# COATS' DISEASE AND CONGENITAL VASCULAR RETINOPATHY

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## INTRODUCTION

A COMMUNICATION IN THE ROYAL LONDON OPHTHALMIC HOSPITAL REPORTS, Volume XVII for the year 1908, was entitled, "Forms of Retinal Disease with Massive Exudation," by George Coats, Curator.[1] This lengthy presentation is now recognized as a classic in medical literature. Few medical publications have engendered such imperious controversy or called forth such a profusion of writing. Many writers have reexamined, restated the nature of, and reclassified Coats' disease and its various forms.

This essay is not just another attempt to reclassify these ocular disorders but rather to invoke a return to Coats' own broad classification and perhaps extend it. I also hope to demonstrate that this entire group of diseases consists of manifestations which I shall call congenital vascular retinopathy. Such a return to Coats' classification was suggested several years ago by Sugar.[2] The present writer cannot agree with Sugar, however, that such a resurrection of the original classification would be "a regression in our thinking."

In my opinion, all disease can be viewed as a continuum, as a spectrum, wherein related disease processes merge gradually into contiguous pathologic entities without the pigeonholing of this disease, that disease, or so-and-so syndrome. I feel that this group of ocular disorders presents us with an excellent example of this concept. I hope to demonstrate that Coats' disease forms a spectrum with loci representing the "classical form" of a particular disease merging into the "atypical form" of a related entity. Within this spectrum of congenital vascular retinopathy will be included not only Coats' disease, Leber's miliary aneurysms with retinal degeneration, and Reese's telangiectasia, but also von Hippel's disease, racemose aneurysms, and related malformations.

The opening sentences of Coats' publication read as follows: "The cases with which the present paper deals form, with other similar records from

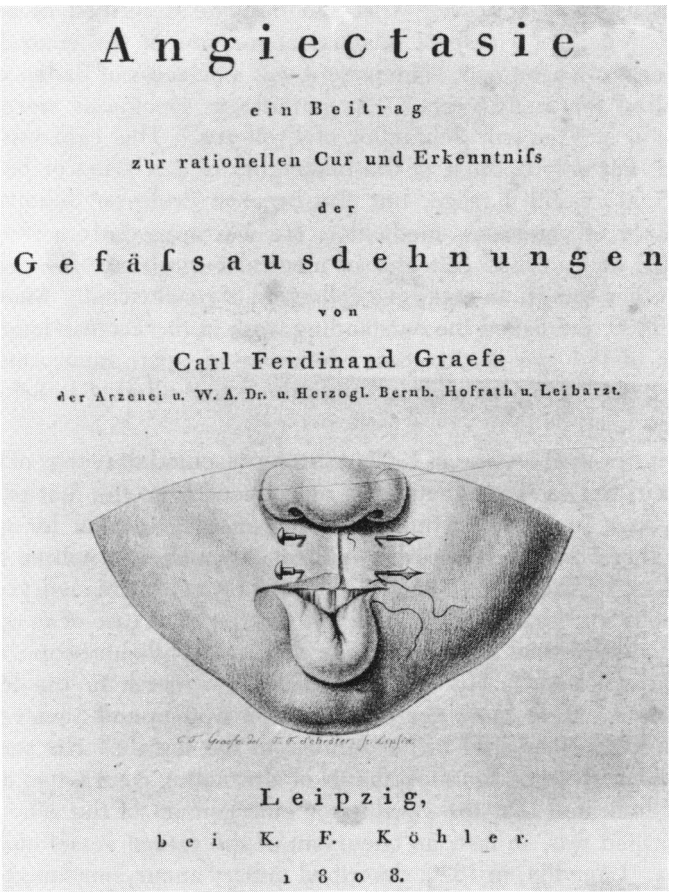
the literature, a fairly well defined group. They are characterized by the presence in some part of the fundus of an extensive mass of exudation. In some instances very peculiar forms of vascular disease are also found." Seven decades later, I hope to demonstrate that the earliest manifestation of this group of diseases at one end of the spectrum is indeed a "very peculiar form of vascular disease" and that "an extensive mass of exudation" is among the latest manifestations and at the other end of the spectrum.

## MATERIALS AND METHODS

This essay presents the findings and observations in a series of cases of Coats' disease and related entities comprising part of a long term, continuing study of retinal vascular anomalies. A total of 47 patients were examined and 54 eyes were affected. Three incomplete pedigrees of families with Coats' disease will be included. With one exception, all patients were personally studied in detail and followed by the writer. The follow-up period ranged from 17 years to the present. At the initial examination, fundus photography was supplemented by detailed retinal drawings. When possible, fluorescein angiography was performed. Prior to treatment, an exhaustive medical work-up was performed. With the exception of initial single treatments with the argon laser in two patients, as detailed below, all treatments and surgical procedures were performed by the writer. A short description of each case is included. The evolution of the ophthalmoscopic picture in this group of disorders will be discussed. From a study of these cases, I hope to demonstrate, in agreement with previous authors, that the basic lesion in all forms of Coats' disease, Leber's retinal degeneration with miliary aneurysms, and Reese's telangiectasia, is a telangiectasia of the terminal retinal vasculature.

## HISTORICAL REVIEW

Carl Ferdinand Graefe of Leipzig published a treatise on the diagnosis and treatment of aneurysms in 1808[3] (Figure 1). He described, "Aneurismen der Zentralarterie" and "Telangiectasie der Retina," in a woman who had complained of pain and photophobia in an eye which became blind over several months. The "Zentralarterie" was the central retinal artery. Thus, the term, "telangiectasia" in relation to the retina was given its place in history. Telangiectasia is derived from the three Greek words, telos = end + aggeion = vessel + ektasis = dilatation.


Angiectasie

ein Beitrag

zur rationellen Cur und Erkenntniſs

der

Gefäſsausdehnungen

von

Carl Ferdinand Graefe

der Arzeneï u. W. A. Dr. u. Herzogl. Bernb. Hofrath u. Leibarzt.

Leipzig,

bei K. F. Köhler.

1808.

FIGURE 1
Frontispiece of Graefe's treatise published in 1808.

In my opinion, telangiectasia is a precise term and aptly describes the initial ophthalmoscopic lesion in Coats' disease. Some modern authors have sought to denigrate the term[4] or have considered it to be of recent origin.[5] A century after Graefe, we find the term in Coats' publication.[1] Graefe described a second case he saw in Dresden and considered it similar to the one described above. No pathologic examination was made. In 1818, Demours described the marked dilatation of the central retinal artery as an aneurysm in a woman who died of metastatic breast carcinoma.[6]

In 1819, there appeared what can only be described as an extraordinary report of a case of bilateral aneurysms of the central retinal artery, again in a woman. The patient was a princess of Baden who had been blind for many years. The pathologic specimens were in the possession of Professor Schmidler of Freiburg.[7] This case was indeed unusual, not only because of the bilaterality of the lesion or because of the patient's noble lineage, but also because Professor Schmidler was a professor of veterinary medicine. He was apparently a remarkable individual and an avid collector of medical curiosities. He possessed, among other things, an extensive collection of renal calculi. MacKenzie, who in 1830, published the outstanding work in the English language on diseases of the eye at that time, commented upon aneurysms of the central retinal artery as a cause of blindness and alluded to Schmidler's case.[8]

In the Bowman lecture of 1965, Ashton presented drawings of fusiform and sacculated capillary aneurysms reproduced from the first pathologic record book in his department of Moorefields Hospital for the year 1854. These drawings were by William Bowman and Ashton felt that these must be the earliest know records of retinal microaneurysms.[9]

In 1865, Sous described what he felt was the third case of an aneurysm of the central retinal artery and gave the first ophthalmoscopic description of the lesion.[10] His case is actually the fourth in the literature to that date. Once more the patient was a woman and Sous remarked that the lesion appeared to be peculiar to the female. His statement, "The tumor executes the movements of alternating contraction and dilatation," indicated that the swelling or enlargement of the artery which he described was, in fact, an aneurysm of the retinal vessel and not an angioma. Liouville, in 1870, described miliary aneurysms found at postmortem in the retina of a patient who had died from a cerebral hemorrhage, originating in rupture of similar miliary aneurysms in the brain.[11]

Bouchut, in his atlas of ophthalmoscopy, published in 1876, gave two examples of aneurysms of the retinal arteries apparently in patients with general paralysis of the insane. One of these figures was reproduced in the German literature with a notation that the lesion occurred in the arteries in association with neuroretinitis.[12] It is possible that these figures were a misinterpretation of changes, or apparent changes, brought about by edema of the nerve head and the adjoining retina. An English contemporary, discussing Bouchut's illustrations of these aneurysms, remarked tersely, "His figures, however, suggest considerable exaggeration."[13]

In 1877, Stephen MacKenzie recorded finding microaneurysms in a patient with glycosuria.[14] This lesion was to be forgotten until rediscovered, as it were, by Ballantyne and Lowenstein.[15] An interesting sidelight on history was the fact that it was Warren Tay of Tay-Sach's disease who discovered the fundus lesions in MacKenzie's patient.[16]

In 1879, Gowers classified aneurysms of the retinal arteries. He stated, "Two forms of aneurisms have been observed: (1) Aneurisms of some size on the primary branches of the central retinal artery on the disc; (2) miliary aneurisms of the arterial twigs upon the retina, and of the small capillary vessels."[17] In the Transactions of the American Ophthalmological Society for the year 1880, Loring published the first report in the American literature with a drawing of an aneurysm of the retinal artery.[18] At the Dublin Ophthalmological Club, in March, 1883, Story and Benson demonstrated retinal miliary aneurysms in the right eye of a 20-year-old man. The aneurysms were present on both the arteries and veins.[19] I believe this is the first description of what later became known as Leber's Miliary Aneurysms with Retinal Degeneration. Doyne, in 1896, reported a similar case and even though the details were sparse, the nature of the lesion is evident from the accompanying sketch.[20]

In Oeller's atlas of 1899, telangiectasia with arteriovenous shunts in the macula was depicted.[21] Oeller's case was very similar to case #44 in the present series. He described a subsequent rupture of the abnormal vessels. This was unusual; the bleeding may have resulted from a detachment of the vitreous. Thus, at the turn of the century, what became known as Leber's retinal degeneration with miliary aneurysms had been well defined in the literature.

Krause and Buckner reported a case with both venous and arterial aneurysms in 1907.[22] Subsequently, both Coats and Leber claimed that this case was an example of each disease. Leber, in his original article of 1912, cited only two personally observed cases, the remaining 11 being culled from the literature.[23] His first case was a 25-year-old man with absolute glaucoma and miliary aneurysms in the retina. It is well known that miliary aneurysms occur in glaucomatous eyes, as well as in many other conditions apart from diabetes mellitus.[24,25]

In 1908, 1911, and 1912, Coats' three articles appeared. Coats' publications engendered much comment and speculation, which have continued to the present. In 1915, Leber elaborated on the nature of retinal aneurysms,[26] and in 1916, he discussed the exudative retinitis of Coats under the term, "Serofibrinous degenerative retinitis and chorioretinitis".[27] Leber now felt that the pathology of the condition that Coats

had described consisted of one to several foci of inflammation and necrosis in the retina with serofibrinous exudation. Eventually, encapsulation of the subretinal debris took place by the formation of surrounding fibrous tissue, some of which, at least Leber felt, was derived from the retinal pigment epithelium.

In 1918, Pringle described bilateral retinal aneurysms in a 19-year-old man. The lesions involved the larger branches of the arterioles. Secondary retinal changes were absent.[28] In 1921, Miyashita and Nisyake described the pathologic findings in a case of multiple miliary aneurysms of the retina.[29] Their case was evidently one of retinal telangiectasia without intraretinal hemorrhage. These authors believed that the disorder they described was due to a hereditary weakness of the blood vessels.

In 1934, Junius endeavored to relate Leber's miliary aneurysms of the retina to hereditary hemorrhagic telangiectasia.[30] In 1938, Lamb gave the pathologic findings in "six early and two late cases" of exudative retinitis. He was much influenced by Leber's views. Under scrutiny, a few of Lamb's cases do not meet the criteria for Coats' disease.[31] Elwyn considered Coats' disease as a telangiectasia of the retinal vessels and classified the disorder as a vascular malformation.[32] In 1956, François and coauthors sought to incriminate toxoplasmosis in the etiology of Coats' disease.[33] Reese, in agreement with Elwyn, felt that Coats' disease was, "Basically a more or less masked telangiectasis of the retinal vessels." Reese also felt that Leber's multiple retinal aneurysms and Coats' disease were probably the same process. Leber, in his initial article on the subject, had suggested that the condition which later became known by his name was an early stage of Coats' disease. Reese described the deposition of mucopolysaccharide in the telangiectatic vessels in his cases.[34] A similar process had previously been described by Faber in the aorta.[35] In 1958, Sugar went even further than Reese, stating, "I believe that the primary pathological process may be telangiectatic or aneurysmal or even of other nature, including von Hippel's disease. This is, indeed, a regression in our thinking to Coats' original description, but I believe it to be accurate."[2] Wise, in 1961, considered local retinal hypoxia to be the primary etiologic factor with hemorrhage and vascular changes as secondary effects.[36] In 1962, Imre suggested an endocrine disturbance as an etiologic factor in Coats' disease.[37] In 1963, Woods and Duke endeavored to link hypercholesteremia with uveitis as etiologic factors in "the adult form of the disease."[38] In 1964, Woods and Duke gave results of experimental work on rabbits in support of their hypothesis.[39] In 1971, Tripathi and Ashton, in an electron-microscope study, suggested that the basic lesion in Coats' disease may be a

functional or structural breakdown of the blood-retinal barrier resulting in aneurysmal dilatations and telangiectasis.[40]

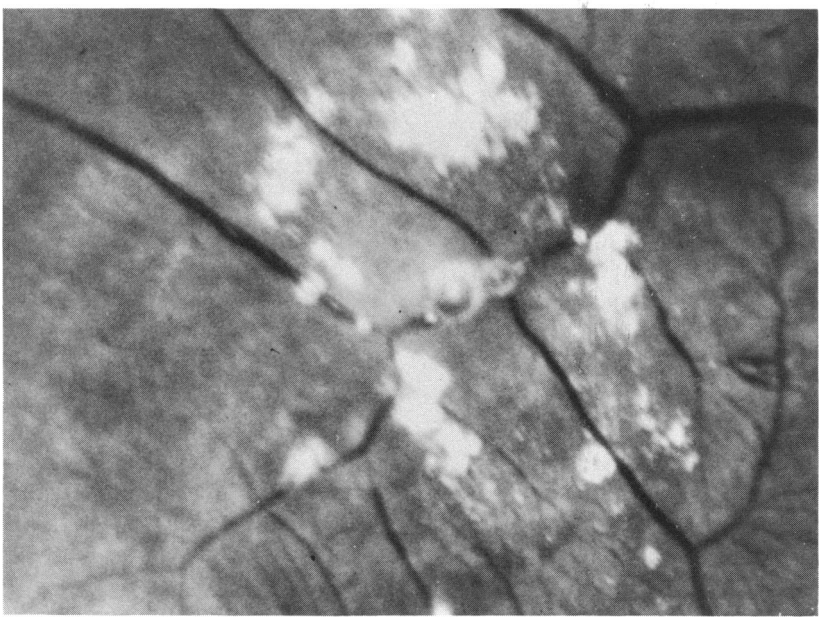


FIGURE 2
Macoraneurysms surrounded by exudate, Case 20, O.S.

## OPHTHALMOSCOPIC APPEARANCE

The pathology of Coats' disease has been emphasized in the literature. All too frequently the cases presented were in the later or terminal stages. On the other hand, in this writer's opinion, the evolution of the ophthalmoscopic picture has frequently been lacking in precise detail. Based on the present series of cases, therefore, certain ophthalmoscopic features will be described.

### *1. Retinal Vessels*

Accentuation of what appeared to be a normal capillary bed forming an obvious network with arteriovenous shunts at or beyond the equator in the temporal half of the eye is perhaps the earliest visible extramacular change in the fundus (Figures 12, 13). This alteration in the vasculature may be localized and remain static, or it may progress to involve all quadrants. Exceptionally, all quadrants may show this initial change.

