# SPONTANEOUS CYSTS OF THE CILIARY BODY SIMULATING NEOPLASMS*

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Frequently single or multiple intra-epithelial cysts occur on the ciliary body. When these cysts are small they are undetected and unimportant clinically, but are seen merely in the routine microscopic examination of globes. When the cysts are large they may manifest themselves clinically and this is usually by way of the iris. From a clinical standpoint, therefore, they are viewed often as an iris lesion.

In the routine microscopic examination of eyes removed for various causes, it is not uncommon to see these single or multiple globular cysts in the valleys between the ciliary processes (Figures 1, 2 and 3). These cysts arise mostly from the anterior portion of the corona ciliaris and particularly at the base of the iris. Their walls are composed in part of the nonpigmented layer and in part of the pigmented layer of the ciliary epithelium. Therefore, the cysts represent really a localized separation of the 2 epithelial layers. I have found these cysts in otherwise normal eyes removed in order to adequately irradiate a malignant tumor of the sinuses as well as in eyes removed for a great variety of causes. Instances in the literature in which such cysts have been found coincidentally in eyes removed for other causes are reported by Rabitsch,[1] Ichikawa,[2] and Loewenstein and Foster.[3]

The microscopic characteristics are that the cyst wall is composed mostly of the nonpigmented epithelial layer which may be from 1 to 4 cells thick. The cysts are filled with a clear fluid which is largely evacuated in the preparation of the

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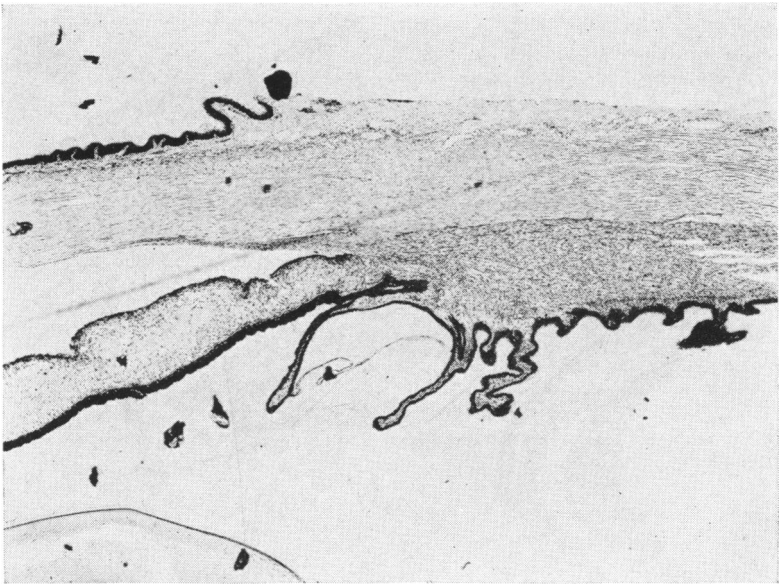

Fig. 1.—There is a cyst between 2 elongated ciliary processes. The medial wall of the cyst composed of the nonpigmented epithelium has collapsed in preparation and is seen folded and lying in the cyst space. The cyst has pushed the root of the iris forward, blocking the filtration angle. The globe shows a diffuse active choroiditis but no history is available regarding the clinical findings.

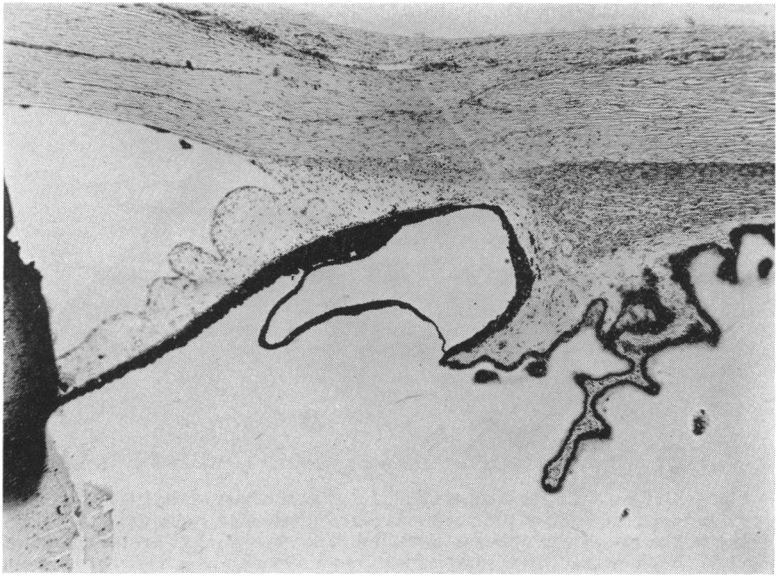

Fig. 2.—There is a cyst at the base of the iris at the junction between the iris and the ciliary body. The inner wall of the cyst is composed of pigmented epithelium which is partially collapsed in preparation. The cyst has pushed the periphery of the iris forward, blocking the filtration angle. This cyst was not detected clinically. The globe was enucleated because of secondary glaucoma, idiopathic keratitis and anterior scleritis.

