THE SURGICAL TREATMENT OF GLAUCOMA COMPLICATING CONGENITAL ANIRIDIA

BY *Eugene M. Blake,* M.D.

IT IS NOT THE INTENTION of this presentation to discuss in detail the embryology, histology, or genetics of congenital aniridia, or irideremia. These phases of the subject have been ably and recently covered by Callahan (1) and Pincus (2). Congenital lack of the iris is always represented by a narrow rim of tissue, is always bilateral, and results from defects in the development of the mesodermal and epidermal layers of the iris. It shows a marked tendency to hereditary transmission and occurs usually as a dominant characteristic. Aniridia is generally complicated by other structural defects of the eye, such as cataract, ectopia lentis, nystagmus, corneal opacities, aplasia of the macula, and especially by glaucoma.

It is impossible to determine exactly how frequently congenital aniridia occurs, perhaps in 0.04 percent of cases. Judging, however, from the small number of cases seen by clinicians of wide experience, the disease is comparatively rare. What proportion of these patients develop glaucoma is even more difficult to determine, but it is certainly a fairly frequent complication of an already unpleasant picture. According to E. Treacher Collins (3):

> Regarding cases of Aniridia from the clinical standpoint alone, we should be inclined to think that it was impossible for them to become subjects of glaucoma; for we fail to see how, when the iris is absent, the filtration area can be blocked. But the microscope seems to show that they are really predisposed to glaucoma, because between the rudimentary iris and the ligamentum pectinatum there are abnormal adhesions. This is so in the eyes examined by Pagenstecker and Rindfleisch and in those that I have recorded.

It would appear that the increased intraocular pressure is not present at birth or one would find enlargement of the globe com-

paratively early in life. As a matter of fact, buphthalmus is rarely seen. Therefore there must be a building up of pressure for a considerable time before we observe the evidences of glaucoma.

The treatment of congenital aniridia with glaucoma presents a difficult and unsatisfactory problem. Belmont quotes Professor Ernst Fuchs (4) who once said that all of these cases are hopeless. While several writers have reported lowering of the intraocular pressure through the use of miotics, the majority have failed to obtain any lasting benefit from these drugs. When reduction of tension does occur, it is probably due to the pull upon the scleral spur or to the effect upon the blood vessels of the uveal tract. In view of the generally unsatisfactory results of miotic medication alone, surgery is usually indicated when the tension becomes definitely elevated. It may be that a combination of miotics and surgery will yield better results.

Many textbooks on ophthalmology dismiss the subject of aniridia with a few scant words and little or no recommendation as to treatment. Because the writer's surgical experience in congenital aniridia complicated by glaucoma is limited to a single patient and because of the diverse opinions expressed by the relatively small number of reports available, it was thought desirable to solicit a sufficiently large number of surgeons, requesting information as to their experience in operations upon these patients. The results of this inquiry requested of some eighty-seven ophthalmologists, mostly members of this society, revealed several significant facts. In some instances, the replies were difficult to tabulate—for instance, the surgeon would reply that he had performed cyclodialysis two or three times on one eye and then had done diathermy, or a sclerectomy combined with cyclodialysis, etc. In assembling the facts here presented, I have endeavored to make clear the procedure which seemed to have yielded the results tabulated. A considerable number of the surgeons replied that they have had no experience in the surgery of glaucoma complicating congenital aniridia but expressed their opinion as to what procedure would be followed if confronted with the problem. These replies seem to me to be of very great value in the consideration of treatment, since no one person admits a large experience with this rare condition.

The results of the questionnaire are tabulated below.

RESPONSE TO THE QUESTIONNAIRE

Number of replies received	87
Number of replies, "No experience"	46
Number of surgeons having operated upon aniridia	41

TYPES OF OPERATION AND RESULTS

	Controlled	*Not Controlled*
Trephine	4	10
Cyclodialysis	13	15
Cyclodiathermy	8	8
Sclerectomy	5	1
Goniotomy	11	1
Extraction of Cataract	1	

PREFERENCES EXPRESSED AS TO TYPE OF SURGERY, IF REQUIRED

Trephine	2
Cyclodialysis	6
Cyclodiathermy	10
Goniotomy or Goniopuncture	5

In spite of the small number of cases of glaucoma operated upon, and in view of the suggestions for treatment when and if surgery were required, it would appear that cyclodiathermy, or cyclo-electrolysis, is to be preferred. In the small series of sclerectomies, possibly with excision of the iris root, this procedure with five good results against one failure is significant. Certainly cyclodiathermy is becoming an increasingly popular operation and modifications in technic are making for better results. Placing the diathermy points farther back of the limbus, together with a longer period of contact with the applicator, is yielding better reduction of tension. Possibly Weve's method of attempting to destroy the ciliary nerves still more posteriorly will present advantages.