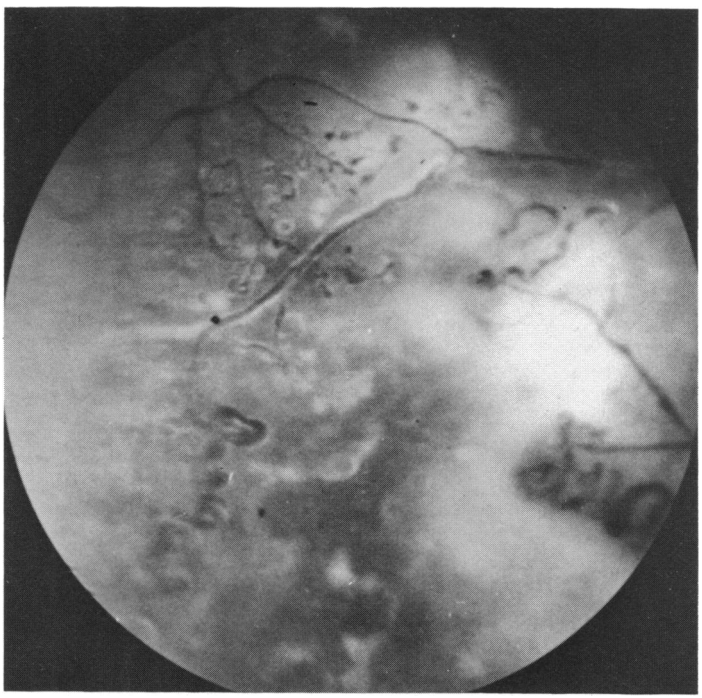


FIGURE 3
Typical peripheral telangiectasia, Case 10, O.D.

In cases showing progression, the affected vessels become enlarged and distended, forming sausage-like dilatations. Balloon-like structures appeared on the arterioles and veins. They were both single and multiple (Figure 2). On the arterioles, such structures, occurring as an isolated finding, have been termed "macroaneurysms" by Robertson.[41] Hemorrhage into the retina was noted with these aneurysms in case #20, similar to those of Shults and Swan.[42] Aneurysmal pulsation, as described by these authors, was not observed. These macroaneurysms are most probably similar, if not identical, to the structures in the optic nerve described by Graefe,[3] Schmidler,[7] and Sous,[10] and in the retina by Loring[18] and Pringle.[28] McDonald and Sarin have recently discussed the treatment of this type of lesion.[43] Beading of vessels, similar to that seen with large retinal angiomas, was an infrequent finding. Irregular loops appear on vessels and miliary aneurysms develop.

As the affected vasculature becomes more floridly distended, masses of telangiectasia result (Figure 3). Small, localized conglomerations of telangiectatic vessels, surrounded by a circinate figure of exudate, may

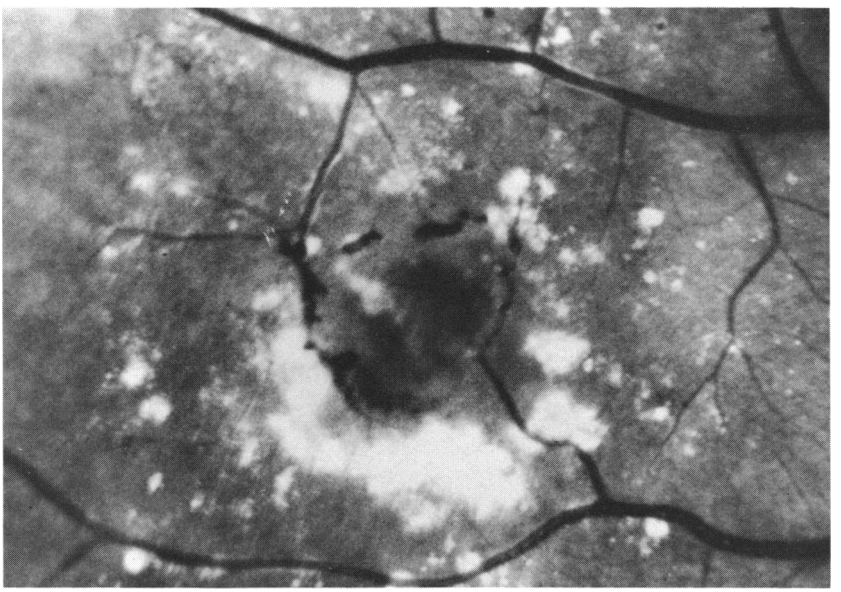


FIGURE 4
Localized telangiectasia, Case 20.

be seen apart from the larger areas of involvement (Figure 4). By the time the mass of telangiectasia involves one quadrant of the retina, and if vascular decompensation is marked, localized elevation of the surrounding retina occurs. In the posterior polar region, the initial changes usually take the form of tiny, balloon-like dilatations, on the terminal vessels, commonly referred to as miliary aneurysms (Figure 5A). They arise on either the arterioles, the veins, or both. The changes, however, may involve the entire macular vasculature. Thus, in case #44, accentuation of the capillary network with large bulbous vessels and areas of telangiectasia was present (Figure 5B). So long as the abnormal vasculature does not decompensate, secondary retinal effects do not occur. Decompensation is indicated by staining of the vessel wall or leakage on fluorescein angiography. Ophthalmoscopically, decompensation is indicated by retinal edema, retinal hemorrhage, or the appearance of exudate. Such decompensation has been noted in the study of arteriovenous communications of the retina.[44] Both edema and exudate may appear in the immediate vicinity of the abnormal vasculature or in uninvolved regions. Thus, commonly the macula will show slight edema with radiating striae, microcystic edema, or less frequently, an extensive mound of exudate. In fully developed areas of telangiectasia at or be-

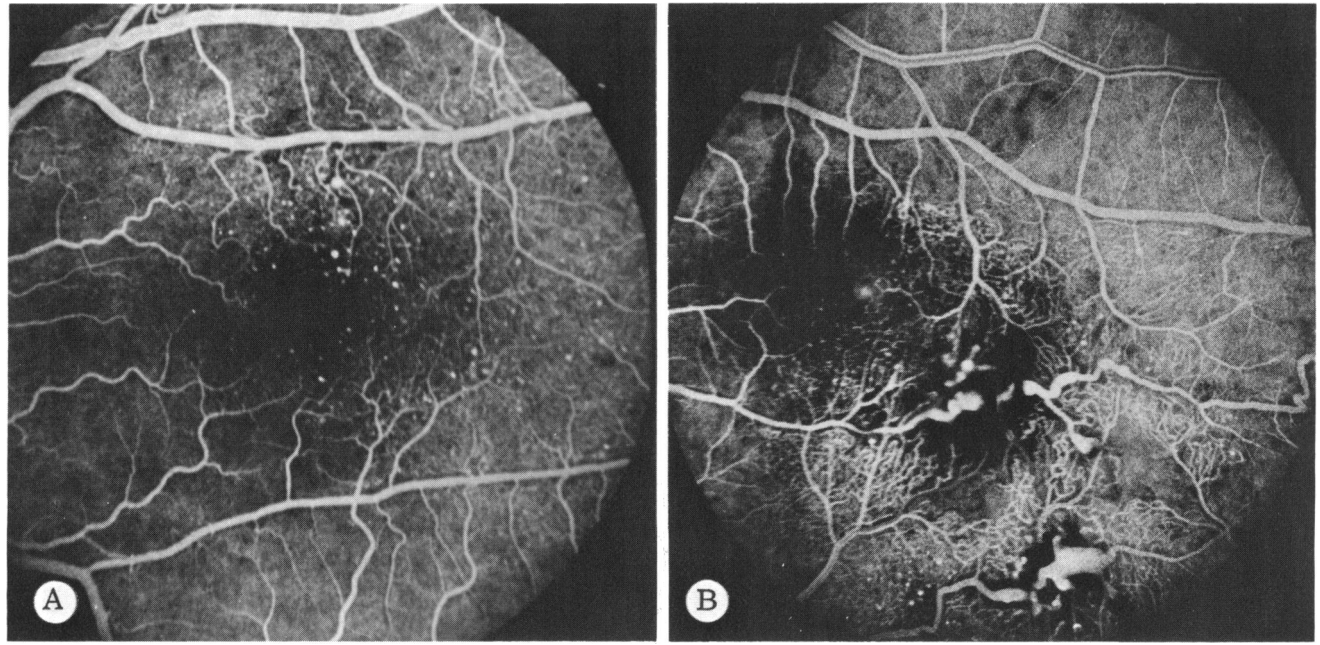



FIGURE 5

A: Angiogram showing central telangiectasia—miliary aneurysms. Case 11, O.S. B: Angiogram of central telangectasia. Involvement of all vascular components. Case 44, O.S.

yond the equator, the more peripherally placed vessels become converted to white, thread-like structures. This change may be the result of a thrombotic process in the more distal reaches of the abnormal vasculature. The thrombosis in these vessels could be a direct effect of stasis in the proximal telangiectatic mass. The main retinal veins draining the area of telangiectasia show varying degrees of dilatation, depending on the bulk of the abnormal vasculature. In the classical von Hippel tumor, as the angioma enlarges, the associated vessels also enlarge. In retinal telangiectasia, however, while a significant degree of dilatation of the draining vein does occur, the feeding arterioles do not usually show a corresponding degree of enlargement. This difference in the vessels in the two lesions may be due to the increased size of the capillary bed in the angioma while telangiectasia is simply an area of shunt formation with lower resistance and stagnation of flow. With the appearance of extensive exudate, both veins and arterioles may show heavy sheathing with this yellow material.

2. *Retinal Hemorrhages*

Small hemorrhages in the superficial retina and on the retinal surface are a widespread and constant feature in all late cases.

3. *Retinal Angiomas*

Single or multiple angiomas may occur in areas of telangiectasia. They may also present as isolated lesions in eyes with telangiectasia in other areas. These angiomas are small in size, the largest in the present series being about one-half disc diameter. Small angiomas were present in cases #9, 13, 19, and 21.

4. *Subretinal Mounds*

Elevated, tumor-like masses are frequently found in the fundus at the initial examination. I use the term, "subretinal mound," because the appearance frequently suggests a mass beneath the retina involving the choroid. Coats describes such a lesion. "The lower quadrant of the fundus is occupied by a large mass apparently in the choroid, which gives in parts a greyish reflex, in others a yellowish white. The central portion of the mass is swollen out into a boss which shows a good deal of retinal haemorrhage on its summit."[1] This description admirably fits the lesion in case #26. These mounds may be multiple and located anywhere in the fundus. Telangiectatic vessels usually course over them. In long-standing cases they may be covered in part or completely with yellow, shining crystalline bodies. These lesions are a

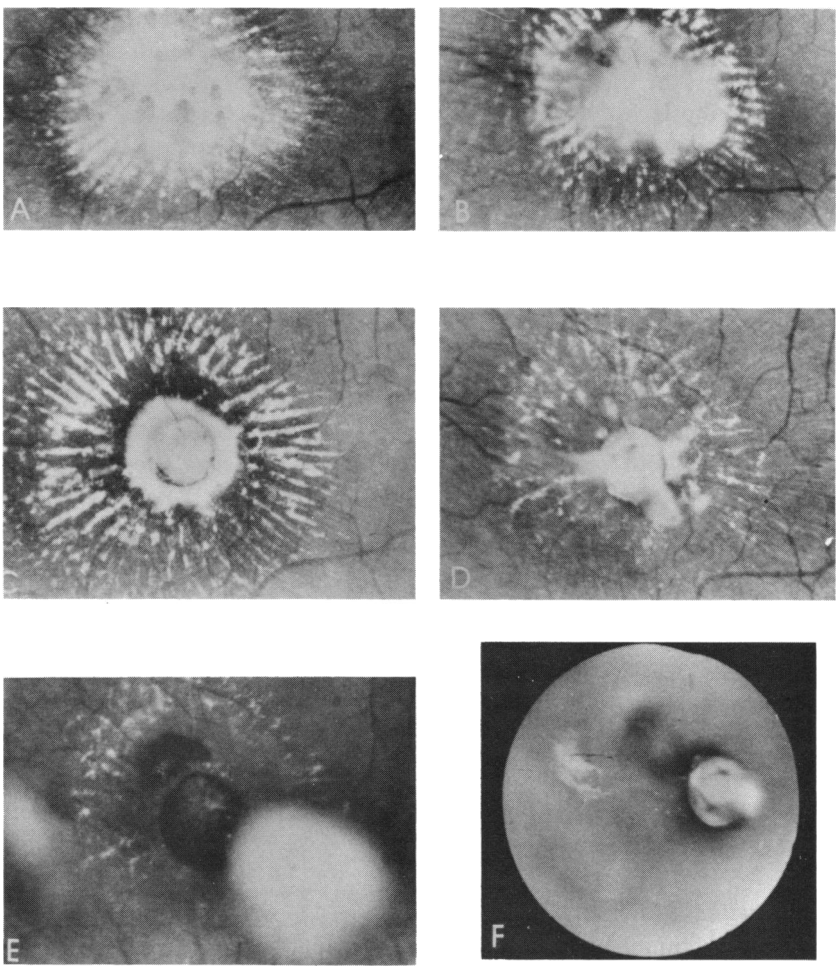



FIGURE 6
Resolution of central macular star with retinal hole formation. Case 30, O.S.

frequent finding in eyes showing rubeosis iridis, retinal detachment, cataract, and glaucoma. These eyes are often enucleated because of the possibility that the eye may harbor a malignant tumor. These lesions are also found, however, in eyes with relatively early changes.

*5. Retinal Exudate*

The formation of retinal exudate is a response to an alteration in the blood-retinal barrier brought about by anoxia. Exudate occurs in two distinct types and both may be found in a single eye.

Type I: This variety appears as a thin, broken, or fragmented layer of light yellow material in the deeper retinal layers in the vicinity of the abnormal vasculature. The edge of the exudate is always separated from the telangiectasia by a clear interval. It may be scattered throughout the entire fundus in stellate figures which coalesce in some areas. Such stellate clumps of exudate, occurring throughout the fundus in association with a localized peripheral mass of telangiectasia, may be the result of a steal-syndrome mechanism, producing hypoxia throughout the entire retina.[45] The left eye of case #30 exemplifies such widespread exudate with a localized mass of abnormal vasculature. Type I exudate forms circinate figures around solitary, larger aneurysms or around isolated telangiectatic masses. It forms a typical star-like mound in the macula when the abnormal vasculature is localized in the temporal periphery. This type of exudate disappears rapidly from the periphery in cases responding to treatment. The star-like figure in the macula leaves a residual nubbin of fibrous or glial tissue which can persist for years, even after the disappearance of all exudate and abnormal vessels. This star-like figure is found only in those eyes where marked vascular decompensation has occurred, in the peripheral telangiectasia. Type I exudate commonly sheathes both arterioles and veins in advanced cases.

Type II: The second variety of exudate occurs in broad sheets in the posterior polar region. Its most striking feature is the continuity of the broad sheets of the material. Like Type I it is yellow in color, but of a decidedly darker hue. In all cases with this type of exudate, subretinal mounds were present. This exudate is due to decompensation, stagnation of blood and its attendant hypoxia, as with Type I, though probably the degree of hypoxia is much more marked in the presence of a subretinal mound than in the presence of superficial telangiectasia alone. The darker yellow color is probably due to absorption of pigment from the breakdown of blood in the subretinal mounds. In case #26, Type II exudate shifted with sustained changes in position. Type II exudate absorbed extremely slowly following treatment, in contrast to Type I, which showed evidence of absorption within days after treatment was begun.

*6. Retinal Detachment*

Elevation of the retina is a frequent finding in a localized area around telangiectasia. It is preceded by the appearance of exudate.

As the mass of abnormal vasculature increases, with concomitant circulatory decompensation, the area of elevation spreads posteriorly to involve the macula and posterior pole. In the presence of subretinal mounds, preretinal fibrosis is common and localized traction detachments ensue. In advanced cases with multiple subretinal mounds, the retina becomes totally detached and disorganized. In case #10, a large intraretinal cyst surrounded by Type I exudate occurred in the elevated retina.

7. *Retinal Pigment Epithelium*

Small, scattered clumps of pigment derived from the retinal pigment epithelium are often seen in areas of telangiectasia, particularly after the retina has become elevated. Where the elevated retina flattens with treatment, the underlying pigment layer in untreated areas shows a moth-eaten appearance. Rarely does pigment sheathing of the vessels occur. On the whole, in cases of superficial telangiectasia, the changes in the retinal pigment epithelium are not striking.

