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[*Reprinted from Transactions American Ophthalmological Society, 1908.*]

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The condition described as macular holes and holes in the retina by English writers, and as retinitis atrophicans sive rarificans centralis by Kuhnt, and as Durchlöcherung der Retina by Haab, is probably not as rarely seen in this country as one might infer from the fact that there are less than half a dozen cases by American writers on record. Until 1900, when three papers on this subject appeared in the medical journals in Germany and England, but little attention was paid to it, although, as Ogilvie has pointed out, a case similar to one of his own was published by Knapp as early as 1869. Knapp's case is, however, a doubtful one, and was misinterpreted by him. The first case about which there can be no doubt was reported to this society in 1871, by Dr. Henry B. Noyes, and published in the transactions for that year. The case is entitled "Detachment of the Retina with Laceration of the Macula Lutea." Loring has evidently seen such a case, for in his text-book of Ophthalmology (1891), in speaking of circumscribed retinitis of the outer layer, he says: a marked feature in some of these cases is the peculiar appearance which the central portion of the macula and fovea present. The latter seems to be so sharply defined that it has the appearance of a clear cut opening — as if indeed the tissue had been cut in this place with a punch. Nothing is said of a relationship to injury.

Dr. Coates of London, who has recently exhaustively reviewed the literature of this subject in the Royal London Ophthalmic Hospital Report, Vol. XVII, part I, page 69, where also a complete bibliography can be found up to 1907,[1] after quoting

[1] Since then Trietmayer (Zeitschrift für Augenheilkunde, Band XVIII, November, 1907, p. 447) has contributed three cases all due to traumatism.

Noyes' case, says: "the history of this condition is interesting on account of the frequency with which it has been rediscovered, and I have given these full extracts from Noyes' paper because there has been a tendency in literature to ascribe the first discovery to Kuhnt and Haab. But after so clear and exact a description, there can be no doubt as to where priority properly rests."

It is not my intention to review here all that has been written on this subject, and would refer any one interested in this subject to Mr. Coates' most interesting paper already mentioned. I may here, however, say that I find that there are nearly thirty-seven cases of so-called hole in the retina on record. In twenty-seven of these a blow on the eye or some injury was the supposed cause of the condition. In ten of these cases an injury could be excluded, and vascular disease, retinal hemorrhage, arteriosclerosis, albuminuria, and irido-choroiditis are given as the probable cause. Amongst these ten cases (non-traumatic) there are three in which a hole in the retina was found in both eyes.

Of all these cases, only six were observed in this country; one of traumatic origin by Noyes; one by Holden, and two non-traumatic cases by de Schweinitz. In the discussion following de Schweinitz's paper before this society in 1904, Dr. Gould mentioned that he had seen this condition, and I mentioned very briefly the case which is here reported as Case II.

My own cases are as follows:

CASE I. *Traumatic perforation of the retina.*— H. D., about 26 years of age, a well-developed man, who up to the time of the accident had had no disease of either eye, and whose vision in both eyes had been excellent, was first seen at the Newark Eye and Ear Infirmary, April 27, 1907. He stated that on July 4, 1904, he had been struck on the left eye by a base ball, and that since that time the vision of the left eye had been much impaired. Present state: the eye is E. S. 6/60 V. F. intact for white and color, a small central scotoma can be made out. Tn. The media are perfectly clear. The pupil is active. The disc

is somewhat blanched in temporal half. The vessels are of normal size. At the macula is a perfectly circular spot, about two-thirds size of disc. It is of a bright deep red color, on which are scattered some minute pigment dots and several small bright white spots. The retina around this red spot is somewhat hazy for some distance, and on the sharply defined margin of the red spot are five or six small bright yellowish-white dots, which appear to be situated in or on the retina. The floor of the deep red spot is on a somewhat deeper level than the grayish sharply defined margin, and is best seen with a — 1 D glass. It gives one the impression of being cupped, and looks as if a piece of the retina had been cut out with a trephine. The opacity of the retina is greatest at the margin of the hole and gradually merges off into the normal retina. Above this hole the pigment of the retinal epithelium is somewhat irregularly distributed, and all over this region of the retina are scattered very fine crystalline particles which sparkle as the light from the mirror strikes them. Otherwise the fundus is normal.