An analysis of these rather scanty figures shows, first of all, that the surgical experience in the treatment of glaucoma complicating congenital aniridia is not extensive. No one operator has seen or treated a sufficient number of cases to have a convincing opinion as to the best method. Secondly, the tabulation shows that nearly half of the cases were unsuccessful, whatever method was employed. It

is to be hoped that all surgeons who operate for this type of glaucoma will report their results with the method used. Only in this way shall we acquire enough information to guide physicians in making a decision as to the best procedure to follow.

CASE REPORT

I wish to describe the single case upon which I have operated for glaucoma complicating congenital aniridia, and the result of treatment. The patient was a boy aged seven weeks when first seen on May 8, 1931. He was the second offspring of healthy parents who suffered from no ocular pathology of any kind. The first-born child was a boy whom I saw at the age of six months. He was afflicted with congenital aniridia and died at the age of three years from influenza. The aniridia was not a complete ring, there being a narrow rim of iris, shaped like a horseshoe and open at the top. Three other children were born to these parents after the second child and none of them has developed any eye disease.

The patient, the second sibling, presented the picture of bilateral congenital aniridia. There were posterior polar cataracts in each eye with the surrounding lens substance transparent. No nystagmus was observed. Six months later the child followed lights and objects and the cataracts were unchanged. The fundi appeared to be normal. At five years of age the corrected vision was 15/200 in each eye. The patient moved to another state and was not seen by me until he was sixteen years old, when vision was found to be 20/140 with the correction of high myopia. The intraocular pressure was 55 mm. in each eye (Schiötz) and the field was limited to a small central area. The right eye was exotropic.

In view of the lens changes and the increased tension, it was decided to extract the right cataract. Because of marked nervousness and nystagmus, the anesthetic chosen was sodium pentothal. Following the removal of the lens in capsule, the boy was extremely excited and required three people to hold him in bed. A choroidal hemorrhage resulted, leading eventually to a shrunken globe. Various miotics, including "floropryl," were used in the attempt to reduce tension in the remaining eye, but with no effect.

Therefore, on November 8, 1947, cyclodiathermy was performed. Two subsequent diathermies were done, resulting each time in a lowered intraocular pressure. A fourth procedure, cyclo-electrolysis, was done below, resulting in tension of 19 which persisted for three years. When seen May 24, 1952, the tension was 30 mm.

The patient is now a junior in college, doing his work with the aid of a reader. In this case, cyclodiathermy, plus miotics, has proved to be

a safe procedure and one which gave a fairly satisfactory reduction of tension.

REFERENCES

1. Callahan, Alston: Aniridia, ectopia lentis, and glaucoma, Am. J. Ophth., 32: 28, 1949.
2. Pincus, M. H.: Aniridia, congenita, report of five cases; genealogy; possibilities of treatment, Arch. Ophth., 39: 60, 1948.
3. Collins, E. Treacher: Aniridia and glaucoma, Ophth. Rev., 10: 101, 1891.
4. Belmont, Owen: Congenital irideremia with secondary glaucoma, Wills Eye Hospital Reports, 4: 19, 1951.

DISCUSSION

DR. WILLIS S. KNIGHTON. Dr. Blake has posed a question which is difficult to answer because so few ophthalmologists see enough of these cases to be able to judge by their own experience. The occasional treatment, which may or may not be successful, leaves one with a feeling of frustration, because there is so little opportunity to compare results with other methods. This is especially true when surgery is involved.

The literature is scanty and discouraging. What little there is, deals mostly with pathology and heredity, with only occasional reference to suggested or actual methods of treatment when glaucoma occurs as a complication. Here the reported results vary considerably. Dennis, quoted by Pincus, said that eserine invariably controls the tension in aniridia, and Pincus added that the glaucoma can be controlled by eserine or trephine. Park Lewis and Parker Heath described lowering of the tension by pilocarpine.

As Dr. Blake has mentioned, others feel that all these cases are hopeless. The truth probably lies somewhere between the two extremes, depending upon the nature and extent of the underlying process itself.

Neber noted that the amount of iris varied from complete aniridia to a small coloboma of the iris, and Nakao and Yasutake agreed that they were only manifestations of the same process.

The angle itself in aniridia has been described as completely closed with rudimentary iris tissue adherent to the cornea in some cases and as open and free from obstruction in others.