8. *Disc*

Apart from occasional hyperemia, the disc is normal until the later stages. The hyperemia is due to the general stagnation of blood flow in the retinal vasculature with resulting local anoxia and capillary dilatation. In one advanced case, optic atrophy was present. In advanced cases with multiple subretinal mounds and retinal detachment, the disc may be obscured from view. The disc may be the actual site of vascular change, as mentioned by Reese,[35] and as described in cases #18 and #35. In cases in which light perception was absent, abnormal vasculature may have involved the retrobulbar portion of the optic nerve.

9. *Vitreous*

Except in advanced, inoperable cases with multiple subretinal mounds, the vitreous is quite clear. In some cases of extensive telangiectasia without subretinal mounds, scrutiny with the slit lamp and contact lens may show a fine haze with pigment flecks. In the later stages, vitreoretinal adhesions and bands are formed with ensuing retinal contracture and detachment. Hemorrhage, rare in early cases, now commonly ensues. Hemorrhage can occur, as in the case of a normal eye, due to vitreous detachment and the symptoms may call for fundus study which reveals an early lesion. Retinitis proliferans does not develop with telangiectasia, except in the late stages, but may occur in the presence of a

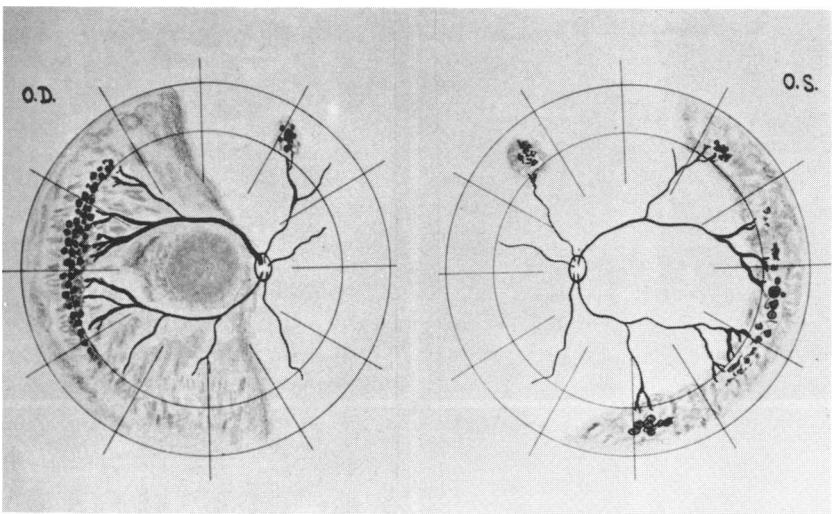



FIGURE 7
Pretreatment drawing of case 3.

still attached retina or with only minimal retinal elevation if subretinal mounds are present, as in case#26.

*10. Macula*

The initial lesion in the macula usually takes the form of multiple miliary aneurysms, which may remain unchanged for years. Circinate exudate may surround clusters of these aneurysms. Both the aneurysms and the surrounding exudate may disappear from one area, only to reappear nearby, as in case #23. With the passage of time, the miliary aneurysms in the macula may gradually increase in number and become part of a generalized telangiectasia involving the entire macular vasculature, as in case #19. Lastly, the initial picture at the macula may be a combination of miliary aneurysms, networks of telangiectatic vessels similar to those which occur in the periphery, with multiple arteriovenous shunts and large, dilated, bulbous vessels (Case #44). Vascular decompensation results in retinal edema, exudate formation, and edema and thickening of the underlying retinal pigment epithelium. As pointed out above, a macular star in association with temporal telangiectasia is a frequent finding. This change may vary from a barely discernible macular fan or wing, corresponding to the quadrant of greatest vessel density, to a full-blown star-like figure with its conical center projecting into the vitreous.

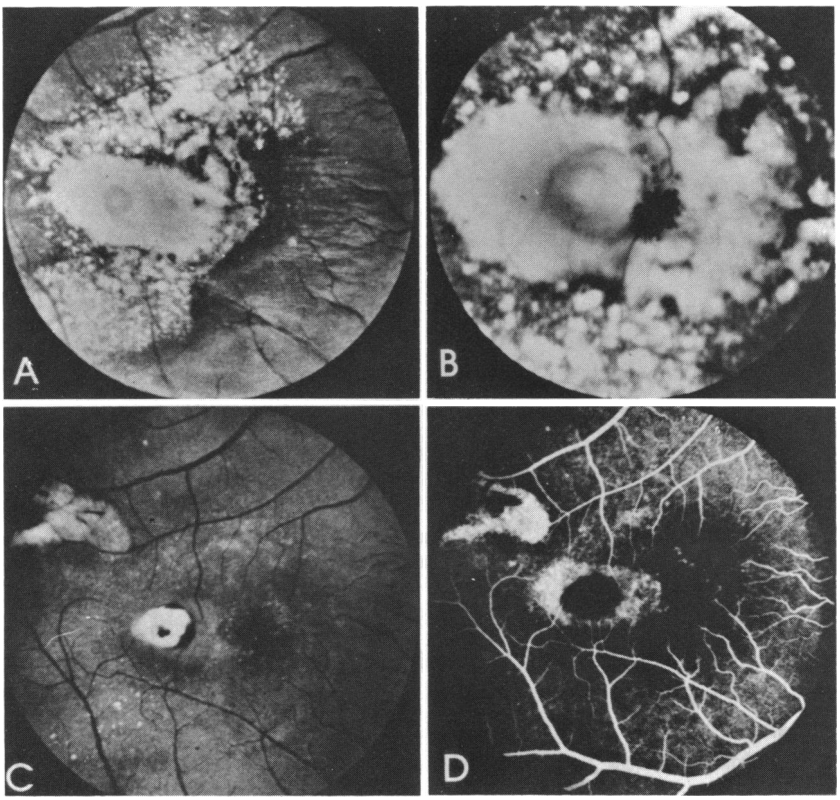



FIGURE 8

Case 3, macula, O.D. A: Early resolution of central mass of exudate six months after commencement of treatment. B: Further absorption of exudate. A small retinal vessel is seen entering edge of glial nubbin just above a superficial hemorrhage. C: Three years post-treatment. Glial nubbin persists. D: More than 10 years post-treatment, central nubbin is replaced by a flat scar. Angiogram reveals early central telangiectasia.

Small retinal vessels may enter the apex of this central mass, as in case #3 (Figure 8B).

Thus, in the evolution of the ophthalmoscopic picture, it is seen that any of the given vascular abnormalities may arise in any region within the retinal substance. Masses of superficial telangiectatic vessels occur more commonly at the equator and beyond, while miliary and macroaneurysms occur in all areas of the fundus. This point is stressed to emphasize that the fundamental process involves the entire retinal vasculature, including that of the optic nerve.

ANALYSIS OF COATS' ORIGINAL CASES

In his initial publication in 1908, Coats described six cases.[1] This was followed by a seventh case in 1911.[46] In his third and last publication on the subject which appeared in the German literature in 1912, 10 cases were described which included the seven cases of 1908 and 1911.[47] In this third publication he also discussed an eleventh case, which he had not examined, and considered it of significance only because of the presence of an epipapillary membrane. Due to lack of detail and the absence of a pathologic report, it is excluded from this analysis. Case number six was not included since "only one or two sections were available for study" and because no ophthalmoscopic details were given. Case number ten was also excluded because in the opinion of this writer it does not meet Coats' own criteria for inclusion. In all probability it represented some form of long-standing total detachment of the retina. Pathologic examination was performed in all cases except number seven. This patient was followed by Coats and his colleagues for 16 years. With the exception of case number three, subretinal mounds were present in the remaining eight cases. Case number three differs from the others in the group in that a total detachment of the retina, except for the macula, was present.

The ophthalmoscopic picture in case three, as given by Mr. W. W. Sinclair, described the macula as covered by a dense, white mass with pigmented edges. This mass was elevated. At its apex a retinal artery was seen to break up into several fine branches. Only at the extreme periphery of the fundus was a slight trace of retinal hemorrhage visible. In the macula, Coats described a nodular mass of fibrous tissue in the outer retinal layers and the subretinal space. It was fully organized and almost homogeneous under the high power. Further, in his pathologic report, Coats states, "Apart from the mass in the macula, the chief changes of the retina are in the periphery and on the temporal side. . . . Blood vessels occur in extraordinary numbers and in places give quite a cavernous aspect to the inner layers of the retina. A few of them are found in the outer layers. They are nearly all crammed with blood. In most cases the wall is thin and not diseased, resembling that of a vein. In a few instances it is thick and the lumen contains coagulum. The enlargement of the vessels ceases a short distance behind the ora serrata."

Case number three, then, is that of a boy, age seven, with a marked reduction in vision in his right eye for about a year. Ophthalmoscopically, a lesion is described in the macula with hemorrhages in the periph-

ery. Pathologically, a total retinal detachment without subretinal mound formation or extensive subretinal fibrotic formation was noted. A nubbin of homogeneous tissue was present at the macula. The retina was intimately connected to this tissue. Markedly dilated, thin-walled vessels were present in extraordinary numbers in the temporal periphery. In my opinion, this third case of Coats was an example of superficial retinal telangiectasia with secondary macular change. This case was identical to a number of the cases comprising this presentation (#3, #9, #10, #28, #30, and #31).

In case one, subretinal mounds were noted ophthalmoscopically and demonstrated in the pathologic sections. Coats considered this case the key to his other cases. He considered the mound to be, in all probability, a subretinal hemorrhage. This writer believes that this hemorrhagic mound originated in deep telangiectatic vessels in the outer layers of the retina and in those areas where vessels appear to grow into this mound from the outer surface of the retina we actually visualize the remnants of deep telangiectatic vessels. These deep vessels became partially obliterated initially by the hemorrhage. Subsequent organization completed the process of obliteration.

Examination of case four revealed what appeared to be two subretinal mounds, one above the disc and one above the macula. Only one half of the eye, however, was available to Coats for pathologic examination. Again, as in case one, a cavity was filled with debris but the process of organization was much more advanced. An extensive sheet of fibrous tissue was found between the retina and the choroid, stretching from the optic nerve to the ora serrata. This case can be interpreted as one of localized deep telangiectasia forming the subretinal mounds, resulting in extensive deposits of exudate throughout the entire fundus. The organization of this exudate resulted in the formation of the sheet of fibrous tissue between the retina and the choroid.

Case number eight is that of a 12-year-old boy with subretinal mounds and widespread yellow exudate, noted ophthalmoscopically. The subretinal mounds were similar to those in case one but were of greater duration since ossification had occurred.

In case nine, involving the right eye of a 7-year-old girl, two subretinal mounds were noted. One was in the macular area and temporal to it and a second one with more elevation was in the temporal half of the eye. The pathology report noted little change in the vessels. The lesion in the macula in this case was similar to the

macular lesion in case number three. The second mound in case nine, occurring temporally, was much less organized and was similar to the mound described in case one. In my opinion, this represents a case of deep retinal telangiectasia with the secondary lesion at the macula similar to the macular lesion in case number three.

In this original group we find one case of superficial telangiectasia with a secondary macular lesion. The remaining cases represent examples of both superficial and deep telangiectasia with subretinal mound formation. The subretinal mounds originate in hemorrhage from the deep telangiectatic vessels. The hemorrhage follows damage to the distended vessel walls as the bulk of the abnormal vasculature increases. This space occupying lesion, within the retinal tissue increases the anoxia in an already stagnant environment, leading to further damage to the walls of the distended and engorged blood vessels. Not all hemorrhage need originate from the same vessel, nor at the same time. Repeated hemorrhages would create a mass with localized swellings or bosses upon its surface as in Coats' case number five or case #26 of the present series.

It has been stated in the literature that Coats changed or modified his views, expressed in his initial article.[48] Apart from the fact that in his third paper he does not divide the cases into groups, such a view is incorrect. In his first article, Coats acknowledged prior reports of this disorder and stated his opinions unambiguously. An examination of his first article reveals the following:

1. Speaking of the nature of the condition, he states, "The first authentic and fully reported pathological examination which I have been able to find is that of Treacher Collins (1889). Possible instances before that date are those of Brailey (1876) and of Cros (1884)."

2. Coats did not reclassify Group 3 of his cases in 1912 since he recognized about eighteen recorded cases of retinal angioma. He acknowledged the first angioma as that of Fuchs, reported in 1882.[49] He noted the frequent bilaterality of this lesion and its familial nature, as first reported by Treacher Collins.[50] Coats said of Group 3, "The best examples of cases watched over a prolonged period are those of von Hippel."

3. Coats left us in little doubt about his view of Group 3 when in his initial paper he stated, "For the present, therefore, it seems best to classify them separately and to accept the cases in group three as examples of a genuine simple tumour of the retina."

4. Speaking of Group 1, he stated, "The explanation which appears to be best suited to account for these cases is that they are due to the

slow organization of haemorrhage." He pointed out that repeated clinical observations of Nettleship, Gunn, and Weinstein supported this view. He stated further, "If these instances are not due to primary vascular disease (*vide infra*), one must fall back on some obscure general condition, possibly connected with alterations in the coagulability of the blood." The suggestion of altered blood coagulability as an etiologic factor in this disorder then is not new.[51]

With regard to Group 2, Coats stated:

1. "Perhaps therefore the haemorrhage is due in some instances to an unknown form of vascular disease, possibly founded on a congenital vulnerability of the vessel wall."

2. In commenting upon the masses in the fundus, the subretinal mounds, he said, "These cases have, however, rarely or never been seen in their earliest stages. Little seems to be known of the ophthalmoscopic appearance of a subretinal haemorrhage, but admitting that it would appear red through normal retina, it must be remembered that the retina in these cases is not normal; it probably becomes oedematous almost immediately and within a short time is thickened and filled with crowds of swollen leucocytes (Figure 2), which hide all details."

In his 1911 article, Coats suggested "Retinitis haemorrhagica externa" as an alternative to "Forms of retinal disease with massive exudation." Both titles head the 1912 article. Further, in his 1911 commentary he stated, "If this be the correct explanation, the disease evidently affects chiefly the peripheral, and perhaps chiefly the capillary circulation; for the haemorrhage seems to occur first in layers only supplied by capillaries . . . it seems certain therefore that ophthalmoscopically recognizable vascular changes are not a necessary precursor of the disease. It may be that changes are indeed present, but that they are too subtle to be detected with the ophthalmoscope or even with the microscope."

In this 1912 article, Coats did not divide his cases into groups nor, by the same token, did he add anything new concerning any aspect of this condition.

## CASE REPORTS

CASE 1

A 3½-year-old child was first examined on March 1, 1962. Examination of the right eye, under general anesthesia, showed a total detachment of the retina. Over the inferior half of the retina abnormal vascularization consisted of telangiectatic vessels, micro- and saccular aneurysms. In the inferior nasal

quadrant, a tumor-like mass was present. Widespread aggregates of exudate were present in the retina and a sheet of dark yellow exudate was seen in the posterior polar region. On April 11, 1973, the right eye was enucleated and the diagnosis of Coats' disease was confirmed. The patient's father had a right-sided nevus flameus which did not involve the eye.

CASE 2

A 45-year-old man was first examined on June 13, 1964. One year previously a diagnosis of angiomatosis retinae of the left eye was made. The corrected vision was 20/15 right eye and 20/200 left eye. The left eye showed a circinate figure of exudate in the macula. In the periphery, between the 2:15 and 7:00 o'clock position, telangiectatic vessels were present. Miliary aneurysms were noted both anterior and posterior to the equator between the 3:30 and 6:00 o'clock position. The vascular changes involved both the arterioles and veins. Many small sheathed vessels were noted distal to the abnormal vasculature. The right eye was normal.

The left eye was treated with xenon arc photocoagulation twice in 1964, once in 1965, and a final treatment was performed in 1968. Thereafter, the condition of the left eye was considered stable and examination at six month intervals showed no recurrence. When examined on December 23, 1974, more than ten years after the initial treatment, the vision in the left eye was 20/70. The central mound of exudate was now replaced by a whitish fibrotic plaque. Between the 12:30 and 2:00 o'clock position a network of dilated vessels was present anterior to the equator. Fluorescein studies confirmed the recurrence of the telangiectasia.