The right eye was normal in every way. The patient was seen again thirteen months later, and the condition found at first examination was unaltered.

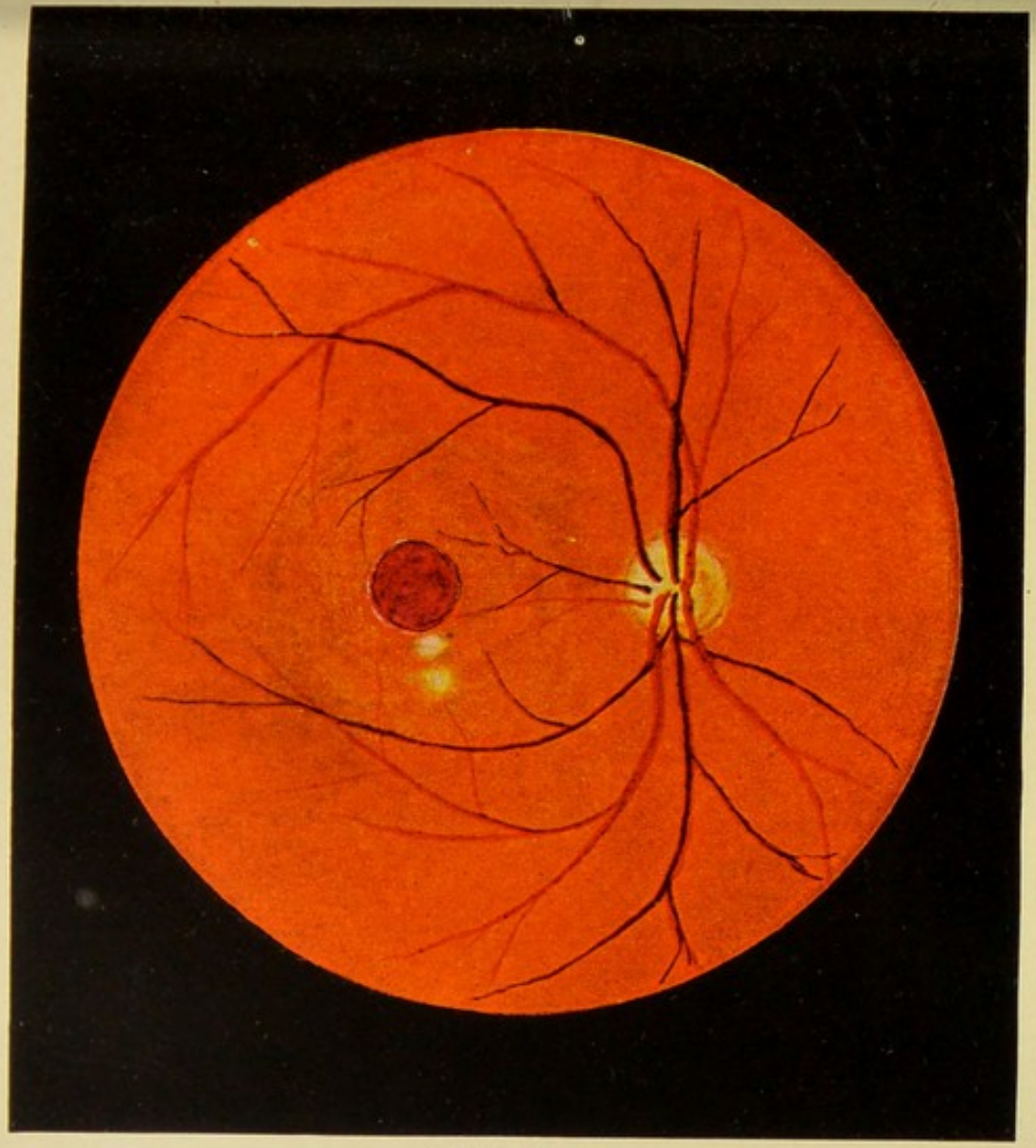
CASE 2. *Macular Hole not the result of injury.*— Miss R. M., 26 years of age, is a well developed and healthy looking woman, whose parents are living and in good health, and whose brothers and sisters are all in good health and without disease of their eyes. She has never been seriously ill, denies syphilis, and her eyes have at no time been inflamed or painful. She was seen by me for the first time in July, 1904, and came to consult me about her left eye, the sight of which had been defective for some time. She could not tell how long the sight had been impaired, but was certain that the eye had never received a blow or other injury. The right eye was as good as ever. The examination of the right eye revealed a mixed astigmatism with S. 5/6. The fundus of the eye was normal. The left eye was practically blind. She could see movements of hands close to the eye. The

