The following examinations were carried out in all cases:

- (1) Slit-lamp examination of the affected eye.
- (2) Ophthalmoscopic record of the fundus.
- (3) Measurement of the visual fields both central and peripheral.
- (4) Measurement of muscular balance on Maddox rod and Maddox wing.

The post-operative follow-up included the examination of the fundus every alternate day for the first 2 weeks, then every week for 3 months, and every month for one year. The central and peripheral fields were checked every 15 days for 3 months, and then every month for one year. The muscular balance was checked to assess any weakness of the lateral rectus due to the implantation in suprachoroidal space.

Premedication.—The patient is given 1.5 g. Nembutal and 25 mg. chloropromazine the night before the operation. 90 minutes before the operation a capsule of 1.5 g. Nembutal is given orally. 45 minutes before the operation the patient receives an intramuscular injection of 50 mg. Pethidine and 25 mg. Largactil.

Anaesthesia.—Surface, facial block, and retrobulbar infiltration anaesthesia is carried out with Lignocaine hydrochloride, containing 1 part of adrenaline to 4 parts of Lignocaine injected into the muscle belly as far back as possible.

Technique (Agarwal's Operation).—The lateral rectus muscle is exposed (Fig. 1) at its insertion. The muscle is freed from the tenon sheath and its expansion. Double-arm sutures are passed through the lower half of the lateral rectus which is then detached from its insertion (Fig. 2). The detached slip of lateral rectus is separated from the upper slip for about 15 mm.

FIG. 1.—The lateral rectus is exposed.

FIG. 2.—Scleral incision to show the formation of the scleral tunnel and the separation of the lower half of the lateral rectus.

The sclera in the region of the equator, *i.e.* about 10 to 15 mm. away from the limbus, is cleaned and two marks are put at 11 and 15 mm. away from limbus. Two 4-mm. vertical cuts are made on these marked points, and the incision is continued till the choroid shines through. With careful dissection, the remaining layers of sclera are cut so that the suprachoroidal space is exposed. Agarwal's muscle spatula (Fig. 3) is passed to separate the choroid from the sclera in order to make a 4-mm. tunnel under the sclera. The two eyes of the spatula are threaded with sutures attached to the detached slip of lateral rectus. The spatula is passed through the tunnel entering at the 15-mm. mark and coming out at the 11-mm. mark.

FIG. 3.—Agarwal's muscle spatula.

The spatula is removed and the muscle is pulled through the tunnel with the help of the sutures already passed. The muscle slips through the tunnel without much difficulty and is again attached to its original insertion (Fig. 4).

FIG. 4.—The lower half of the lateral rectus is passed through the scleral tunnel over the choroid and reinserted in its original position. By this means a portion is implanted in the suprachoroidal space.

The conjunctiva and Tenon's capsule are stitched and both eyes are bandaged for 24 hours.

Observations

The patients were followed for from 3 to 13 months.

(1) *Fundus Examination*.—Direct ophthalmoscopic examination of the area showed hyperaemia and haemorrhage. In some cases the area appeared white, and was surrounded by hyperaemia which was probably due to damage to the choroid during the operation. In two cases only we observed new vessel formation in the area of the damaged choroid.

(2) *Visual Improvement*.—Nine out of fifteen eyes operated on showed visual improvement varying from one to three lines on Snellen's chart for

distance. The other cases showed no improvement in vision but there was no further deterioration.

(3) *Subjective Changes*.—Almost every patient felt able to see and work better. Two patients with retinitis pigmentosa reported that they could cross the road with more confidence and without help which they could not do before the operation. One woman reported that she could write letters and that her vision was more steady.

(4) *Visual Fields*.—The peripheral and central fields showed no appreciable changes.

(5) *Muscle Balance*.—No imbalance occurred in any of the operated cases, the pre-operative and post-operative readings on the Maddox wing or the Maddox tangent scale being the same.

Complications

In two cases a vitreous bead came out; it did not materially affect the post-operative result in one, but the other eye was lost because of severe haemorrhage in the vitreous leading to haemorrhagic endophthalmitis.

Discussion

The management of retinal degeneration is both difficult and unrewarding. Several types of treatment, both medical and surgical, have been tried but with little success. Burnside (1956) produced evidence to show that the implantation of inferior oblique muscle in the suprachoroidal space led to an increase in retinal circulation in that area. Campos (1956) reported beneficial results in retinitis pigmentosa by interscleral implantation of extra-ocular muscle. Agarwal (1961) devised a new technique of implantation of the horizontal recti in the suprachoroidal space, passing the muscle under the bridge of the sclera in the equatorial region. An attempt has been made to evaluate the results of Agarwal's implantation in fifteen eyes of twelve patients with retinitis pigmentosa. Fundus examination revealed hyperaemia and congestion in the operated area in twelve eyes. One eye was lost due to severe vitreous haemorrhage and in two eyes an atrophic patch was seen in the operated area.

In the cases so treated there has been no further deterioration either in the visual acuity or in the field of vision. A large number of cases showed some improvement in acuity (one to three lines on Snellen's chart) and almost all claimed subjective improvement. These findings are encouraging and hold out hope for the future, though it is as yet premature to assess the results, as spontaneous remissions are not uncommon. The results do appear, however, to warrant further trial of this therapeutic procedure.

As regards the mechanism of action of this operation, Burnside (1956) thought that the muscle implantation, besides giving support to the retina, improved the collateral circulation and the nutrition of the tissues. He also

demonstrated experimentally the establishment of neovascularization. Tamesis (1960) thought that the implanted muscle either made good the deficiency of enzymes in the remaining living rods and cones, or helped in the drainage of stagnant fluids or metabolites. We feel that the operation has a triple action: it establishes a collateral circulation, helps in the metabolic processes of the retina by better diffusion though the close proximity of the vascular muscular tissue, and acts as a biogenic stimulator. In view of the beneficial effects of horizontal muscle implantation it is proposed to treat a few cases by implanting all four recti muscles.

Summary

(1) The technique of lateral horizontal muscle implantation (Agarwal's operation) is described.

(2) Fifteen eyes from twelve cases of retinitis pigmentosa were subjected to this operation and the patients were followed-up for from 3 to 15 months.

(3) Fourteen eyes from eleven cases showed subjective improvement, and nine of these showed definite visual improvement.

(4) It is thought that the operation is promising, but no definite claim is made in view of the short follow-up.

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