CASE 3

A 10-year-old boy was first examined on September 5, 1964. The corrected vision in the right eye was 8/400 and that in the left eye 20/30. In the right eye the retina was elevated between the 5:15 and 1:00 o'clock position. Abnormal vessels between the 7:30 and 11:15 position, at about the level of the equator, consisted of dilated veins and arterioles, telangiectatic vessels and multiple arteriovenous shunts. Exudate throughout the area of elevated retina was most dense in the region posterior to the abnormal vessels. The entire macula was occupied by a circular, elevated mound of light yellow exudate, similar in color to that in other areas of the fundus. Both temporal veins were markedly dilated.

Peripheral telangiectasia was present in the left eye. The macula was normal (Figure 7).

Xenon arc treatment of both eyes was performed on several occasions over the next five years. In the right eye the macular exudate absorbed completely but what at first appeared to be a cyst became a nubbin of glial tissue. When examined on April 21, 1972, the corrected vision in the right eye was 20/25 and that in the left eye 20/20. In May, 1973, recurrences were treated in both eyes. On September 9, 1974, the corrected vision in the right eye was

20/25 and that in the left eye 20/20. No abnormal vessels were present in the right eye and the glial nubbin had become completely flat and covered with black pigment. Fluorescein studies, performed on the right eye in December 1974, revealed miliary aneurysms in the macula, indicating a recurrence of the telangiectasia after 10 years (Figure 8).

CASE 4

A 41-year-old man, referred with a diagnosis of Leber's miliary aneurysms of the retina in the right eye, was first examined on December 29, 1964. Examination of the left eye did not reveal any abnormalities. On January 26, 1965, the central macular area was treated with the xenon arc machine. For the next eight years the right eye remained unchanged. The vision remained at 15/400. When examined on April 3, 1974, telangiectatic vessels were seen in the temporal periphery of the right eye, and fluorescein angiography in September 1974, confirmed the presence of the abnormal vasculature (Figure 9).

CASE 5

An 8½-year-old girl was first examined on March 4, 1965. She was referred with a diagnosis of Leber's miliary aneurysms in the right eye. The corrected vision was 20/20 in each eye. In the right eye there was an area of miliary aneurysms and telangiectatic vessels slightly anterior to the equator between the 7:30 and 10:30 o'clock position in association with a stellate configuration of light yellow exudate. Treatment was advised but the parents were told there was no urgency as it was unlikely the lesion would change rapidly. When the child was reexamined, however, on June 8, 1965, the area of telangiectasia had definitely increased and a fresh area of abnormal vasculature was noted anterior to the equator in the 8:30 meridian. In June 1965, xenon arc treatment was performed on the right eye. Ten years later there was no recurrence of the lesion and the vision remained at 20/20.

CASE 6

A 5-year-old boy was first examined on June 15, 1965. He was referred with a diagnosis of Coats' disease, right eye. Examination of the left eye was negative. In the right eye the retina was elevated from the 7:00 to 2:00 o'clock position. The upper temporal quadrant showed extensive abnormal vasculature consisting of telangiectatic vessels with miliary and saccular aneurysms. Extensive sheets of deep yellow exudate in the temporal and posterior polar areas involved the entire macula, reaching the temporal edge of the disc. Various forms of treatment performed over the next three years resulted in failure. Subretinal mounds, not seen initially, became apparent following the attempted treatment.

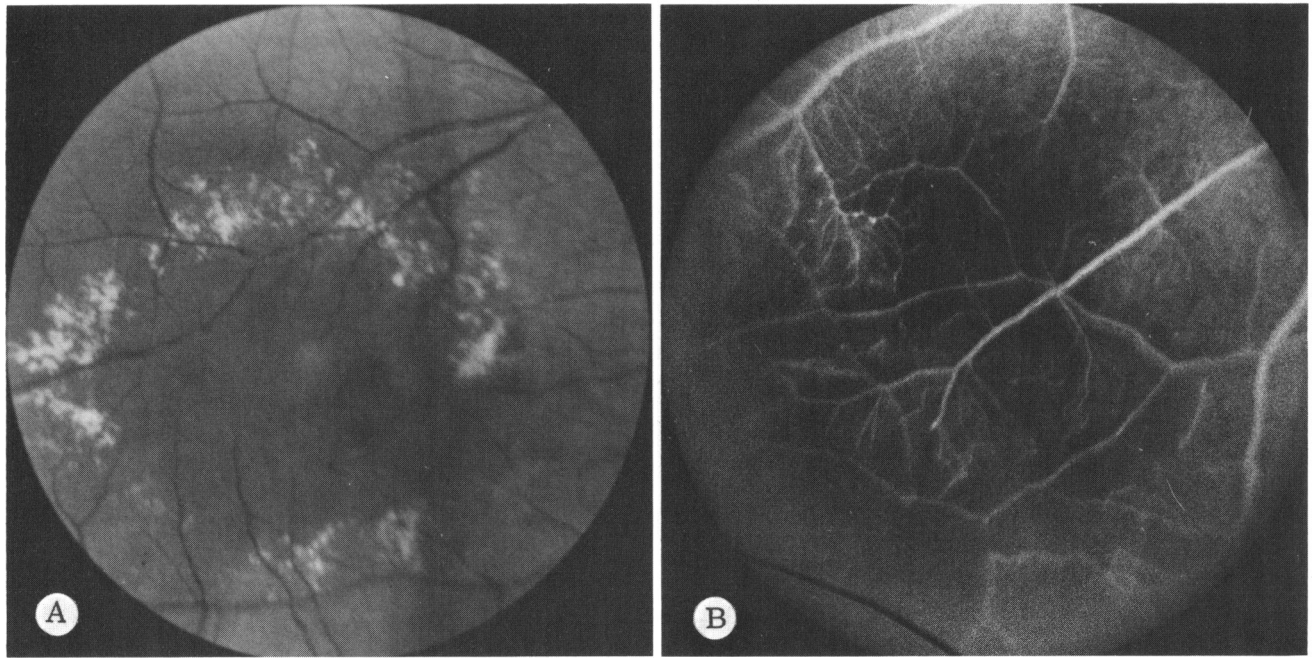



FIGURE 9

A: "Leber's miliary aneurysms" in macula surrounded by exudate. Case 4, O.D. B: Case 4, O.D., showing peripheral telangiectasia which appeared 10 years after treatment of macular lesions.

CASE 7

A 12-year-old boy was first seen on September 16, 1965. The corrected vision in the right eye was 20/20 and that in the left eye 20/20. In the right eye the retina was elevated between the 6:00 and 1:15 o'clock position in the periphery. Between the 7:00 and 10:00 o'clock position the elevation extended posterior to the equator. Extensive exudate was noted along the posterior border of the elevated area. Abnormal vasculature was present only in the area of elevated retina and, for the most part, anterior to the equator. The abnormal vessels consisted of dilated veins and arterioles, arteriovenous shunts, foci of dilated capillaries, with vessels showing beading and sausage-shaped components. The macula was normal with a good foveal reflex. Repeated treatment was performed in 1965 and 1966. On April 2, 1968, no abnormal vessels or exudates were seen. The vision was 20/20, corrected. In December 1974, fluorescein studies showed the condition had recurred, after more than nine years.

CASE 8

A 16-year-old youth was found to have retinal telangiectasia in the course of a routine examination. He was first examined on November 5, 1965. The corrected vision in the right eye was 20/20 and that in the left eye also 20/20. The right eye was normal. The left eye had abnormal vessels in all quadrants, most marked between the 12:00 and 5:00 o'clock positions, at the level of the equator. These abnormalities consisted of sausage-like dilatations, large and small, berry and saccular aneurysms, with arteriovenous shunts and accentuation of the capillary network. The left eye was treated repeatedly over the next four years. When last seen on October 12, 1971, the corrected vision was 20/30 and no abnormal vasculature was present in the fundus. The previous accentuation of the capillary network in the macula had disappeared.

CASE 9

A 7-year-old girl was referred with a diagnosis of retinal degeneration, left eye. At about three years of age, her parents were told the child had "scar tissue in the left eye." She was first examined on July 24, 1965. The corrected vision right eye was 20/20 and that in the left eye counting fingers at two feet. The right eye was normal. A marked left exotropia was present. In the left eye a large, high mound of exudate, conical in shape and whitish-yellow in color, was present in the macula. A wide ring of exudate, which followed the arcs of the temporal vessels, surrounded the central lesion. At about the level of the equator, between the 1:00 and 6:00 o'clock positions, was a band of abnormal vasculature consisting of angiomas, saccular aneurysms, arteriovenous shunts, telangiectatic vessels, and microaneurysms. The telangiectatic vessels varied in size from massive, sausage-like dilatations to networks of fine arborizing vessels. Little or no exudate was seen in the immediate vicinity of the abnormal vasculature.

Treatment with the xenon arc machine was performed on four occasions. The mound in the macula absorbed and flattened considerably. On March 19, 1968, the corrected vision in the left eye was 20/400. Three years later this eye was free of abnormal vessels and was straight. The macular lesion continued to shrink slowly. On February 16, 1973, the corrected vision was 20/300. A fresh area of new vessel formation was noted in the three o'clock meridian posterior to the scar. This area was treated on March 6, 1973. On March 15, 1974, the vision was still 20/300.

CASE 10

A 10-year-old girl was first examined on September 23, 1966. The corrected vision was 20/50 right eye and 20/20 left eye. The left eye was normal. In the right eye abnormal vasculature was present in elevated retina between the 6:30 and 10:00 o'clock positions (Figure 3). The retinal elevation extended posterior to the equator and was rimmed posteriorly by a wide band of exudate. An oval intraretinal cyst, approximately three disc diameters in its long axis, was present astride the equator in the 7:15 meridian. This cyst was surrounded by a closely applied ring of exudate. The telangiectasia consisted of dilated veins and arterioles, arteriovenous shunts with beading and sausage-shaped elements. In many areas, adjacent to the ora serrata, the abnormal vessels were heavily sheathed and white loops of what appeared to be "ghost vessels" or shrunken telangiectatic channels were present. Marked dilatation of the inferior temporal vein extended as far as the disc. Fine pigment stippling was present in the macula and a small glial nubbin was noted just below the fovea.

The right eye was treated on four occasions with the xenon arc machine. On September 20, 1974, the corrected vision was 20/25. No abnormal vessels were present in the eye. The small glial nubbin, noted at the initial examination, persisted, though smaller in size. The inferior temporal vein was sheathed right up to the disc. This vein was "flat" and easily collapsible.

CASE 11

A 19-year-old man was found to have widespread abnormal retinal vessels in his left eye on routine examination. He was first examined on May 27, 1967. The corrected vision in the right eye was 20/20 and that in the left eye also 20/20. The right eye was normal. In the left eye, in the macular area, were myriads of miliary aneurysms (Figure 5A). No hemorrhages or exudates were noted in the fundus. The disc was hyperemic but otherwise normal. At the level of the equator, in all quadrants, extensive telangiectasia was noted, most marked between the 12:00 and 6:00 o'clock positions. In the latter area, the telangiectatic vessels consisted of numerous loops of dilated sausage-like channels in the form of arcades bowed toward the ora serrata. Definite arteriovenous shunts were present. The retina anterior to these large arcades was totally avascular. Groups of large aneurysms were seen in the 8:45 and 11:00 o'clock meridians. Between the 6:00 and 7:00 o'clock and between the 8:30 and 12:00 o'clock positions the abnormal vasculature was similar to but much

less florid than that in the temporal fundus. Fluorescein angiography showed extensive leakage from all abnormal vessels.

On June 23, 1967, the left eye was treated with the xenon arc machine. Treatment was repeated on July 12, 1967. Areas in both the macula and temporal half of the fundus were treated. Treatment was again carried out on July 31 and November 24, 1967. Over the next two months many areas of new vessel formation appeared in the regions adjoining the treatment scars. It was felt that aggressive treatment would be required if the eye was to be saved. The treatment seemed to trigger the rapid formation of new vessels and the question arose: was it correct to treat the eye in the first place? Treatment was repeated six times over the next four years. In October 1974, the corrected vision was 20/80 in the left eye and further treatment was planned. In March 1975, early telangiectasia in the temporal periphery of the right eye was noted for the first time.

CASE 12

A 30-year-old woman was first examined on May 23, 1967. The corrected vision in the right eye was 20/25 and that in the left eye 20/200. She was referred with the diagnosis of retinal angioma, left eye. The right eye was normal. In the left eye a large angioma was flanked by miliary aneurysms and telangiectatic vessels. Following hospitalization, renal arteriography demonstrated an anomalous left renal artery. Over the next year the angioma in the left eye was treated with the xenon arc machine. All tumor tissue was completely obliterated. The eye remained free from abnormal vasculature until February, 1975, when an area of telangiectasia was demonstrated on fluorescein angiography in the upper nasal quadrant outside the area of previous treatment. The examination of the right eye at that time, while not showing any definite areas of telangiectasia, did show some suspicious areas. The patient had complained of headaches for the last three months. She was now also found to have a mass in the right lobe of her thyroid gland.

CASE 13

A 39-year-old woman was first examined on June 27, 1967. She was referred with a diagnosis of retinal degeneration, right eye. She had been treated for repeated episodes of uveitis in both eyes. The corrected vision was 20/20 right eye and 20/20 left eye. In the 5:00 o'clock meridian in the right eye was an angioma about three-fourths disc diameter in size. Between the 7:00 and 9:00 o'clock meridians, a ridge of elevated retina was surmounted by a row of abnormal vessels consisting of telangiectasia and berry aneurysms. Two small angiomas in the 11:00 and 12:00 o'clock meridians were interconnected by telangiectatic vessels. Duplication of the inferior temporal vein for a short distance along its course was interpreted as a developmental anomaly. Fluorescein angiography showed extensive leakage from all abnormal vasculature. The left eye was normal. Xenon arc treatment of the right eye was performed on four occasions. On September 25, 1968, the vision was 20/40 right

eye and 20/40 left eye. All areas of treatment in the right eye were flat but marked posterior polar edema and bilateral uveitis were present. The right eye gradually deteriorated over the next few years. On March 12, 1974, a fibrotic retinal detachment was present in the eye.

CASE 14

A 43-year-old man was first examined on September 17, 1968. He was referred with a diagnosis of vitreous hemorrhage, retinal detachment, right eye. The corrected vision right eye was hand movement and that in the left eye 20/20. In the right eye the retina was almost totally detached. An area of preretinal hemorrhage, about three and a half disc diameters in extent, partially obscured the disc and macula. Just at the upper pole of the hemorrhage, a dark, raised area, about one disc diameter in size, was interpreted as a retinal angioma. In favor of an angiomatous process in the retina was the presence of extensive dilatation of the small vessel network in the area of flat retina, as well as in the elevated retina in the inferior temporal quadrant. Most significantly, the inferior nasal vein was seen to be markedly dilated. Examination of the left eye revealed a fresh horseshoe tear in the 11:00 o'clock meridian and a fresh oval hole in the 12:30 o'clock meridian. At the level of the equator, in the 2:30 o'clock meridian, was an area of accentuation of the capillary network with a few miliary aneurysms. Hemorrhage in this area was considered to be the result of a recent vitreous detachment. The subsequent course of the right eye confirmed the presence of an angioma. An anomalous arrangement of the retinal vessels in the right eye was similar to the case of Story and Benson.[52] Fluorescein studies confirmed these findings.

Following successful retinal surgery on October 7, 1968, all areas of retinal telangiectasia were treated with xenon arc photocoagulation over the next seven months. The tears in the left eye were treated on September 30, 1968. It was ascertained that fundus photographs of the right eye had been taken in 1958. An experienced observer had recorded his description of the right eye in 1958: "It is so rare that I must say I have never seen it before, at least the vascular configuration in the vessels at the posterior pole of the right fundus." Much further detail was given, including the comment, "The aneurysms in the periphery at several points, especially nasal and below and down and temporally, are also amazing and there seem to be some new-formed vessels, as you stated." No mention was made of the obvious angioma now present above the fovea. The retina in the right eye redetached two years and one month later and has remained so until the present time. There has been no change in the telangiectatic vessels in the left eye.

CASE 15

A 12-year-old girl was examined on October 14, 1968. The corrected vision in the right eye was hand movement and that in the left eye 20/20. The right fundus had a yellow exudate (Type II) throughout most of the posterior polar region. Multiple, dome-like elevations of the retina, interpreted as subretinal

mounds, were noted in all quadrants. The mounds were covered with exudate similar to that in the posterior polar area. Networks of telangiectatic vessels with saccular aneurysms showed brilliantly against the exudate. Scattered areas of shimmering crystalline deposits were present. The left eye was normal.