exterior parts of the eye were entirely normal. The iris was of same color as that of right eye and of normal texture. The pupil was active and of same size as that of the healthy eye. The lens and the vitreous were perfectly clear. The disc was rather pale in its temporal half, and on this was an almost black spot of oval form; two other pigment spots were situated just outside of its temporal margin in the choroid. The retina was perfectly transparent, except in the region of the macula where it was decidedly hazy. The haziness was most marked around the central area which was of a deep red color, and gradually faded into the normal. The central portion of the macular region was occupied by a bright deep red spot in size about one-half diameter of the disc; it was circular in shape and was sharply defined in outline. The spot was not of uniformly red color, but on it or in it were many small yellowish-white dots. This deep red region appeared to be situated slightly deeper than the opaque retina surrounding it. The retinal vessels were of normal caliber, and there were no other fundus changes. I prescribed calomel in small doses, which she took for several months, but without the slightest benefit to the eye. Nearly three years after her first visit to me, I had an opportunity to examine the eyes again and found them in about the same condition. The only change I could discover was that the pigment spot on the disc had apparently sunken into the disc, giving it the appearance of a cavity with a dark colored bottom.[1]

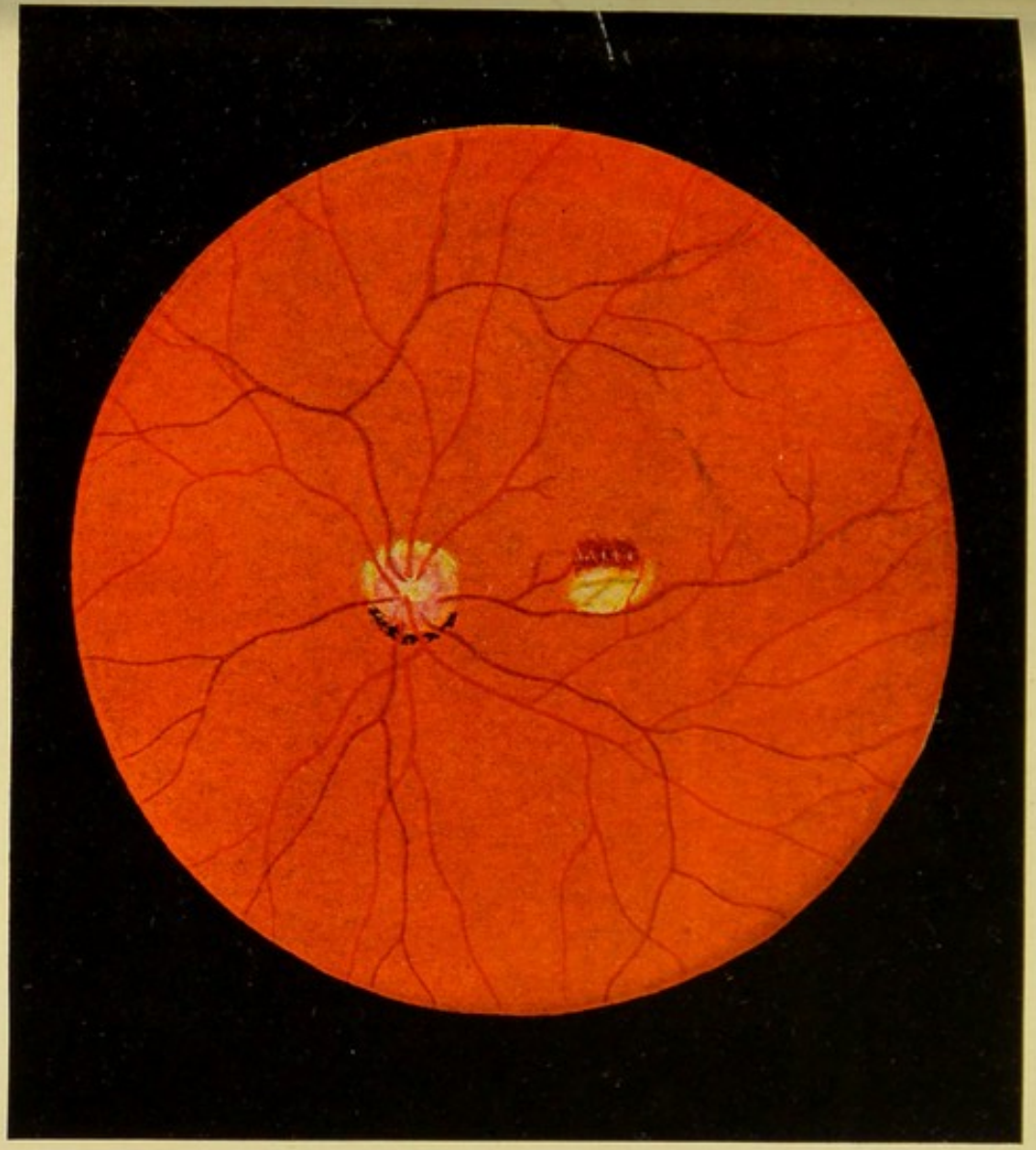
CASE 3.— J. K. is 32 years of age, is a well developed muscular man, of florid complexion, by occupation a laborer, employed by a telegraph company in putting up telegraph poles. Family history negative. He says that he has never been ill and is now in good health. He denies having had syphilis. He is a very moderate drinker of alcoholic beverages, but chews tobacco excessively. His urine is free from albumin and sugar. He does