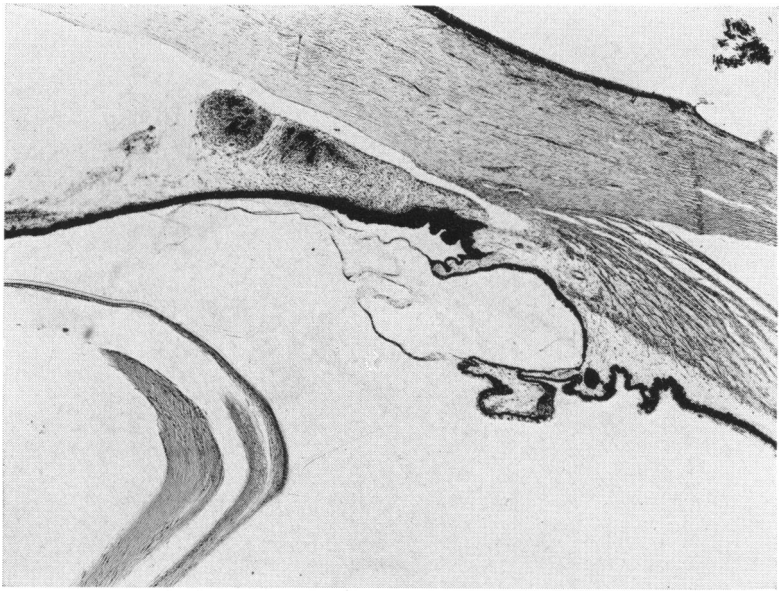

Fig. 3.—There are 3 cysts. Two of these are between ciliary processes and one at the base of the iris. The wall of the latter cyst is composed entirely of pigmented epithelium and in the cyst space are some proliferated desquamated pigmented epithelial cells. These cysts were not detected clinically. The eye was removed because of malignant melanoma of the choroid.

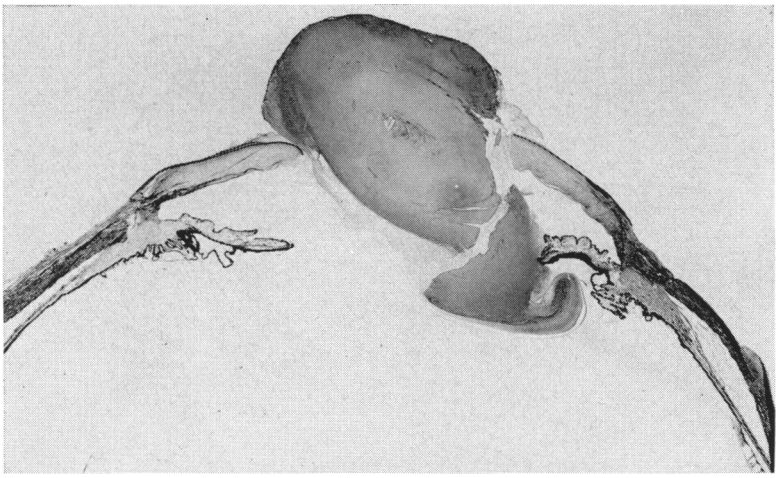

Fig. 4.—There is a large intra-epithelial cyst at the base of the iris and smaller cysts between the ciliary processes. Similar but smaller cysts are noted elsewhere in the eye in the opposite meridian. This was the left eye of a woman aged 63. Back of the iris extending from 5 to 8 o'clock was a dark brown mass which appeared to be in the ciliary body. The anterior chamber was somewhat more shallow from 3 to 4 o'clock due to the protrusion forward of the iris. The lesion failed to transilluminate light. The lesion simulated a melanoma of the iris and ciliary body. (Patient of Dr. Joseph Mandelbaun.)