The associated anomalies of cornea, lens, retina, and so on, also appear to vary in severity. It is more than likely that most of the good results of medication or operation were obtained in those eyes which had not suffered the full force of the pathological process.

A review of the records of the New York Eye and Ear Infirmary for the past 10 years and of the Eye Institute of the Presbyterian Hospital

for the past 20 years disclosed only 9 cases of congenital aniridia. Of these only 5 had glaucoma. Miotics were of questionable benefit in one case where the tension varied between 30 and 40 after treatment. Goniotomy was unsuccessful in another, and in a third case a tension of 35 was brought down to 27 by trephining. The other 6 cases apparently received no treatment or operation.

My own experience is limited to 2 cases. In one of these the tension was controlled with miotics. In the other a sclerectomy combined with cyclodialysis held the tension down for several months before the patient disappeared. Neither case showed severe internal involvement.

In view of the different degrees of ocular involvement that can be encountered in congenital aniridia it seems questionable whether there is any single method of choice in the management of the glaucoma that frequently occurs.

DR. M. ELLIOTT RANDOLPH. I have been fascinated by Dr. Blake's excellent paper. I recently looked up the cases in the Wilmer Institute, stimulated by three members of one family with aniridia whom I had the doubtful privilege of looking after. I found 26 such cases in 13 patients, all of whom were white. The family history showed four in one family, and of the remaining nine, only two had antecedents with aniridia. Bilateral glaucoma was present in 8 of the 13 cases. Cataracts were present in 12 of the 13. In one of these no note was made of any lens changes. The cataracts were either nuclear, anterior, posterior polar, or hypermature.

Several points interested me: first, if glaucoma was not present, the surgical outlook for cataract extraction was relatively good. Among the first cases admitted to the Wilmer Institute, trephinings were done; these were unsuccessful, and cyclodialysis was performed. As a primary procedure cyclodialysis was followed by normal tension in 5 eyes, without further surgery. When glaucoma and cataract were present, and cataract extraction was the primary procedure, 3 out of 4 eyes went into phthisis bulbi following this initial operation.

Another point noted was that in the description of each optic nerve, pallor was described but there was no mention of glaucomatous cupping. Finally, in none of our cases did aniridia occur in Negroes.

DR. WILFRED E. FRY. May I relate my experience in regard to one case of aniridia requiring surgery? During the past 15 years at the Overbrook School for the Blind the records show that 8 cases of aniridia were admitted as pupils. In all of these cases the condition was extreme and I have operated on none of them. The one I want to refer to is a woman who is now in her early sixties, who had lost one eye as the result of uveitis; presumably that eye had aniridia. The remaining eye had aniridia. The patient went along fairly well until several years

ago, when she began to develop a cataract in that eye, and with the development of cataract the tension increased. About six months ago it was necessary to remove the cataract. I was able to do an intracapsular extraction. Following the removal of the cataract, the tension, which previously had been in the neighborhood of 35 to 38, dropped to normal. The visual acuity as a result of the extraction has not been particularly good, although the patient is now able to get around by herself quite normally, feels perfectly safe in traffic, and can read with strong condensing lenses. I think it is too early to evaluate the final result as far as the extraction is concerned, but at least the early result is reasonable.

DR. W. S. ATKINSON. Dr. Blake has presented some interesting statistics on a rather rare condition. It is one which does not lend itself readily to the more common operations for glaucoma.

I would like to report a case of a lady aged 63 with congenital aniridia, glaucoma, and senile cataract.

I first saw her two years ago. Her vision was reduced to 18/200 in the right eye. The left eye had been enucleated following a cataract extraction eight years before. There was a small, scarcely visible ring of iris and general haze of the lens which was more dense centrally. The fundus was not clearly seen, but marked cupping was detected. The tension was 32 and the field quite contracted.

An extracapsular instead of intracapsular extraction was done with the thought that the posterior capsule would support the vitreous. It was hoped that removing the lens would control the tension.

The operation was followed by an uneventful recovery, with vision of 20/50. The fundus could then be seen more clearly, and there were central macular changes which probably reduced the vision somewhat. In the spring of 1952, the patient returned and said that she had had some discomfort in her eye and right side of her head. The vision was 20/60+2, the field about the same, and tension in the thirties. It had been as high as 55 and partially controlled with pilocarpine and suprarenin bitartrate 1 percent.

In April, 1952, oradiathermia was done. It consisted of ten surface diathermia applications 8 mm. from the limbus preceded by a paracentesis to control the tension. Following recovery from the operation the tension has remained normal with vision and fields about the same. It is hoped that the above procedure will continue to control the tension as well as it has done in other types of glaucoma on which it has been used.