[1] Since this paper was read, a case similar to this one has been published by Dr. W. Reis in Zeitschrift für Augenheilkunde, June, 1908, p. 505. In both cases there was, in addition to the hole at the macula, a cavity with dark bottom in temporal half of disc. In my case there were also two pigment spots in choroid near margin of disc. The lines radiating from the macular hole were not present in my case.

not remember that either of his eyes has ever been red or painful. Sixteen years ago, the house in which he lived at the time, was struck by lightning and a fragment of wood struck his face, cutting the upper lid of his right eye. He thinks that since then the vision of his right eye is impaired. No scar could be found on the lids of the right eye. His left eye has always been good, it is slightly myopic S. 6/6. Examination of the right eye revealed that it was E.; vision was reduced to counting fingers at a few feet. The movements of the globe were in no way impeded, and the eyelids were entirely normal, the globe itself was free from injection. The cornea was normal, the anterior chamber was of normal dimensions, the iris was of normal color and texture. The pupil was of same size as that of the left eye and active. The lens and vitreous were perfectly transparent. The optic disc was somewhat whiter than normal, its outline was well defined, and the retinal vessels were of normal size. The retina is transparent everywhere except in the region of the macula, where it is somewhat opaque and looks like a rigid membrane with a somewhat uneven but polished surface. It may be compared to polished milk-glass. The opacity gradually increases from periphery to center. The size of the opaque area is equal to about three diameters of the disc. On it are numerous very minute transparent particles, which sparkle like crystals as the light from the mirror strikes them on a certain angle. The opaque portion of the retina is best seen with a + 2 D glass, while the disc is best seen without any glass. In the center of the macular region there is a sharply defined space of about three-fourths of the breadth of the disc, slightly oval, the longest diameter vertically, in which the retina appears to be absent. It looks precisely like a hole in the retina made with a sharp punch. The bottom of this hole is of a bright deep red color with yellowish-white dots in it. The color is that of a raspberry. The margins of the hole are very sharply defined, and are best seen with a + 2 D glass, while the bottom is best seen without any glass. Adjoining the lower margin of this hole



Right Eye.



Left Eye.