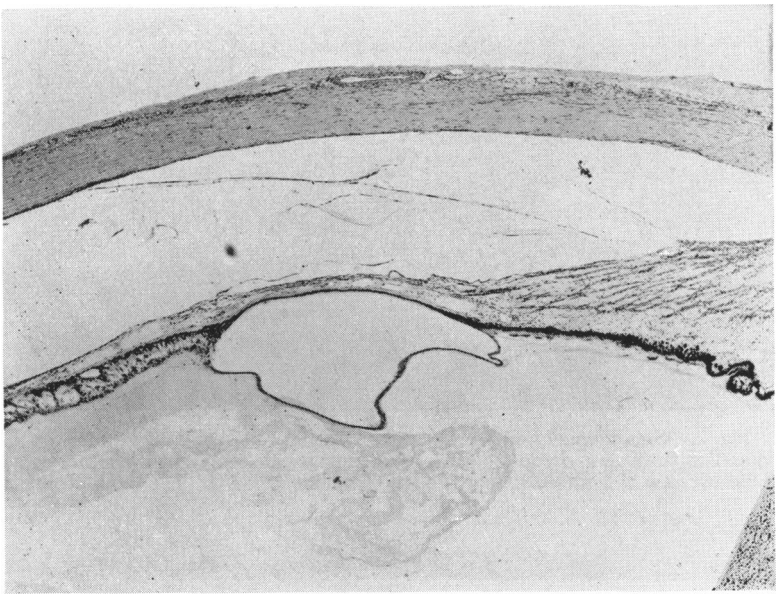

Fig. 5.—A cyst of the flat portion of the ciliary body. The inner wall is composed of the nonpigmented epithelium and the outer wall of the pigmented epithelium. The eye was removed because of glaucoma secondary to occlusion of the central vein.

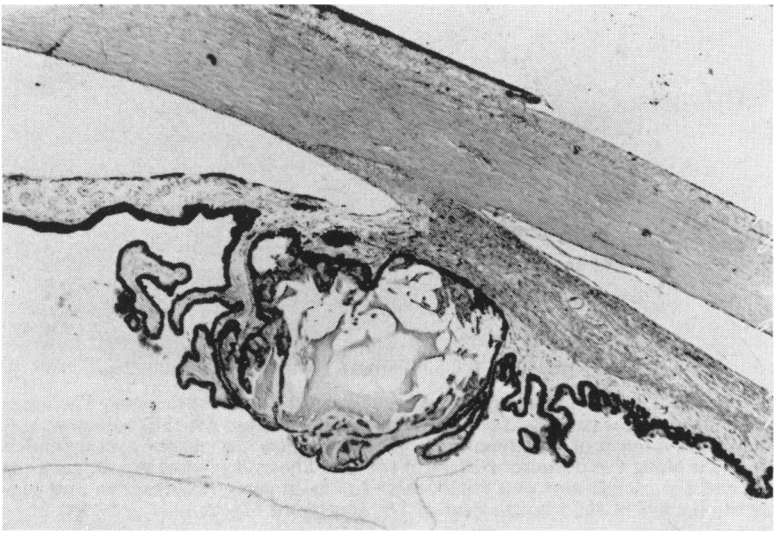

Fig. 6.—A cystic adenoma (benign epithelioma) of the ciliary body resembling a cyst. The lesion was not detected clinically. The eye was removed because of malignant melanoma of the conjunctiva.

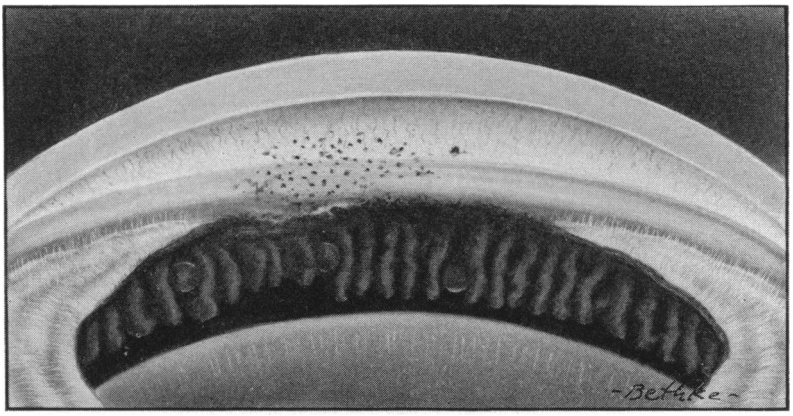

Fig. 7.—Small translucent cysts can be seen between the ciliary processes as viewed with the gonioscope through an operative coloboma. An iridectomy was done because of a melanoma of the iris. (Patient of Dr. J. H. Dunnington.)