Attempted xenon arc treatment was a failure. The patient was lost to follow-up until January 14, 1975, when she was located as a member of a family under study by the writer. On that date, examination of the right eye showed no light perception. Apart from moderate lens changes, the anterior segment was normal. The intraocular tension was normal. Only in the inferior temporal quadrant was there any semblance of the original fundus picture. In this region a few telangiectatic vessels coursed over a yellow-grey mass. Many heavily sheathed or obliterated vascular channels were visible. In the rest of the fundus the retinal vessels were markedly attenuated and the extensive exudate and subretinal mounds were replaced by white fibrotic tissue. Thus, after an interval of more than six years, the original florid picture was replaced by one of atrophy. The left eye remained normal.

CASE 16

A 47-year-old man was first examined on March 17, 1969. The corrected vision in the right eye was 20/100 and that in the left eye 20/20. The right eye showed a marked corneal dystrophy. Three separate circinate figures were present in the fundus, one just above the macula, a second nasal to the disc, and a third in the inferior temporal quadrant astride the equator. In the center of each circinate figure the retina was thickened and edematous. Miliary aneurysms and a few berry aneurysms with a few telangiectatic vessels were present within each figure. Examination of the left eye was normal. Following three treatments with the xenon arc machine, all exudate and abnormal vessels disappeared. On May 2, 1970, the corrected vision in the right eye was 20/40 and no abnormal vessels were seen. On December 2, 1970, the patient died. The diagnosis was subarachnoid hemorrhage. An autopsy was not obtained.

CASE 17

A 43-year-old woman was first examined on June 13, 1969. The corrected vision in the right eye was 20/40 and that in the left eye 20/20. Examination of the left eye was normal. In the right eye a small amount of fresh blood was present in the inferior vitreous. A recent detachment of the vitreous had occurred. In the inferior temporal quadrant, astride the equator, between the 6:30 and 8:00 o'clock meridians, a conglomeration of abnormal vessels was noted. The abnormal vasculature consisted of telangiectatic vessels, saccular aneurysms, and arteriovenous shunts. The retina in the immediate vicinity of these structures was edematous. The abnormal vasculature was bounded posteriorly by a semicircle of exudate. The disc was normal. Following treatment with the xenon arc machine, all abnormal vessels and exudate

disappeared. On February 20, 1975, the eye had no abnormal vessels and the corrected vision was 20/20.

CASE 18

An 18-year-old girl was first examined on June 17, 1969. She was referred with a diagnosis of Coats' disease, right eye. There was no light perception right eye, and normal vision in the left eye. The right eye showed a total detachment of the retina with yellow exudate and scattered deposition of what appeared to be cholesterol crystals. In both temporal quadrants tumor masses were visible. A tumor mass in the immediate vicinity of the disc was encroaching upon it. These masses were interpreted as subretinal mounds. Examination of the left eye was normal. Over the next five years a mature cataract developed in the right eye. The eye remained white, normotensive, and free of rubeosis iridis.

CASE 19

A 38-year-old man was first examined on September 3, 1969. Retinal hemorrhages and circinate figures had been noted in the fundus of the left eye. The corrected vision in the right eye was 20/20 and that in the left eye 20/25. The right eye was normal. In the central macular area in the left eye, the retina was cystic and multiple aneurysms were noted. Astride the equator, between the 12:45 and 2:30 o'clock meridians, telangiectatic vessels were present. Centered on the 3:30 o'clock meridian, posterior to the equator, was a broken circinate figure. Within this ring was loops of telangiectatic vessels, a few saccular aneurysms and what appeared to be a few tiny solid angiomas. Centered on the 5:00 o'clock meridian was a second circinate figure surrounding a cluster of bulbous aneurysms. A third circinate figure was located at the same level in the 6:00 o'clock meridian. In the 8:00 o'clock meridian, at about the level of the equator, was a small, solid angioma about one-third disc diameter in size. The retina in the vicinity of the tumor was normal. It was felt that with destruction of all abnormal vasculature outside the macular region, the aneurysms in the macular region might regress.

In September 1969, xenon arc treatment was performed. Treatment was repeated twice in 1970. On November 11, 1970, the corrected vision was 20/25. With the exception of the macular area, all abnormal vessels and exudate had disappeared. The cystic condition of the macula was unchanged. It was felt that perhaps the miliary aneurysms were less in number (Figure 10A). On February 13, 1971, the corrected vision remained at 20/25; however, for the first time it was thought that perhaps treatment of the macular region with the argon laser would have to be undertaken as there was no evidence of regression of the pathology in this area. Fresh miliary aneurysms, as well as definite accentuation of the capillary bed, were noted in the upper nasal quadrant. It was considered most likely that this area too would eventually call for treatment. On May 22, 1972, the state of the macula seemed unchanged. A few fresh miliary aneurysms were noted in the 2:00 o'clock

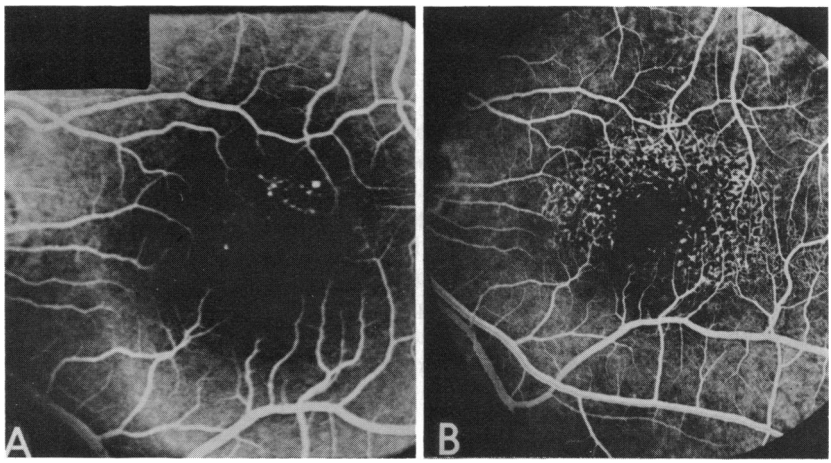



FIGURE 10
Case 19, O.S. A: Angiogram showing a few miliary aneurysms in the macula; B: Angiogram five years later showing generalized central telangiectasia.

meridian. On January 19, 1974, the corrected vision in the left eye was 20/30. The patient was advised to consider treatment of the macular region, as had been recommended by both the writer and a consultant in 1971; however, fluorescein angiography, performed on October 16, 1974, demonstrated an extensive meshwork of telangiectatic vessels with microaneurysm formation in the central macular region (Figure 10B). Treatment was deferred.

CASE 20

A 45-year-old man was first examined on December 10, 1969. The corrected vision was 20/20 right eye and 20/200 left eye. Examination of the right eye showed darkening and dilatation of the retinal veins with mild arteriovenous nicking and minimal narrowing of the arterioles. The left eye showed widespread retinal edema with abnormal vasculature throughout the entire fundus, about equally distributed in all quadrants, both anterior and posterior to the equator. The abnormal vessels were predominantly miliary and saccular aneurysms, with scattered telangiectatic vessels and arteriovenous shunts. Large saccular aneurysms were present on the arterioles. Extensive exudate with circinate figures was noted. The retinal veins were uniformly dilated and darkened without any regional accentuation of these changes. Over the next three years xenon arc photocoagulation was performed on the left eye on seven occasions. On February 26, 1973, a small posterior polar cataract was noted. The macula was unchanged. When seen on September 11, 1974, occlusion of the superior temporal vein in the right eye had occurred. The corrected vision in the right eye was 20/40 and that in the left eye 20/200.

Fundus study of the left eye showed new vessels in the periphery and at the 10:00, 11:00, and 12:00 o'clock meridians. The vessels in the macula had remained unchanged since June, 1972. Heavy sheathing of the superior nasal arteriole was now present but there was no indication of occlusion of this vessel. In addition, there was slight but definite accentuation of the capillary and small vessel network around the disc. No exudate was present in the fundus.

CASE 21

A 27-year-old woman was first examined on December 10, 1959. The corrected vision in the right eye was 20/20 and that in the left eye 20/30. The right eye was normal. Ophthalmoscopic examination of the left eye showed an angioma about one disc diameter in size with classical feeding and draining vessels at the ora serrata in the 6:00 o'clock meridian. Between the 4:30 meridian and the edge of this angioma was a series of six smaller angiomas, also at the ora serrata, interspersed with a few saccular aneurysms. The saccular aneurysms and small angiomas were interconnected by telangiectatic vessels. Between the 3:45 and 7:30 meridians, the retina was slightly elevated. From the temporal end of the area of elevated retina, a wide band of yellow exudate (Type I) coursed posteriorly to a point 1.5 disc diameters from the fovea, thence, to the ora, ending in the 7:00 o'clock meridian. Radial striae were present in the perifoveal area. Following xenon arc treatment of the angiomas on December 12, 1969, the retina became quite elevated, so much so that it was thought retinal surgery would be required; however, the retina flattened and on January 6, 1970, the entire area of abnormal vasculature was treated with cryotherapy. Additional cryotherapy was applied on March 5, 1970. Definite retinal contracture appeared below the disc, producing marked kinking of the retinal vessels. When seen on June 19, 1970, extensive vitreous hemorrhage obscured inferior fundus detail. On October 23, 1970, the vitreous had cleared and the retina was flat throughout and devoid of abnormal vasculature. The macula was clear. The corrected vision was 20/30. Four years later the patient reported she "could read quite well" with her left eye.

CASE 22

A 62-year-old woman was examined on April 20, 1970. The corrected vision in the right eye was 20/20 and that in the left eye light perception. A typical racemose aneurysm was present in the left eye (Figure 11). The right eye showed an area of telangiectasia between the 7:30 and 9:00 o'clock positions. The patient has not been available for follow-up.

CASE 23

A 49-year-old man, referred for evaluation of the right eye because of retinal hemorrhages, was first examined on February 8, 1971. The corrected vision in the right eye was 20/40 and that in the left eye 20/30. Examination of

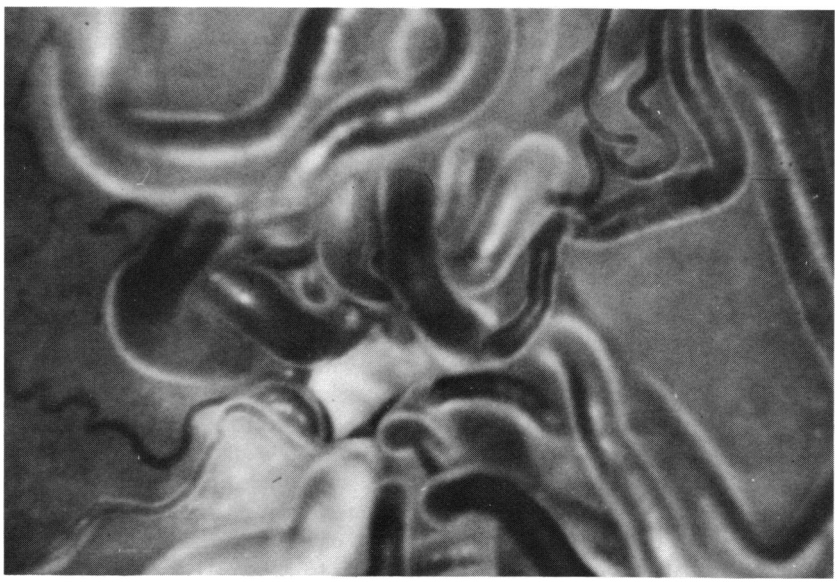


FIGURE 11
Case 22, O.S. Racemose aneurysm of retina.

the left eye was negative. The right eye showed retinal edema with numerous miliary aneurysms within the arcs of the temporal vessels. A tiny circinate figure surrounded a few aneurysms just above and temporal to the fovea. The disc was normal. Observation of the right eye was decided upon. No change was noted until mid-October, 1971, when the visual acuity improved to 20/30, corrected. The central macular region was less edematous. The number of miliary aneurysms had increased. Fluorescein angiography showed slight leakage from these structures. No leakage was noted in the immediate vicinity of the fovea. On January 31, 1972, the number of microaneurysms had increased. Areas of early telangiectasia were now present. The exudates had all but disappeared. On April 24, 1972, the corrected vision in the right eye was 20/40. The abnormal vasculature was unchanged but all exudates had disappeared. By July, 1972, exudates were again seen in the macular region and miliary aneurysms were noted in relation to both the veins and arterioles. On October 9, 1972, the corrected vision in the eye was 20/25-1. All exudates had cleared. On July 2, 1973, the posterior polar area was unchanged. On April 22, 1974, the corrected vision in the right eye was 20/40. There was no significant change in the number or distribution of the microaneurysms or in the extent of the retinal edema. Some of the miliary aneurysms could now be classed as berry aneurysms. On February 24, 1975, the corrected vision in the right eye was 20/30. Fluorescein angiography performed on

March 3, 1975, demonstrated a marked reduction in the number of miliary aneurysms with regression of the telangiectasia in the macula.

CASE 24

A 51-year-old man was first examined on October 26, 1971. The corrected vision in the right eye was 20/20 and that in the left eye 20/20. The right eye was normal. In the left eye, in the 6:30 meridian, anterior to the equator, was a vascular structure about two and a half disc diameters in size which appeared to be an angioma. Careful examination, however, showed absence of typical feeder vessels with the draining veins. In addition, the color of the mass was not uniformly red but greyish-white with large bulbous vessels on its surface. A typical angioma is covered with a diffuse network of fine vessels, whereas the surface of this structure presented large telangiectatic vessels. The lesion was interpreted as an example of an early stage of superficial and deep telangiectasia. On November 9, 1971, the lesion in the left eye was treated with xenon arc photocoagulation. Over the next 12 months, regression of the abnormal vasculature continued but a large central vein persisted. This vessel was the main drainage channel in the original lesion. When last seen on February 10, 1975, the vision in the left eye remained at 20/20. The large vein described above persisted and appeared to connect with deeper vessels in the retina. Further treatment is planned.

CASE 25

A 47-year-old man was first examined on February 14, 1972. The referring diagnosis was Coats' disease right eye. The patient had been seen by an ophthalmologist in 1967, because of black spots in the right eye. At that time localized areas of retinal elevation with what was apparently telangiectasia, were noted in the right eye. Large areas of yellow exudate were described. The left eye was normal. There was a subtotal retinal detachment in the right eye with two large subretinal mounds superiorly and one inferiorly. Treatment with the xenon arc machine was a failure.

CASE 26

A 21-year-old man, referred with a diagnosis of Coats' disease left eye, was first examined on January 19, 1973. The corrected vision right eye was 20/20 and left eye 20/200. The right eye was normal. In the left eye low elevation of the retina extended from the 12:45 to the 9:00 o'clock meridian, well beyond the equator, and was bordered by a wide sheet of yellow exudate (Type II), which reached almost to the horizontal meridian. Temporally between the 1:00 and the 4:30 o'clock meridians, telangiectatic vessels, miliary aneurysms, and arteriovenous shunts were seen. In the 4:30 meridian, at the equator, was a large tuft of retinitis proliferans. Between the 6:00 and the 7:30 o'clock meridians, a subretinal mound appeared to arise in the choroid and extended from the ora serrata to behind the equator. It was surmounted by a conical, dome-like knob. The elevated area was studded with

miliary aneurysms and telangiectatic vessels. The disc was hyperemic but otherwise normal. The inferior temporal vessels, especially the vein, were markedly enlarged. Fluorescein angiography showed massive leakage from all abnormal vessels. The elevated mound transilluminated readily. Ultrasonography confirmed the presence of a solid mass in the inferior fundus. The consistency was irregular and the possibility of calcified areas within the mass was mentioned. Following successful obliteration of all telangiectatic vessels in the upper half of the fundus, an attempt was made to wall off the area of the subretinal mound. Sequestration of the lesion with a wide barrier of scar tissue would be followed by direct treatment of the mound; however, a localized inferior retinal detachment occurred before the subretinal mound could be surrounded completely. Over the next two years the sheet of exudate in the posterior pole became converted into whitish fibrous tissue. The subretinal mound gradually shrank. When seen on March 11, 1975, the vision in the left eye was hand movement. Lens changes were evident. The subretinal mound in the inferior fundus was less than one-quarter its initial size. It had lost its solid, tumor-like quality and now appeared translucent. A single retinal vein entered the summit of the mound as if connecting with deeper vessels.