is a yellowish-white spot, round in form and about one-fourth of the disc's diameter, which shines indistinctly through the retina, and another spot of same color and a little larger in size is situated a short distance below the other one. Neither of these spots is surrounded by pigment. The retinal vessels are veiled on this translucent part of the retina, but can be followed up to the hole, but not over it. The *left* eye is apparently normal, except for the changes to be described. The disc is about normal color. The sclerotic ring is broadened above and below, giving the disc apparently an oval shape; above there is no pigment, but a little below it has a fringe of dark pigment. The retinal vessels are of normal size. In the region of the macula there is an area, somewhat oblong in shape, roughly of the size of the disc, in which not only the retinal pigment, but also the stroma pigment of the choroid, is absent. The white sclerotic is covered by a few choroidal vessels, dividing it in three white triangular spaces. Adjoining the upper part of this rarefied space is an area somewhat square in shape which is of a bright deeply red color, in which there are yellowish-white dots arranged in lines running vertically. The color of this area is very much like that of the bottom of the hole in the other eye. The retina of this entire region appears perfectly transparent, and the retinal vessels can be traced to the region of the fovea lutea. There is no collection of pigment to be seen at the margin of the light colored area or in its neighborhood. This region, like the disc, is best seen without any glass behind the mirror. Repeated and very careful examination of the visual field showed a very small central scotoma for form and color. The light sense of the left eye was much reduced. With a smoked glass before his eyes he could see only 6/60, while I could see with same glass before my eyes 6/8. On questioning about his vision at night, he admitted that he has difficulty in getting around at night, and much more so at present than in former years.

The patient has been under my observation for the last sixteen months, and at this time the picture in both eyes is about the

same as when first seen. I saw the patient about once in every fortnight and examined his eyes under homatropine most thoroughly. With regard to the right eye, I can say positively that in the region around the hole, the surface of the retina was most distinctly seen with a + 2 D glass, while the floor of the hole was like the disc, best seen without any glass. The retina was therefore either thickened in this region 2/3 mm. or else was pushed forward by a plastic exudate behind it to that extent. There were no folds in the retina and no other signs of detachment. The two light-colored spots seen below the hole did not impress me as being opacities in the retina itself, but rather as circumscribed exudates under the retina, or as perhaps spots of atrophy in the choroid similar to the large patch of choroidal atrophy in the left eye.

In the left eye there were seen slight evidences of a former choroiditis around the disc. The bright deep red area situated above the patch of atrophy of the choroid, seemed to me almost identical in color with the color of the floor of the hole in the right eye. I assume that here the retina was thinner, more transparent than in other parts, so that the pigment epithelium was visible here. In the light-colored area below this, not only the pigment of the retinal epithelium, but also the stroma pigment of the choroid, had disappeared. There were no collections of pigment to be seen anywhere around the atrophic region, nor, indeed, anywhere in the ophthalmoscopic field, except just below the lower margin of the disc. That the deep red area was not an extravasation of blood is quite certain, as it did not change in appearance in the least during the period the eye has been under observation, and moreover it did not have the uniform red color of a blood extravasation. The retina over both the deep red and the light-colored areas was perfectly transparent, and the retinal vessels could be distinctly seen in it.

We have therefore two very dissimilar pictures of the eye-grounds of this patient. In the right eye the picture does not differ from that described by Noyes and others since, under the

name of a hole in the retina or macular holes. In the left eye the choroid is in a state of atrophy in the macular region and the retina is present, but apparently much thinner over a small area, just above the atrophic spot in the choroid. From the history of the case it seems probable that the injury to the right eye 16 years before he came under observation, is responsible for its present condition. It is not impossible also that the left eye received a blow at the same time that the right eye was injured, although the patient does not remember it. In fact the patient was not aware the left eye was defective before he sought my advice. It must therefore remain undecided whether the changes in the left eye are due to traumatism or disease. It may possibly be a congenital defect. It may also represent the result of an incomplete rupture of the choroid, but its location, its form, the absence of collections of pigment at its edge or on it, and the fact that choroidal vessels were present seems to me to make this impossible. To me it looks much more like an atrophic area following a circumscribed inflammation of the choroid. An œdema of the retina caused by the choroiditis would perhaps also account for the atrophic area of the retina seen just above the atrophic area of the choroid. I have been unable to find amongst the recorded cases, one in which a hole in the retina was present in one eye and choroidal atrophy in the macular region in the other eye. There are several cases on record in which a hole in the retina in the macular region was present in both eyes, but in all of these it arose spontaneously. There is one case on record in which the two eyes of a man received blows at different times and in which a macular hole was found in each eye, some time after the injuries.