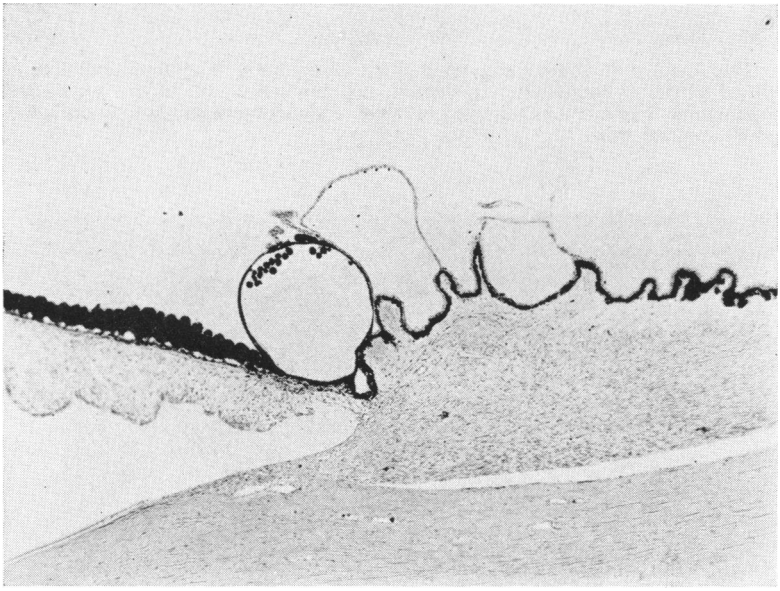

Fig. 8.—There is a large collapsed cyst between 2 ciliary processes. The inner wall of the cyst is composed of nonpigmented epithelium which is collapsed and in folds as a result of the preparation of the specimen. *In vivo* the cyst extended quite far along the posterior surface of the iris. The cyst pushed the iris forward against the cornea and as a result there has been some proliferation and pigment changes in the iris. (Patient of Dr. Raymond Meeks.)

section. Around the base of the cyst the epithelial wall undergoes a transition into the pigmented epithelium, which is more or less unchanged. The cysts may be multiple and the process, instead of being confined to just the corona ciliaris, may involve the same epithelial layers over the periphery of the iris (Figure 4) and over the pars planum of the ciliary body (Figure 5). When in the former place the entire epithelial wall is pigmented, and when in the latter place the cyst is flat instead of globular. A cystic adenoma (benign epithelioma) of the ciliary body may simulate such a cyst (Figure 6).

It is not possible to state the incidence of these cysts but in some degree they do not appear to be uncommon. Histologically, as a coincidental finding, their presence has been noted 46 times in our collection of slides. Clinically, I have seen 12 lesions which I thought belong to this group. In viewing the ciliary body with a gonioscope through an operative coloboma of the iris these cysts may be seen. The case reported by Francois[4] is such an instance and Dunnington[5] also has had such a case (Figure 7).

The internal and external layers of the secondary optic vesicle are not united but merely lie in apposition. In the postnatal eye also the structures derived from the one layer and the other are in loose apposition. In the posterior portion an actual separation of these 2 layers gives a detachment of the retina. Over the iris and ciliary body a separation of the 2 layers manifests itself as an intra-epithelial cyst. In the iris the site of predilection is the pupillary area where the marginal sinus remains patent.

I have no explanation as to why these cysts occur. A localized incomplete coaptation of the inner and outer layers of the secondary optic vesicle may serve as a patent space which later enlarges. Around the sixth fetal month the forerunners of the ciliary processes are seen as ridges or folds of the 2 epithelial layers. Sometimes these folds are characterized by a separation of the 2 epithelial layers, particularly at the junction between the iris and the ciliary body. It is

possible that the intra-epithelial cysts described in this paper have a predilection to occur at the base of the iris because at this transitional area the tendency for the 2 epithelial layers to be separated is carried over from fetal life. This explanation seems inadequate in that it does not include an impetus for the cyst formation. As previously stated, the cysts occur not only in apparently normal eyes but in eyes with a great variety of pathology. There are no indications that the cysts are related to the pathology for which the eyes are enucleated.

These intra-epithelial cysts of the ciliary body are, in my experience, far more common both microscopically and clinically than the similar type of intra-epithelial cyst of the iris arising in the pupillary area supposedly from a patent marginal sinus.

Greeff[6] described the blebs occurring in the ciliary body of rabbits due to the separation of the epithelial layers from the stroma. If, after a paracentesis of the anterior chamber, the rabbit's eye is enucleated, 10 minutes later there is present a hyperemia of the ciliary body and a detachment of both layers of the ciliary epithelium. In this way cysts are formed between the stroma and the epithelium and they are filled with an eosin-staining fluid. This lesion was interpreted as a proof of the secretory function of the ciliary body. The cysts discussed in this paper occur between the nonpigmented and the pigmented epithelial layers of the ciliary body and not between the layers of the ciliary epithelium and the stroma, and it seems highly improbable that they are related to Greeff's blebs or to the production of aqueous by the ciliary body.