CASE 27

A 34-year-old woman had been told she had an angioma in her left eye and was advised to have it treated. When examined on March 1, 1973, the corrected vision in the right eye was 20/20 and that in the left eye 20/20. Examination of the right eye was normal. The left eye had an area of telangiectatic vessels astride the three o'clock meridian between the equator and the ora. Just posterior to the vessels was an area of light yellow colored exudate. The abnormal vasculature showed definite shunts. A small area of lattice degeneration was present in the 12:00 o'clock meridian. The patient was not available for follow-up.

CASE 28

An 18-year-old girl was first examined on March 12, 1973. A diagnosis of Coats' disease had been made in the right eye and treatment with the argon laser had been performed on one occasion. The vision in the right eye was hand movement and that in the left eye 20/20, corrected. In the macula of the right eye, the center of a large mound of exudate was occupied by a globular whitish structure to which the vitreous was adherent. Retinal edema was present throughout the posterior polar area temporal to the disc. In the periphery between the 5:00 and 10:00 o'clock meridians the retina was elevated. Between the 6:30 and 9:30 meridians, abnormal vasculature consisted of telangiectatic vessels, miliary aneurysms, and dilated capillaries at the level of and anterior to the equator. Exudate was scattered throughout the involved region. Discrete areas of pigmentation, interspersed with the abnormal vessels, were interpreted as laser burns. On March 30, 1973, xenon

arc photocoagulation was performed. Since that time all abnormal vessels have slowly disappeared and the central macular mass has continued to shrink. Vitreous traction on the central glial nubbin in the macula has persisted.

CASE 29

A 25-year-old man was first examined on March 28, 1973, as a member of a family under study. His right eye was blind since infancy. The vision in the left eye was 20/20 without correction. Telangiectasia was present astride the equator between the 10:30 and 4:30 o'clock positions. No retinal edema or exudate was noted. The telangiectatic vessels were clearly shown in the fluorescein angiogram performed on December 10, 1974. Arteriovenous shunts were present (Figure 12C). Along the course of the inferior temporal arteriole, about 1¾ disc diameters from the disc margin, a cluster of miliary

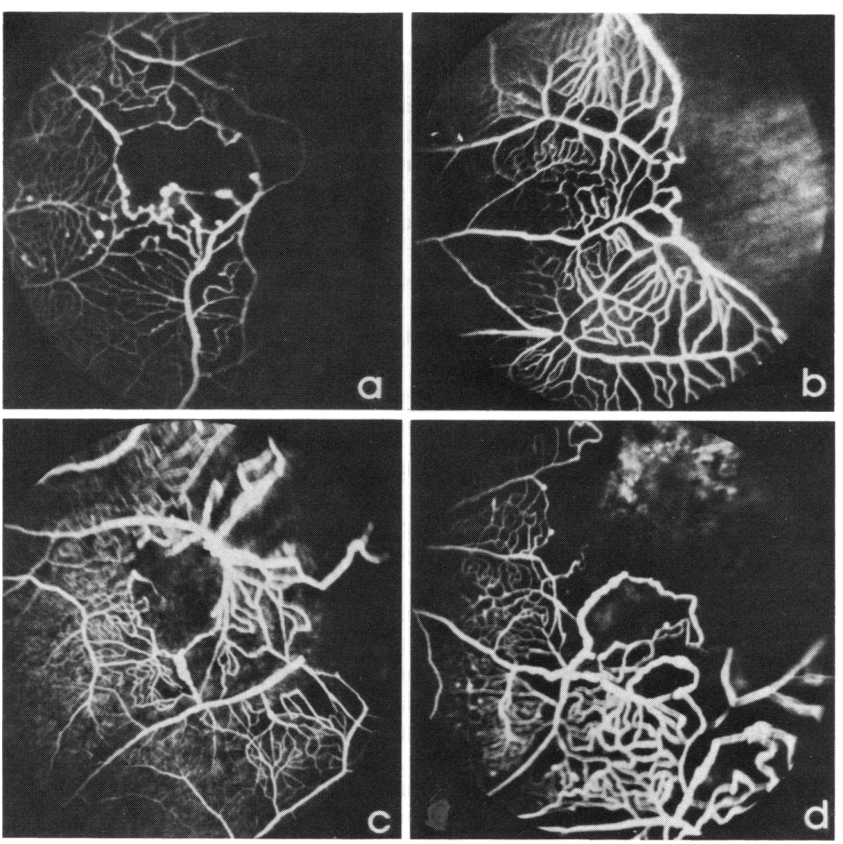



FIGURE 12

Familial peripheral telangiectasia. A: Case 43, O.S. B: Case 42, O.S. C: Case 29, O.S. D: Case 36, O.S. See Table III.

aneurysms was seen. The last examination on January 24, 1975, showed definite advance in the telangiectasia. This one-eyed patient remains under close observation in anticipation of treatment.

CASE 30

A 16-year-old girl was referred with the diagnosis of Coats' disease, left eye, made in October, 1973. Treatment with the argon laser had been attempted on one occasion. She was first examined on January 23, 1974. The corrected vision in the right eye was 20/20 and that in the left eye 20/400. A few tiny tortuous vessels in the 10:00 o'clock meridian behind the ora serrata were the only abnormalities in the right eye. The left eye had small stellate figures of exudate throughout the entire fundus, extending in some meridians almost to the point midway between the equator and the ora serrata. The distribution of the exudate was not related to vessel pattern but simply scattered at random. Between the 2:00 and 6:00 o'clock meridians, abnormal vessels presented as a broad band astride the equator and extending anterior to it. The abnormal formations comprised sausage-shaped, dilated segments with arteriovenous shunts, miliary and saccular aneurysms, in addition to networks of small vessels of irregular caliber. In the 4:30 meridian, about midway between the equator and the fovea, a single large, saccular aneurysm surrounded by a ring of exudate was located on the inferior temporal vein. An extensive star-shaped mound of exudate (Type I) occupied the central macular region (Figure 6). Between the 1:30 and 5:30 o'clock meridians, the retina anterior to the equator was elevated and edematous. A singular observation was made concerning the entire venous system. All of the retinal veins were markedly dilated and darker than normal. This generalized venous dilatation with widespread exudate formation was interpreted as a form of "steal syndrome."

On January 30, 1974, the telangiectasia was treated with the xenon arc machine. This was repeated on April 17, 1974. The vision had now improved to 20/200 and all exudate was absorbing rapidly. A nubbin of fibrous tissue was present in the central macular region (Figure 6C). On June 26, 1974, all remaining areas of abnormal vasculature were treated. By September 16, 1974, the central mass in the macula showed marked shrinkage. The exudate nasal to the disc was disappearing at a significantly slower rate than that in the rest of the fundus. The corrected vision was 20/100. Examination of the left eye on November 4, 1974, showed a large macular hole (Figure 6E). The nubbin of glial tissue had been avulsed by a posterior vitreous detachment and was floating freely in the vitreous. A girdle of black pigment was present on this fibrous body (Figure 6F). On February 5, 1975, the exudate in the left eye had all but disappeared. The macular hole was unchanged and the corrected vision was 20/100. Examination of the right eye showed an area of unmistakable telangiectasia in the temporal periphery. This was confirmed by fluorescein angiography (Figure 13).

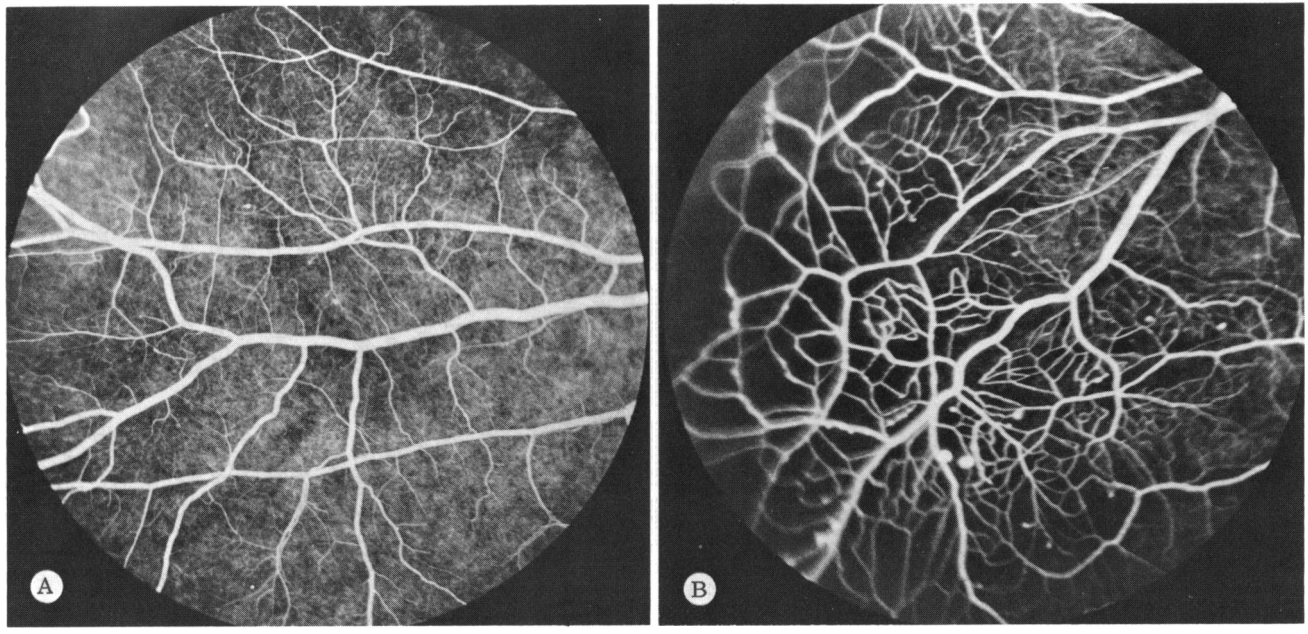



FIGURE 13
Case 30, O.D. A: Showing earliest peripheral telangiectasia in present series in inferior temporal quadrant. B: More advanced changes in superior temporal.

CASE 31

A 48-year-old man was first examined on December 12, 1973. The corrected vision was right eye 20/40 and left eye 20/20. The right eye had an asteroid hyalitis of such an extreme degree that a special effort was required to visualize fundus detail. An area of telangiectasia was present in the temporal half of the fundus at the level of and anterior to the equator between the 8:00 and 11:15 o'clock meridians. Preretinal fibrosis and a lamellar hole were present in the central macular region. The left eye was normal. Treatment with the xenon arc machine was performed on two occasions. On February 24, 1975, the retina was completely flat and free from all abnormal vasculature. The macular region was unchanged, as was the asteroid hyalitis. The corrected vision remained at 20/40.

CASE 32

A 58-year-old man, referred with a diagnosis of Coats' disease left eye, was first examined on April 15, 1974. The corrected vision in the right eye was 20/20 and that in the left eye light perception. Examination of the right eye was normal. In the left eye, posterior synechiae and moderately advanced lens changes were present. The retina was totally detached. Many vitreoretinal adhesions were present. Telangiectatic vessels, interspersed with hemorrhages, were scattered over the detached retina. In the upper temporal quadrant, posterior to the equator, a round, globular mass appeared to arise from the choroid. A solid retinal detachment was noted between the 10:00 and 12:00 o'clock meridians. A similar elevation of the retina in the inferior half of the eye stretched from the ora serrata to well posterior to the equator. These areas of solid elevation of the retina were interpreted as subretinal mounds. The diagnosis of Coats' disease was confirmed.

CASE 33

A 6-year-old boy was first seen on July 2, 1974, with the diagnosis of possible intraocular tumor in the right eye. No light perception was present in the right eye which was quite soft, with band keratopathy and marked rubeosis iridis. The retina in the right eye was totally detached. Two solid tumor masses were present in the posterior polar region. Clumps of exudate were scattered throughout the fundus. A diagnosis of Coats' disease was made and the parents were told that in the not too distant future the eye would in all probability become irritable and require enucleation. A second opinion was obtained and enucleation of the eye advised because of the danger of the blind eye harboring a tumor. The pathologic diagnosis was Coats' disease.

CASE 34

A 25-year-old man was first examined on July 3, 1974. The corrected vision in the right eye was 20/20 and that in the left eye 20/60. Examination of the right eye was normal. Examination of the left eye showed a normal anterior segment. The vitreous contained many large floaters. Retinal edema in the

peripapillary area and radial striae in the macula were noted. In the temporal periphery, between the 1:30 and 3:00 o'clock positions was a round, elevated, greyish mound, extending well posterior to the equator. The superior and inferior borders were flanked by a narrow rim of dark yellow exudate, Type II. Scrutiny with the slit lamp and contact lens showed the outlines of vessels beneath the surface of the mound. On its surface were tiny scattered hemorrhages, without any abnormal vessels. Fluorescein angiography failed to show any vessels on the mound but circumscribed areas of leakage were demonstrated. It was thought that the lesion represented a form of subretinal mound in which the involved vessels were deep in the retina and from which very little bleeding had yet occurred. When seen on May 5, 1975, the corrected vision in the left eye was 20/20. Small, isolated telangiectatic vessels were now present on the surface of the temporal mass, which had become smaller. Traction on the mass by numerous vitreous strands was now evident. There was a definite increase in the exudate along the superior border of the mound. The peripapillary edema and macular striae had disappeared.

CASE 35

A 50-year-old man, referred with the diagnosis of vitreous hemorrhage, possibly retinal tear left eye, was first examined on September 16, 1974. The corrected vision in the right eye was 20/25 and that in the left eye 20/50. Fundus examination of the left eye revealed extensive hemorrhage in the vitreous. The retina was in place throughout. A mass of telangiectatic vessels was present on the upper half of the disc with an area of superficial telangiectasia at about the level of the equator between the 1:30 and 4:00 o'clock meridians. The right eye was normal. Fluorescein studies confirmed the presence of the abnormal vasculature in the left eye.

CASE 36

An 11-year-old boy, first examined on November 11, 1974, as part of a family study, had no ocular complaints. The uncorrected vision, right eye, was 20/20 and that in the left eye also 20/20. In the right eye the only abnormality was confined to the retinal vasculature in the inferior temporal quadrant. In this area, anterior to the equator, the vessel network was dilated with arteriovenous shunts. There was no sheathing of the vessels, no distinct irregularity of caliber, and no retinal edema. This change was considered to be among the earliest manifestations of retinal telangiectasia in this series. Examination of the left eye revealed abnormal vasculature in the temporal periphery between the 11:45 and 5:00 o'clock meridians (Figure 12D). From the 1:00 to 4:00 o'clock meridians, many large, dilated, sausage-like vessels with arteriovenous shunts were seen. A few bulbous aneurysms were noted. Scattered patches of exudate (Type I) were interspersed with the abnormal vessels. Retinal edema was noted in the immediate vicinity of the large vessels.

CASE 37

A 50-year-old woman was first examined on November 18, 1974. She was referred with a diagnosis of Coats' disease left eye. The corrected vision in the right eye was 20/30 and that in the left eye 20/300. Extensive posterior synechiae were present in the left eye. A large angioma was noted in the upper nasal quadrant with smaller angiomas astride the 5:00 and 7:00 o'clock meridians. An area of superficial telangiectasia with miliary aneurysms was present astride the equator between the 2:00 and 3:00 o'clock meridians. A glomerulous-like tuft of vessels was seen at the equator in the 4:00 o'clock meridian. The retina was detached between the 8:00 and 12:00 o'clock meridians. A left nephrectomy confirmed the presence of carcinoma. Over the next three months four separate angiomas developed in the right eye. The primary diagnosis was von Hippel-Lindau disease.