I venture the suggestion that in my case a choroiditis attacked both eyes some time in early life, and that the disease of the choroid caused the change in the retina in the right eye, and the so-called hole in the macula. I look upon the two light-colored spots below the hole as patches of atrophy of the choroid shining through the cloudy retina. In the left eye the region immediately

adjoining the disc shows evidences of a former inflammation of the choroid, and the large atrophic patch is doubtless also the result of such an inflammation. Kuhnt has pictured a case of hole in the retina, which has also several light colored spots in or under the retina near the circumference of this hole, and in addition a large atrophic patch in choroid close to the disc. This case would seem to confirm the view I have suggested: that the hole in the retina is the sequence of a choroiditis.'

As regards the pathology of this condition, I can only repeat what others have written. Coates in the paper repeatedly referred to, says: Reis, in discussing his own cases and those in literature, very properly points out that since exactly the same appearance may arise after a contusion and after certain diseases, especially irido-cyclitis and retinal vascular diseases, it is necessary to presume some pathological factor common to these two conditions. He elaborates the theory already foreshadowed by Fuchs, that retinal œdema is the underlying lesion. This is opposed to Monteith Ogilvie's supposition that the hole is a tear by contracoup — by the convergence on the posterior pole of waves transmitted through the fluids of the eye, and that therefor it occurs at the moment of injury. Against this theory it may be observed that on the one hand a hole has never been observed sooner than 60 hours after the contusion (Ogilvie, Case 5). On the other hand opacity of the retina without a hole *has* been observed after injury in a case in which a hole subsequently developed (Reis, Case 3, Holden). Oedema of the retina with subsequent formation of a hole has also been observed in cases in which there has been no injury (Kuhnt, Case 3, de Schweinitz). The theory of contracoup furnishes no explanation of the cases arising in connection with iridocyclitis and retinal vascular diseases, whereas retinal œdema is not only probable in such cases but furnishes the desired factor common to them and to cases of injury (Berlin's opacity). Moreover such pathological evidence as we possess all points in the same direction. The cases of Pagenstecher and Genth, Fuchs, Mura-

kami, and v. Hippel, whether they arose from injury or from disease, had this in common, that large cystic spaces were present in the fovea and in the retina around.

The state of our knowledge from the pathological point of view may be summarized as follows: *No case has been seen ophthalmoscopically and examined microscopically.* Coates adds four cases of his own which he hopes will add something to the pathological data. In neither of these cases was the hole seen ophthalmoscopically. He summarizes his conclusions as follows: Macular holes are produced by an œdema of the retina at the posterior pole. The œdema may not be confined to the region of the fovea, but the appearance of the hole will only be produced if there is a defect at least of the inner layers of the retina. Possibly for the complete typical picture without membrane or shred, a total defect of all the layers of the retina is necessary, and that such a complete defect may arise from retinal œdema is proved by Case 2, above. The œdema may result from a contusion, in which case it is the same as the œdema which produces Berlin's opacity; or it may arise from toxines in the vitreous, the result of iridocyclitis; or from retinal vascular disease.

Rupture of the retina at the time of injury is not the cause of macular holes. This is proved: (1) by those instances which occur without injury in cases of iridocyclitis, choroido-retinitis, retinal vascular disease, albuminuric retinitis, etc. (2) by those cases in which diffuse opacity of the retina *without* hole has been observed after contusion and in which a hole has subsequently developed. (3) by Case 4 (Coates' case) in which such rupture occurred, but in which the measurements and appearances could not be brought in line with the clinical picture of a macular hole.

DISCUSSION.

Dr. Wm. Zentmayer, Philadelphia: Some 15 years ago there was brought to the Wills' Eye Hospital a colored man who had received a severe blow on the eye which was followed by

intraocular hemorrhage with greatly increased tension. That evening I was called by the resident as he feared the necessity of an enucleation of the eye on account of the severe pain. I did a paracentesis and the glaucomatous symptoms subsided. Examination of the fundus some days later showed the deepest excavation of the nerve head that I had ever seen, and a circular area at the macula about the size of the head of a pencil, the floor of which was a little darker than the surrounding chorioid. The only change that occurred later was that the central portion became granular and the lesion presented all the appearances of, what to-day is known as a "hole at the macula."

NOTE.— Since this paper was written, Dr. Alt and the writer of this paper have published in the *American Journal of Ophthalmology* for August, 1908, a case in which a hole in the macula was seen ophthalmoscopically twenty-four hours after an injury, and in which the histological examination of the eye confirmed the diagnosis.