When these ciliary body cysts are small they assume no clinical significance but when they are large they may manifest themselves in a number of ways. The periphery of the iris may be pushed forward so that a distinct localized narrowing or obliteration of the angle is noticed (Figures 1, 2, 8 and 9). This process may even embarrass the filtration angle sufficiently to produce glaucoma, as illustrated in cases

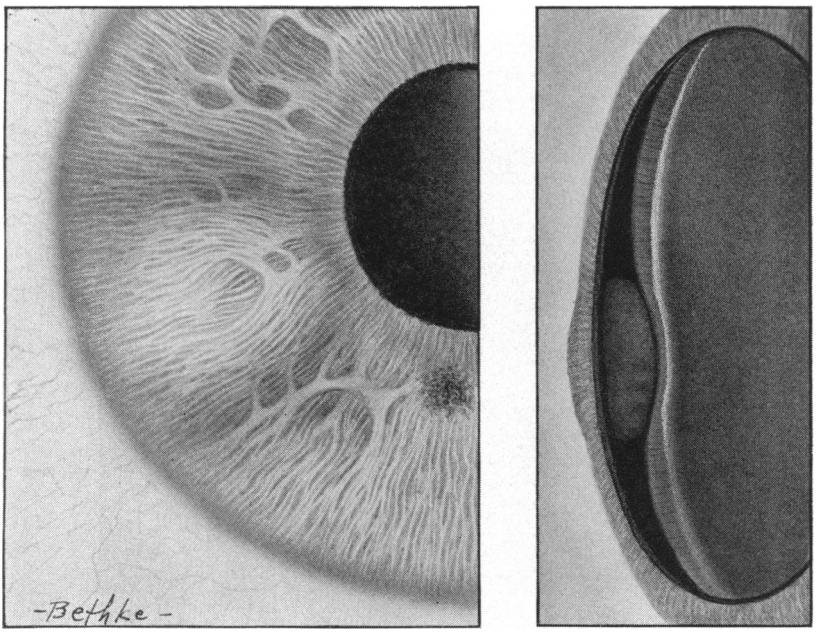


Fig. 9.—In the right eye of a 35-year-old man the iris bulges forward almost to the point of touching the posterior surface of the cornea. The angle is greatly narrowed but could be seen with a gonioscope. The lesion transilluminates light well. Through a dilated pupil a globular cyst can be seen posterior to the iris at the site where the iris is pushed forward. Over the area where the cyst indents the lens the zonules are missing. Posterior to the cyst several ciliary processes can be seen magnified. Over a period of 2 years and 3 months observation the lesion has not changed. (Patient of Dr. Daniel Rollet.)

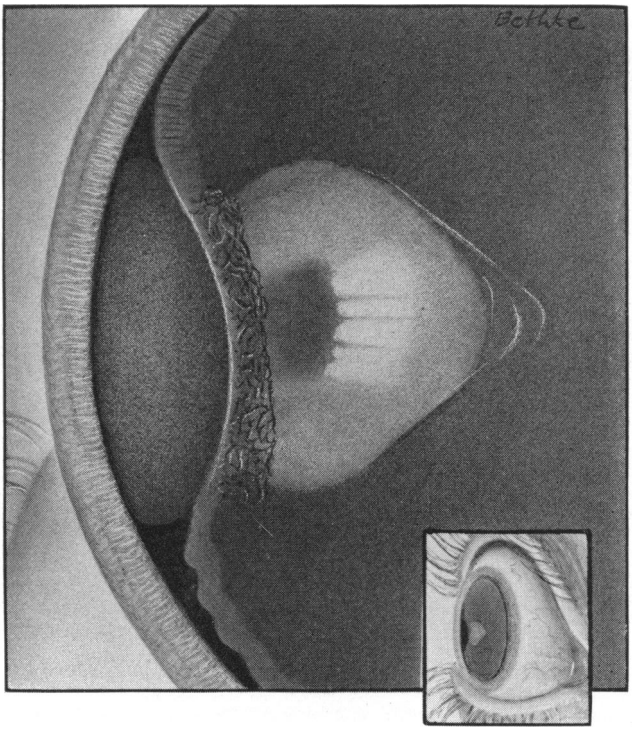

Fig. 10.—During a routine eye examination of a girl aged 16 a small coloboma of the lens was noted in the right eye at 7 o'clock. Through a dilated pupil it could be seen that a cyst over the ciliary body was responsible for the lens change. If the eye is moved while the lesion is being observed some tremulousness of the cyst is noted. There were some cataractous changes around the lens coloboma. The eye has been observed at yearly intervals for 4 years and no change has been noted. (Patient of Dr. Arthur Yudkin.)