CASE 38

A 59-year-old woman was first examined on December 4, 1974 and the corrected vision in the right eye was 20/50 and left eye 20/400. The left eye had a marked asteroid hyalitis. The retina was elevated from the 8:00 to the 12:00 o'clock meridians, extending posteriorly to the disc. Multiple areas of telangiectasia were present with miliary aneurysms and tiny angiomas between the 9:00 and 11:00 o'clock meridians. The vessel arrangement at the disc was abnormal in that all the large vessels entered the inferior half of the disc while numerous small vessels entered the superonasal quadrant of the disc. These latter vessels drained the telangiectasia and angiomas. An area of localized telangiectasia was present astride the equator between the 2:00 and 3:00 o'clock meridians. Detailed study of the right eye showed the retina to be in place throughout. A few tiny, irregular vessels were present along the course of the superior temporal vessels, 1.5 disc diameters from the disc margin. On December 19, 1974, retinal surgery resulted in replacement of the retina and destruction of most of the abnormal vasculature in the upper nasal quadrant. Subsequently, residual lesions were treated with the argon laser. On March 21, 1974, the retina in the left eye was securely healed and the vision correctable to 20/400. Three months later unequivocal telangiectasia was demonstrated in the right eye.

CASE 39

In the course of a routine examination on December 24, 1974, as a member of a family study, this 11-year-old girl was found to have areas of telangiectasia in the temporal periphery of the right eye. The abnormal vasculature consisted of arteriovenous shunts, miliary aneurysms, sheathed vessels, and accentuation of the capillary network. Slight elevation in the peripheral retina was present in the involved area. Similar changes, though less marked, were noted in the left eye.

CASE 40

On December 24, 1974, a 33-year-old man, the father of the preceding patient (case 39), was found to have arteriovenous shunts, sheathing of vessels,

and accentuation of the capillary network in the upper temporal quadrant of his right eye. The left eye was normal.

CASE 41

A 13-year-old boy was examined as a member of a family under study for Coats' disease on November 13, 1974. The corrected vision in the right eye was 20/20 and that in the left eye 20/100. Cataractous changes were evident in the left eye. Masses of telangiectatic vessels involved the temporal half of the eye. Subretinal mounds in the inferior temporal quadrant extended posterior to the equator. Subretinal hemorrhage was present in the macular area. Examination of the right eye revealed extensive superficial telangiectasia in the temporal half and the superior nasal quadrants.

CASE 42

A 12-year-old boy was first examined on December 24, 1974, as part of a family study. The corrected vision was 20/20 in each eye. Early retinal telangiectasia was present in the left eye (Figure 12B).

CASE 43

A 10-year-old girl was examined on December 24, 1974, as a member of a family under study. The corrected vision was 20/20 in both eyes. Early retinal telangiectasia was present in the temporal periphery of the left eye (Figure 12A).

CASE 44

A 16-year-old boy was first examined on March 6, 1975. The corrected vision in the right eye was 20/20 and that in the left eye 20/100. Detailed fundus study of the left eye revealed extensive telangiectasia in an area four disc diameters in size just temporal to the fovea. The abnormal vasculature involved the arteries and veins. It consisted of markedly dilated, sausage-like configuration, miliary aneurysms and a diffuse network of dilated capillaries with only minimal exudate. No other findings were noted. Fluorescein angiography outlined the lace-like telangiectasia surrounding the abnormal central vasculature (Figure 5B). The right eye was normal.

CASE 45

A 15-year-old girl, sister of a preceding patient (case 7), was seen as part of a family study on March 17, 1975. The corrected vision in the right eye was 20/20 and that in the left eye also 20/20. The right eye had retinal telangiectasia between the 8:00 and 10:30 o'clock positions, slightly anterior to the equator. The left eye had minimal telangiectasia, also in the temporal half of the eye.

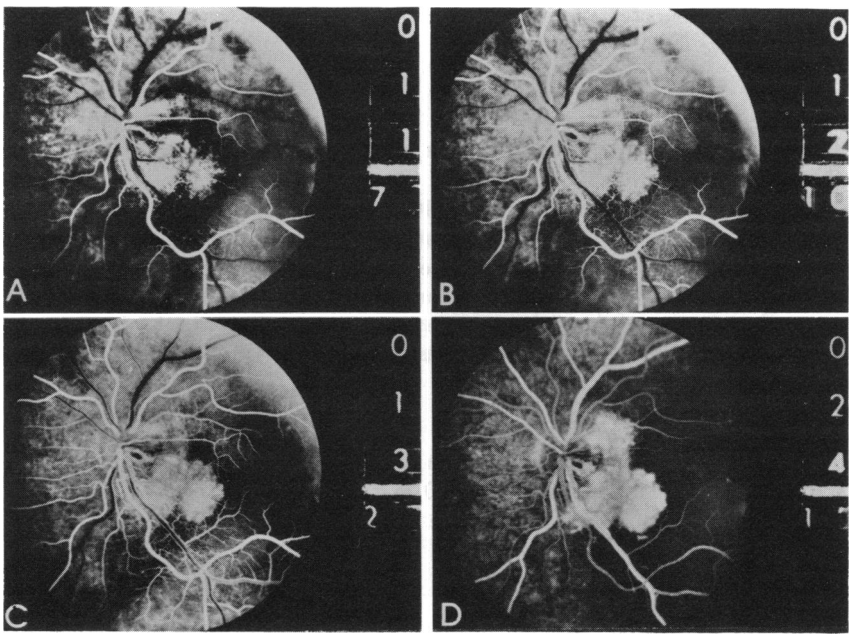



FIGURE 14
Case 47, O.S., showing sequential filling of lobulated vascular mass at temporal margin of disc. Retinal telangiectasia is present below and temporal to the mass (b).

CASE 46

A 52-year-old woman was examined as a member of a family under study for retinal telangiectasia on February 28, 1975. The corrected vision in the right eye was 20/20 and that in the left eye 20/20. The right eye had superficial telangiectasia without secondary retinal change between the 8:00 and 10:30 o'clock meridians. Minimal changes were noted in the temporal periphery of the left eye. This patient is the aunt of a preceding patient (case 30).

CASE 47

A 16-year-old boy was first examined on March 14, 1975. The corrected vision in the right eye was 20/20 and that in the left eye 20/50. Examination five years previously found his left eye to be normal. In the left eye what appeared to be an angioma was present along the temporal margin of the disc. The area between the disc and the fovea was edematous. A circinate figure surrounded the lesion. Fluorescein angiography showed a multilobulated mass along the temporal border of the disc. Telangiectasia of the surrounding capillary network was present (Figure 14). The patient's mother died of spinal hemangioblastoma.

TABLE I: CONGENITAL VASCULAR RETINOPATHY

| | |
|---|---|
| Total number of patients | 47 |
| Bilateral | 10 |
| Male | 28 |
| Female | 19 |
| Youngest | 3½ yr. M. |
| Oldest | 59 yr. F. |
| Enucleated | 2 |
| Treated | 26 |
| Not available for follow-up | 2 |
| Died | 1 |
| Recurrences | 8 |

## ANALYSIS OF CASES IN PRESENT SERIES

The present series consists of 47 cases (Table I). There were 28 males and 19 females. The youngest of the group was a 3½-year-old male and the oldest a 59-year-old female. Bilateral lesions were found at the initial examination in seven patients. Bilateral involvement developed in two patients while under observation. Two eyes were enucleated and the diagnosis of Coats' disease was confirmed in both instances. Twenty-six eyes were treated. Two untreated patients were not available for follow-up. Recurrences were observed in eight patients. One patient, who had been treated, died. The diagnosis was "subarachnoid hemorrhage." The series may be divided into three groups: (Table II).

### *Group I*

In these eight patients, subretinal mounds were present in the fundus at the initial examination or, as in one patient (case #6), the lesion became apparent during treatment. Four patients in this group were treated. Only one patient (case #26) could be considered as having a reasonably successful result.

One patient (case #15), underwent marked regression over six years. The two patients in which enucleation was performed were in this group.

TABLE II

| | |
|---|---|
| Group I: | 1, 6, 15, 18, 25, 26, 32, 44 |
| Group II: | 2, 3, 4, 5, 7, 8, 9, 10, 11, 12, 13, 14, 16, 17, 19, 20, 21, 24, 28, 30, 31, 37, 38 |
| Group III: | 22, 23, 27, 29, 34, 35, 36, 39, 40, 41, 42, 43, 44, 45, 46 |

*Group II*

Subretinal mounds were not present in this group. Of these 23 patients, all of whom were treated, eight showed one or more recurrences. Sixteen of the treated patients showed the same or better vision at the end of the follow-up period. Treatment was abandoned in one patient because the macula would have to have been sacrificed. In two patients, the macular region was treated directly with consequent loss of acuity. One patient (case #24) was an example of both superficial and deep telangiectasia in the same area of the retina. Hemorrhage was not present in the lesion. Angiomas of the von Hippel type were present in six patients in this group.

*Group III*

Two of these 15 patients refused treatment. Treatment was initially planned for one patient (case #23), but observation showed regression of the lesions. Though there was recurrence, there was no loss of visual acuity over a period of four years. Another patient (case #34) was an example of telangiectasia of the deep vessels alone. No hemorrhage had occurred in the lesion. One patient (case #41) demonstrated superficial and deep telangiectasia with identifiable deep retinal hemorrhage and subretinal mounds in the left eye with early superficial telangiectasia in the right eye.

One patient (case #35) had disc involvement with normal vision. One patient (case #18) had extensive subretinal mound formation involving the disc. The fact that no light perception was present in the eye at the initial examination probably indicated optic nerve involvement by the abnormal vasculature.

Three incomplete pedigrees were included in this series (Table III).

## DISCUSSION

In any discussion of this group of disorders, only too frequently the facts were drawn from a small group of cases or even from a single case. Reports of long term follow-up by a single observer were rare. Pronouncements were made upon, and conclusions drawn from the

TABLE III: INCOMPLETE PEDIGREES

1. Cases #3 and #36 are brothers and half-brothers of case #29.
   Cases #3 and #36 are cousins of cases #39, #40, #41, #42, and #43.
2. Case #30 is a niece of case #46.
3. Case #7 is a brother of case #45.

examination of very late cases. Even cases referred to as "very early" were remarkably advanced when compared with the earliest changes described in this presentation. Lack of detail in the evolution and interpretation of the varied ophthalmoscopic picture resulted in biased or inaccurate reading of the pathologic findings. Based solely on the ophthalmoscopic picture in the present series of cases, the following types of telangiectasia were documented:

1. The most common form involved the superficial vessels in the temporal half of the eye. This variety was associated with a characteristic macular lesion consisting of a stellate mass of exudate with a central nubbin of glial or fibrous tissue (Cases #3, #17, #30).
2. Telangiectasia of the superficial vessels in the macula. This usually commenced as miliary aneurysms and progressed to a florid picture of dilated vessels. (Leber's miliary aneurysms with retinal degenation). (Cases #4, #16, #23).
3. The occurrence of types I and II in the same eye (Cases #11, #20).
4. Telangiectasia of both the superficial and deep vessels in the same area of the retina (Case #24).
5. Telangiectasia of the deep retinal vessels only (Case #34).
6. Hemorrhage from deep telangiectatic vessels producing subretinal mounds in association with any of the above types of superficial telangiectasia (Cases #15, #26, #41).
7. Any type of telangiectasia described above in association with retinal angiomas (Cases #9, #14, #19, #21).
8. The advanced picture of multiple subretinal mounds and superficial telangiectasia where the retina was detached and disorganized. Small retinal angiomas, which may have been present, were not visualized. The abnormal vasculature may have involved the optic nerve (Cases #18, #32, #33).

The literature yielded numerous examples of retinal telangiectasia without deep or subretinal hemorrhage producing subretinal mounds. Case III of Coats depicts dilated peripheral vessels, though Figure 14 of this case was incorrectly designated "Angiomatosis Retinae" by Duke-Elder (Figure 15).[53] Similar cases were those of Klein,[54] Reese,[35] and Sugar.[2] This type of case with only extramacular telangiectasia responsed well to treatment. Similar successful treatment results have been reported in the past.[55] Every effort should be made to identify this type of telangiectasia clinically in view of the favorable response to treatment.

A macular star with its central pearl-white dome should suggest superficial telangiectasia. This stellate figure of exudate in the macula has received scant attention in the literature. Coats' description of his third case, both of the ophthalmoscopic and pathologic picture of this lesion, cannot be improved. This lesion occurred in cases #3, #9, #28, and #30. The description of the right eye in case #3 exactly parallels Coats' case III. Over a period of 10 years the lesion absorbed completely (Figure 8). In case #30, resolution took place rapidly until the stage where only the central glial nubbin remained. At this point a vitreous detachment occurred and the nubbin was avulsed from the retina, leaving a lamellar hole in the macula but an intact retina. The glial mass with a girdle of black pigment was seen floating freely in the vitreous (Figure 6F). In case #28, the vitreous was adherent to the central nubbin but had not yet detached. It is important to realize that this lesion is a secondary macular change, developing in association with superficial telangiectasia. It is different from the subretinal mounds which originate in hemorrhage. Previous authors have depicted this central macular lesion but without significant comment.[56,57] Klein's case was clearly an example of superficial telangiectasia with this macular component. She described it as a psammoma-like body.

Subretinal mounds usually presage a treatment failure; however, every effort must be made to establish the correct diagnosis since regression with retention of a cosmetically acceptable eye can occur, as for example cases #15 and #26. Coats considered the subretinal mounds to be hemorrhagic in origin. The pathologic picture of this lesion will vary, depending on the degree of organization of the hemorrhage. If the hemorrhage is localized and seen relatively early, the mound will be confined to a small area, as in case #26. In advanced cases with multiple subretinal mounds and retinal detachment, rubeosis iridis appears. It is worthy of note that rubeosis iridis did not occur in association with subretinal mounds in the absence of retinal detachment (Cases #6, #15, #26). When the diagnosis of far advanced Coats' disease is made in the infant or newborn, we must assume the existence of the lesion prenatally, just as cutaneous angiomas and telangiectasia occur before birth.[58] In such cases it is possible that hemopoiesis, which has been reported in association with the eye, may play a role in the fundus lesion.[59]

Just what particular features of the ophthalmoscopic picture Coats had in mind when he spoke of "massive exudation" remains unclear. In the title of his first paper he used the words, "massive exuda-

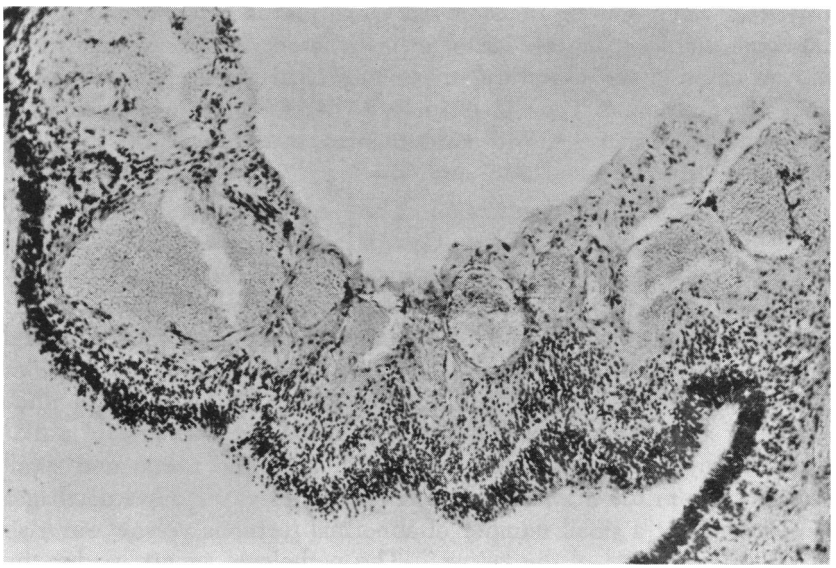


FIGURE 15
Illustration from Coats, showing telangiectasia in temporal periphery of Case III.

tion." In the second sentence of this paper he spoke of "an extensive mass of exudation." By "massive exudation," did he mean those lesions which I have described as "subretinal mounds"? Did the term also include the large sheets of yellow material which I have called Type II exudate together with the true exudative retinal detachment seen in association with this vascular retinopathy? In this writer's view, "massive exudation" was used by Coats to describe one of the hallmarks of the ophthalmoscopic picture, the extensive sheets of exudate present in all his cases, except case III. That Coats was very much aware that the term, "massive exudation" could be misinterpreted was shown by his concern, voiced in his 1911 publication, in which we are told: "This nomenclature was used to avoid possible confusion with any other disease to which the name, "exudative retinitis" may have been applied." In the following sentence he suggested the alternative name, "retinitis haemorrhagica externa." Collins accepted Coats' view that the subretinal mounds were hemorrhagic in origin but he apparently assumed that Coats meant only this particular lesion by massive exudation.[60] Collins also felt that the extensive plaques of subretinal fibrous tissue described in pathologic sections by Coats and others were derived from subretinal hemorrhage. These plaques,

however, have not been accurately correlated with the ophthalmoscopic picture. In this writer's view, based on the present series and on cases in the literature, these subretinal fibrous plaques result from organization of Type II exudate. Therefore, such plaques were found only in association with subretinal mounds. Such a change was apparent in cases #2 and #15, and #26.