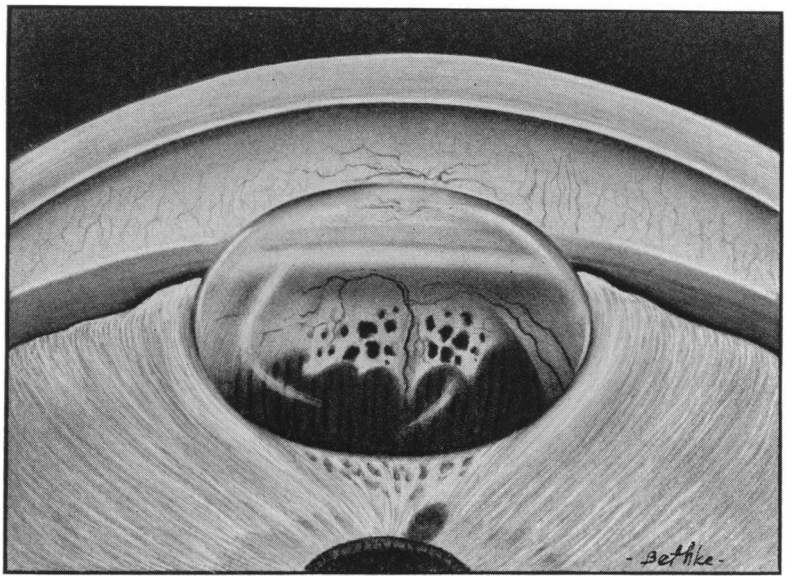


Fig. 11.—A spontaneous cyst in the periphery of the iris. A man aged 43 noted the lesion for the first time in 1946. An examination at this time showed a cyst 3 by 1.5 mm. The cyst was transparent and apparently extended through the iris stroma. The cyst enlarged over the succeeding 5 months. (Patient of Dr. J. H. Dunnington.)

reported by Wintersteiner[7] and Pagenstecher.[8] In the latter instance the author observed the cyst develop over a 6-month period and the glaucoma produced was rather transitory. The cyst may push the anterior surface of the iris against the posterior surface of the cornea and when this occurs the friction of the iris stroma against the cornea may incite proliferation and pigment changes which make the lesion simulate all the more a melanoma. Such a sequence occurred in my opinion in the case reported by Meek[9] (Figure 8). Although the case is reported as a melanoma of the ciliary body and iris, the author states that there was difference of opinion regarding the true nature of the lesion. I have had the opportunity to study sections of Meek's case and I believe it is primarily a cyst of the ciliary body with secondary iris changes and, therefore, belongs to the group described in this paper.

When the cyst extends along the posterior surface of the iris (Figure 4) it more often simulates a melanoma because the cyst wall is composed of pigmented epithelium, thus interfering with transillumination of light and giving the lesion an especially dark color which may be noted in the pupillary area. Important differentiating points in cases involving the iris are that the cyst protrudes from the posterior surface of the iris in the periphery, is smooth, and may be slightly tremulous. When the cyst arises from its usual site in the corona ciliaris (Figures 9 and 10), it has a smooth surface, a dark color, mostly because of its inaccessible location, transilluminates light, may be tremulous, and may indent the lens with or without secondary localized cataractous changes. A bright focal light thrown on the lesion may penetrate the cyst sufficiently to give a reddish reflex or hue from the underlying uvea. Also, under these circumstances, magnified ciliary processes may be identified posterior to the cyst (Figure 9). Because of the clear fluid content and the lack of pigment in the epithelium composing the cyst wall facing the posterior chamber, transilluminated light may have a refractile quality

which accentuates the cyst in direct contrast to what occurs in the case of a melanoma.

A type of translucent cyst may occur in the periphery of the iris, producing a lesion simulating an iridodialysis. Such cases have been reported by Elschnig[10] and by Villard.[11] Dunnington[12] also has had such a case (Figure 11). All 3 have a surprisingly similar clinical appearance. I suspect that these cysts belong to the group I am discussing but I am not sure. My doubt is based mainly on the fact that some tissue excised from the wall of Dunnington's case revealed an epithelial lining unlike the epithelium of the ciliary body. It was a stratified cuboidal epithelium more like the conjunctiva, although there was no history of operation or injury. Following excision of the cyst wall in both Elschnig's and Dunnington's cases there were recurrences.

Most of our cases have shown, over a period of years, no appreciable enlargement of the cyst. Therefore, the lesion is significant usually only insofar as it may be confused with a neoplasm or may produce glaucoma. Because of the usual location in the ciliary body it seems impossible to completely excise the lesion without doing irreparable damage to the eye. If the cyst is predominantly in the iris such an excision may be possible. No surgical procedure is indicated unless for diagnostic purposes or the integrity of the eye is jeopardized by progression of the lesion. Irradiation has never been tried.

## SUMMARY

A not uncommon type of spontaneous intra-epithelial cyst of the ciliary body is discussed. The lesion manifests itself usually as an iris lesion, may be confused with a neoplasm, and may produce glaucoma.

## BIBLIOGRAPHY

1. Rabitsch: Centralbl.f.prakt.Augenh., **28**:321–324, November, 1904.
2. Ichikawa, K.: Klin.Monatsbl.f.Augenh., **47**:73–82, 1909.
3. Loewenstein, A., and Foster, J.: Arch.Ophth., **37**:8–17, January, 1947.
4. Francois, J.: Ophthalmologica, **116**:313, 1948.
5. Dunnington, J. H.: Personal Communication.

6. Greeff, R.: Arch.f.Augenh., **28**:178, 1894.
7. Wintersteiner, H.: Ber.d.ophth.Gesellsch. (Heidelberg), **33**:345–359, 1906.
8. Pagenstecher, H. E.: Arch.f.Ophth., **74**:290–293, 1910.
9. Meek, R. E.: Arch.Ophth., **8**:864–870, 1932.
10. Elschnig, H.: Klin.Monatsbl.f.Augenh., **74**:476–483, 1925.
11. Villard, H., and Dejean, C.: Arch.d'ophth., **50**:91–121, 1933.
12. Dunnington, J. H.: Personal Communication.

## DISCUSSION

DR. DERRICK VAIL, Chicago, Ill.: In presenting this subject for our consideration Dr. Reese has made a contribution to our knowledge that is noteworthy for its clinical significance. I should like to add a report of a case of congenital iris cyst in an eye that was removed with the diagnosis of melanoma.

A student nurse, aged 21, was referred in consultation on November 5, 1940. In the course of a routine examination her ophthalmologist, Dr. Nancy Finney, of Cincinnati, had discovered a brown mass in the iris of the left eye between 3 and 4 o'clock. On examination, the iris angle on the temporal side showed a bulge that blocked the angle. The iris stroma in the affected area was atrophied and the pigment was increased in the extreme periphery. Transillumination was doubtful. The ocular tension was normal (19 mm. Hg., Schiötz). The ocular media were clear and the fundus normal.

The pupil was dilated with adrenalin, and one could then make out a flat, dark brown mass peeping from under the pupil margin at 3 o'clock. It seemed to be adherent to the anterior capsule of the lens, although the slit-lamp and biomicroscopic examinations showed no dissemination of pigment material on the anterior capsule, nor was there any evidence of lens opacification.

The gonioscopic examination showed an obliterated angle in the affected area, and the iris bulge was in contact with the cornea at this point, flattening the surface of the iris. The mass in the iris seemed to be sharply defined.

The patient was seen at frequent intervals until January 22, 1941, 10 weeks later, when both Dr. Finney and I thought that the mass had increased in size, especially toward the pupil margin. A faint haze in the posterior surface of the cornea when it was in contact with the lesion was now noticed. The tension was normal. Transillumination gave no further information.

A diagnosis of malignant melanoma of the iris was made and the eye was enucleated by Dr. Finney shortly thereafter.

Microscopic study of the depigmented specimen showed that the mesodermal part of the iris and iris muscles are normal. The pars iridae retinae shows multiple cyst formations which are mainly

located in the ciliary portion. The walls of the cysts are composed of cubical and flat cells derived from the pigment epithelium of the iris, the base of the cells facing the outside. The largest cyst, on the temporal side, extends along almost the entire posterior surface of the iris and adheres to the lens extensively. On the temporal side, the proliferation and tendency to cyst formation extends to the pars ciliaris retinae, where strands of proliferated cells, partly adherent to the larger cysts, have pulled some of the ciliary processes inward. The posterior segment is normal.

The fact that the cells were adherent to the anterior capsule of the lens in a firm fashion suggested to my mind that one of the causes of this formation of iris cyst might be some anomalous condition of the zonule fibers arising from the ciliary processes, with adhesion of the ciliary epithelium and the iris epithelium to the zonule fibers. I should like to ask Dr. Reese what he thinks of that idea.

Rapid growth of a heavily pigmented mass on the posterior surface of the iris, visible through the dilated pupil, obviously suggested the clinical diagnosis of malignant melanoma, and enucleation seemed advisable. The extensive and rapid cyst formation would soon, probably, have led to increased intra-ocular pressure and visual loss.

How can we recognize these lesions in the living eye? Dr. Reese mentions the following points: (1) the cyst may protrude from the posterior surface of the iris; (2) it is smooth; (3) it may be somewhat tremulous; (4) it may or may not transmit light, depending on the intensity of its pigmentation; (5) if it arises in the corona ciliaris it may indent the lens opacification. But the differentiation may be exceedingly difficult, as was revealed in my case, and even if a cyst is suspected, its rapid increase in size justifies the removal of the globe. As the specimen shows, there were multiple cysts present and conservative treatment, such as evacuation of the cyst or cysts, radiation, destruction with diathermy or electrolysis very likely would have resulted in loss of the eye in any event.

DR. ARTHUR M. YUDKIN, New Haven, Conn.: Dr. Reese has presented a most interesting contribution on cysts of the iris and ciliary body. I would like to record a case that has been included in his study.

In a routine examination of Miss N. P., aged 13, in 1945, I observed a small coloboma of the lens at 7 o'clock in the right eye. A small cysticlike enlargement of the ciliary body seemed to be responsible for the lenticular anomaly. The periphery of the

coloboma of the lens showed some cataract formation. She was examined in 1946, 1947 and 1948. I was satisfied that there was no increase in the mass located in the coloboma of the lens. However, she was referred in 1947 to Dr. Reese for consultation. I might add that her father had a malignancy of the lower bowel in 1947 and metastasis to the brain, and died 1 year later.

DR. F. H. VERHOEFF, Boston, Mass.: I want to compliment Dr. Reese on his paper. I think it is a remarkably good presentation of the subject. Of course all of us who examine many eyes have run across such cysts frequently, but I could not get together such a collection as he has shown here. I should like to ask him whether he thinks the cysts which have been described as free floating cysts in the anterior chamber do not have this origin. As to the clinical diagnosis, I have found that transillumination is not of much help. I remember a case in which transillumination showed a very bright light coming through a supposed cyst of the iris. I operated and found it was a solid tumor, which proved to be a malignant melanoma, unpigmented.

DR. RAMÓN CASTROVIEJO, New York City: In this very interesting paper by Dr. Reese the fact was particularly impressing that some eyes affected with benign cysts were enucleated because of the possibility of malignancy. I wonder if in these cases it would not be feasible to insert a fine needle through the limbus into the anterior chamber and into the suspicious iris growth to determine whether the supposed growth is solid or consists of liquid. If it is fluid and the growth collapses upon insertion of the needle it may be treated as a cyst by injection of a solution which will destroy it. If, on the other hand, the growth is found to be solid and the suspicion of malignant growth is confirmed, enucleation of the eye can be carried out immediately.

DR. DONALD J. LYLE, Cincinnati, O.: These 2 slides show a spontaneous transparent cyst apparently springing from the base of the iris at the angle. The cysts prevented dilation of the pupil with mydriatics or cycloplegics. There was no history of trauma or sign of inflammation. The nature of this cyst is not known to me, and it is shown here with the hope that Dr. Reese will discuss this type of cyst in his closing remarks.

DR. C. A. VEASEY, JR., Spokane, Washington: Apropos of differential diagnosis, I would like to describe a clinical experience I had about 18 months ago. A man 66 years of age presented himself for refraction. A short time before he had been refracted elsewhere, and

was dissatisfied with his glasses. His vision in the left eye, which was the faulty eye, was 20/50 with correction. Ophthalmoscopic examination showed a shadow at 6 o'clock behind the lens. With the dilated pupil you could see what appeared to be a spherical growth or cyst just behind the lens inferiorly.

In this case if the transilluminator was well behind the lesion it appeared dark. If the light came through the lesion it transilluminated quite well. By direct illumination it appeared yellow, perhaps somewhat fleshy, with 1 or 2 blood vessels visible on the surface. Because it transilluminated I was afraid it might be a cyst. Certain differential diagnosis did not seem to be possible clinically. Under local anesthesia I dissected up a conjunctival flap at 6 o'clock. Then about 4 mm. below the limbus, after sparking the area with diathermy, I introduced a No. 22 needle on a tuberculin syringe, expecting to aspirate some fluid. The needle was introduced with a rotary motion. No fluid was obtained. There was a minimum of moisture in the needle, perhaps aqueous from an aqueous vein. The contents of the needle were blown on a slide, dried and stained. The needle had bored out a core of cells which appeared to be malignant. The eye was enucleated and examination disclosed a nonpigmented malignant melanoma.

DR. ALGERNON B. REESE, closing: Dr. Vail's case illustrated the varied clinical manifestations these cysts may assume. I think his idea of the adhesions of the zonule to the ciliary epithelium as a source of origin of the cysts is an interesting and plausible one.

I am very glad to have Dr. Yudkin add the additional facts in regard to the case of his I used in this study.

Dr. Verhoeff asks whether or not these cysts may be the origin of the rare appearance of a free cyst in the anterior chamber. I think they might well be, although these cysts of the ciliary body do not tend to be pedunculated.

Dr. Verhoeff mentioned the use of transillumination in diagnosing the condition. I believe these lesions transmit light if they are on the corona ciliaris, because the inner layer is nonpigmented. If they are at the base of the iris, where the entire wall of the cyst is lined with pigmented epithelium, the light tends not to be transilluminated.

Dr. Castroviejo mentioned the possibility of inserting a needle for diagnostic purposes. I think this is a sound idea.

Dr. Veasey has related to us what in my opinion is the first instance in which an aspiration biopsy has been employed for an intra-ocular tumor.