In this connection, examination of the two cases of Manschot and deBruijn is revealing.[61] In their Case #1, the right eye of a 4½-year-old boy, four days after cataract surgery, "revealed greenish-grey masses, a local retinal detachment, and some retinal haemorrhages." The pathologic report noted: "the postequatorial segment harboured a thick area of irregular subretinal fibrosis with tumour-like nodules containing cholesterol clefts." In their Figure 2—Case #1, a thick plaque of subretinal fibrosis is clearly seen. Case #2, that of a 3½-year-old boy, "showed large exudates all over the retina and small whitish spots in the macular region of the right eye. No haemorrhages were found but a small number of abnormal tortuous vessels was visible in various parts of the retina." The pathologic report, under the heading, "subretinal fibroblastic proliferation," stated, "The external retinal layers showed minor folds which seemed to be caused by a thin subretinal fibroblastic layer which at many places consisted of only one or two cell layers (Figure 9)." In more detailed discussion, under the heading, "subretinal fibrosis," the very thin nature of the subretinal fibrosis in case #2 was stressed. It was pointed out that possibly part of the fibrosis arose from the detached retinal pigment epithelium. These authors fail to comment upon the marked and obvious layer of subretinal fibrosis which they show in their Case #1. In the opinion of this writer, Case #1 of Manschot and deBruijn, based on the information given, was an example of superficial and deep telangiectasia with subretinal mounds. The organization of exudate Type II resulted in the thick plaque of subretinal fibrosis. Case #2, an example of superficial telangiectasia, was, as these authors pointed out, similar to those of Reese and others, mentioned above. In case #2, one would not expect to find extensive subretinal fibrosis since Type II exudate was not present.

Farkas and coauthors, in a single case diagnosed as Coats' disease, reported on the microchemical analysis of the subretinal fluid.[62] These authors stated, "The results are entirely compatible with the postulate that the cause of Coats' disease is subretinal leakage of blood plasma." No mention was made of the presence of what could be interpreted as subretinal mounds or subretinal fibrous plaques. The case of Far-

kas and coauthors, based on the data given, was one of superficial telangiectasia, in which case the finding of components of whole blood would not be expected. Had subretinal mounds been present with Type II exudate, break-down products of hemoglobin arising from the organizing hemorrhage may have been present in the subretinal fluid. Furthermore, the lack of correlation of the ophthalmoscopic details with the pathologic findings in a glaucomatous eye tends to invalidate the results of this study.

In cases of superficial telangiectasia, it is essential that the diagnosis be made as early as possible. In this regard, the examiner must be alert to note any irregularity of the vessel pattern, especially in the periphery of the fundus in a young person or child. The introduction of fluorescein angiography has provided an excellent tool in diagnosis.[63] Even in eyes which appeared normal on ophthalmoscopic or slit lamp examination, fluorescein angiography showed an abnormal pattern. Repeated fluorescein angiography during a six month period can demonstrate significant advance. Fluorescein angiography is invaluable in following the course of a particular case and as a guide to treatment and follow-up of treated cases. In case #19, the evolution of the macular lesions from a few miliary aneurysms to a picture of diffuse telangiectasia was followed over a five year period (Figure 10). Similarly, in case#3, in which good vision had been maintained for 10 years following treatment, discrete areas of leakage have recently occurred in the macular region in the right eye (Figure 8D). In case #2, a 45-year-old man, telangiectasia recurred 10 years after the commencement of treatment. There had been a gradual improvement in vision over this period of time. Fluorescein angiography demonstrated the development of typical temporal telangiectasia in a 41-year-old man (case #4) more than 10 years after photocoagulation of the macula for Leber's retinal degeneration with miliary aneurysms (Figure 9B). Furthermore, in all families where a diagnosis of Coats' disease has been made, it is strongly recommended that at least the younger members have annual fluorescein studies.

In this series the recurrence rate was surprisingly high, as indicated both by extension of the macular lesions or recurrence in the periphery. In all eyes, the recurrences took the form of a lace-like network of telangiectatic vessels, regardless of the form or location of the initial lesion. In case #12, telangiectasia appeared seven years after the successful treatment of a large angioma. As noted above, telangiectasia and miliary aneurysms occurred in association with this angioma.

Goldberg and Duke recently reported the histopathologic finding of microaneurysms and angioma in the same eye.[64]

The diagnosis of Coats' disease does not invariably lead to intractable glaucoma with loss of the globe. Regression of the lesions does occur. Friedenwald reported such a case in 1914.[65] He provided a detailed color plate. The case was one of superficial, widespread telangiectasia, both central and peripheral. Fifteen years later all vessels had become quite attenuated and all exudate had disappeared.[66] No subretinal mounds were present in Friedenwald's case, which was similar to that depicted in Oehler's atlas, referred to above, and to cases #23 and #44 in the present series. On the other hand, in Coats' seventh case, which had been followed for 16 years, subretinal mounds were present, judging from his description, and the eye eventually became glaucomatous and blind.

When case #15 of this series was initially examined, this writer felt quite sure the course would be rapidly downhill with loss of the globe. Seven years later, the subretinal mounds were receding and the superficial telangiectasia had all but disappeared. The broad sheets of yellow exudate (Type II) had become converted into what appeared to be whitish fibrous tissue. Vessels at the disc were markedly attenuated and displaced. Friedenwald had commented on this vessel displacement and demonstrated it in his case. Over a period of two years, case #26 showed marked regression of a subretinal mound following treatment. The large sheet of Type II exudate was replaced by a sheet of whitish fibrous tissue. The rapidity with which superficial telangiectasia can advance was shown in Snell's case in which a 13-year-old girl was found to have normal vision and a normal fundus at the initial examination. Three years later the vision had fallen to 10/200 and a picture of widespread, massive telangiectasia was present in the central and peripheral areas.[67] Snell reported the case as "angiomatosis retinae."

As noted by Reese, the disc may be the primary site of improvement. Case #35 showed both disc and peripheral lesions. No light perception was recorded in cases #18 and #33. In case #15, no light perception was found six years after the vision had been recorded as hand movement. Could these cases then represent involvement of the retrobulbar portion of the optic nerve? In the present historical review three, or at most four, cases of vascular lesions of the optic nerve were reported in the nineteenth century. In the present century, only three cases of such involvement were recorded up to 1974.[68] In the case of Archer and Krill, optic atrophy was present in the right eye and peripheral retinal telangiectasia in the

left eye with a normal disc. The patient's grandfather had a tumor of the optic nerve.[69] The presence of telangiectasia in the optic nerve in this particular case was a real possibility, in my opinion. In a recent case, a vascular lesion was found in the intracranial portion of the optic nerve 25 years after the appearance of the atrophy.[68]

If telangiectasia of the retina is viewed as a pathologic entity and a significant locus in a spectrum of retinal anomalies in which other components of the spectrum have been demonstrated in the brain, it is reasonable to assume, on developmental grounds, that retinal telangiectasia has a cerebral counterpart. Streeter showed that aneurysms resulted from persistence of the primitive vascular network prior to differentiation of the vessels into arteries and veins.[70] Foster Moore alluded to the possible association of retinal and cerebral miliary aneurysms. He was apparently unaware of Liouville's report.[71] Cushing and Bailey defined telangiectasia of the brain and discussed its relationship to other cerebral vascular anomalies.[72] A most recent classification of brain tumors added nothing new.[73] In the text by Blackwood, Dodds, and Sommerville (Figures 308 and 309), the similarity of capillary telangiectasia of the brain to the retinal lesion is striking.[74] Hoyt considered the cerebral and retinal lesions to be identical histologically.[75] It must be remembered that the von Hippel angioma in the retina had been described well over a quarter of a century before Lindau reported a similar lesion in the brain.[76]

Arteriovenous malformation or racemose aneurysm of the retina and optic nerve is part of the syndrome of Bonnet, Dechaume, and Blanc.[77] In this syndrome, according to Wyburn-Mason, an intracranial, vascular malformation is always associated with the retinal vascular malformation.[78] More than half a century elapsed from the first description of this lesion in the eye by Magnus[79] until Krug and Samuels reported it in the eye and brain.[80] Telangiectasia in the brain is usually silent.[81] In reported cases of this vascular anomaly, the diagnosis has been made at autopsy, death being almost invariably preceded by a vascular accident. The actual telangiectasia may be obscured by overlying or associated hemorrhage.[82] McCormack and Hardman's finding that the posterior fossa, the location of the Lindau lesion, is the commonest site of telangiectasia is of great significance.[83] Bryson and Wolter have reported a case in which miliary aneurysms of the retina were associated with central nervous system dysfunction.[84] An unusual case of symptomatic capillary telangiecatasia of the brainstem without hemorrhage has been recorded.[85] One patient (case #16) in the present series died from "subarachnoid hemorrhage". This

writer thinks it is only a matter of time until it will be possible to demonstrate intracranial telangiectasia in association with the retinal counterpart in a certain percentage of cases.

Computerized tomography, a fundamentally new technique of radiologic diagnosis developed by Hounsfield[86] and first clinically evaluated by Ambrose,[87] may provide a method for the detection of small cryptic intracranial lesions, including telangiectasia. The value of this new, noninvasive technique has been demonstrated in the field of neuro-ophthalmology.[88] Of course, the lesion in the brain, as has been repeatedly shown at autopsy, may be extremely small and obscured or lost in associated hemorrhage, just as lesions of the von Hippel type, in certain instances, have been found only in microscopic sections of autopsy specimens.[89,90]

Attempts have been made to relate retinal telangiectasia to various systemic disorders.[30,51] Retinal lesions in hereditary hemorrhagic telangiectasia were first described by François in 1938,[91] followed by the first American report in 1971.[92] In 1863, Virchow described telangiectasia in the cerebral hemispheres and pons with the lesion in the liver.[93] Visceral involvement is now recognized in hereditary hemorrhagic telangiectasia.[94,95] Eventually, retinal telangiectasia may be found to be a component of the systemic angiomatosis of Ullman.[96]

In the introduction to this essay, this writer expressed the view that these disorders form a spectrum. An examination of the cases presented supports this hypothesis. Cases #5, #10, #17, and #23 represented instances of localized, early, superficial telangiectasia. Both central and peripheral telangiectasia were exemplified in cases #2, #4, #11, #14, #19, #20, #26, and #41. Case #24 is an illustration of superficial and deep telangiectasia in the same area of the retina. Following treatment, a vein was seen entering the depths of the lesion. This picture was similar, if not identical, to that following treatment of case #26. Case #34, like #24, was also an intermediate example of telangiectasia. Only the deep retinal vessels were involved and hemorrhage had not occurred in the area of the abnormal vasculature. On the other hand, in case #41, superficial telangiectasia with subretinal mounds and recognizable subretinal hemorrhage was present in the left eye with early peripheral telangiectasia in the right eye. These cases, thus far, formed a continuous spectrum of telangiectasia. Angiomas with telangiectasia were present in cases #9, #13, #14, #19, #21, #37, and #38. In case #14, an angioma developed after telangiectasia had been present in the eye for at least nine years. Cases #14 and #38 showed an anomalous distribution of the retinal vessels. In

case #38, the peripheral telangiectasia exhibited a pattern different from all the other cases. The abnormal vasculature terminated in fan-like projections towards the ora serrata in contrast to the usual lace-like shunts of peripheral telangiectasia. Small angiomas were associated with the telangiectasia. This fan-like arrangement of the vessels may indicate the potential for the vessels in question to develop angiomas. Egerer and coauthors depicted a similar case in their Figure 4.[48] In case #47, the peripapillary lesion at first was considered to be an angioma. Fluorescein angiography, however, showed it to be composed of clusters of telangiectatic masses, possibly early angiomas. Diffuse telangiectasia was present in the retina in the vicinity of the lesion (Figure 14). The lesion may well be similar to that in case #38 but in a different location. In case #12, telangiectasia reappeared seven years after successful treatment of a large angioma. In case #37, multiple angiomata were treated in the left eye, as were areas of telangiectasia and glomerulous-like formation of vessels. A left nephrectomy confirmed the diagnosis of carcinoma. While under treatment, the earliest lesions of the von Hippel angioma developed in the right eye. In the earliest stages, multiple fine vessels formed a shunt-like communication between an arteriole and a vein. The new vessels, however, unlike those of telangiectasia, displayed marked leakage from the earliest stages. Finally, case #22 was that of a classical racemose aneurysm in the left eye with early peripheral telangiectasia temporally in the right eye. These numbered cases ranged in age from 3½ to 59 years. They exhibited the entire range of the anomalies of congenital vascular retinopathy. Retinal telangiectasia, the basic lesion in Coats' disease, was the common element. Atypical cases of angiomatosis and telangiectasia representing "intermediate forms" of these anomalies have been reported by Witmer and coauthors[97] and by Gronwall.[98]

Retinal telangiectasia, like retinal angiomas, arises from apparently ophthalmoscopically normal vasculature. In case #37, angiomas appeared in the right eye while the left eye was under treatment. Stern first described the earliest stage of a retinal angioma.[99] Ditroi,[100] followed by Gamper[101] and more recently by Welch,[102] described the early von Hippel lesion. The retinal angioma was thought to develop from congenital rests. An entire vascular unit, arteriole, capillary bed and venule were involved.[103] Jesberg and coauthors have suggested that the angioma may not arise from such rests but may develop *de novo* at any age in an individual with the proper genotype.[104] While the angioma is a circumscribed lesion in which new channels are formed, telangiectasia is a diffuse process which consists of dilatation of the pre-existing capillaries.

In 1894, Collins showed the retinal angioma to be familial.[50] The fact that retinal telangiectasia can occur in a familial form, as seen in the present series, brings it closer to other anomalies, such as arteriovenous shunts and angiomas.

The basic lesion in telangiectasia may be an alteration in the control of blood flow within the capillaries, resulting in vessel dilatation. In the peripheral retina in the human, the function of shunting vessels is a matter of controversy.[105] In the cat and dog, shunt vessels are a prominent feature. Thuranszky's studies on the cat, though open to technical objections, support the view that a shunt-like mechanism plays a significant role in the retinal circulation.[106] In the present series, a lace-like pattern of large caliber shunting vessels was demonstrated in all areas of the retina. Certainly, in the earliest stages of telangiectasia—judging from the ophthalmoscopic and fluorescein angiographic studies, altered intraluminal pressure may be a factor. Following dilatation of the vessel, secondary changes may stem from stagnation and local anoxia. Changes in the vessel wall with altered permeability ensues.

Changing concepts of the development of the retinal vessels may shed light on the development of these anomalies. Since the closing years of the last century, the retinal vessels were thought to develop from the hyaloid system by a process of budding. Michaelson, updating this concept, conceived the retinal capillaries as arising from the veins. The growth of the capillaries was inhibited in the neighborhood of the arterioles, leaving a capillary-free zone.[107] More recently, Ashton has shown that retinal arterioles and veins arise from the capillaries and not the reverse, as was previously believed.[108,109]

This writer has little doubt that had Coats lived out a normal life span, he would have shed light on the etiology of the condition since known by his name. Indeed, Coats may well have begun the search for an animal model for his "massive exudation" for in 1911 he published a report on tubercular choroiditis in the cat. In the detailed color plate of the cat's fundus, the similarity of this condition to Coat's disease is evident.[110,111]

Because of the widening interest in animal ophthalmoscopy, this writer believes it is only a matter of time until retinal telangiectasia is reported in a subhuman species. A racemose aneurysm was recently described in a primate,[112] and a new form of macular and generalized retinal degeneration was reported in the baboon.[113] In a new text on veterinary ophthalmoscopy, peripapillary anomalous vessels were depicted in the cat and dog.[114]

Sixty-seven years after its description by George Coats, the disorder remains an enigma. His clinical and pathological descriptions remain unequalled. Little, if anything, has been added to Coat's possible etiologic factors. In the introduction to this essay, quotations were taken from Coats' original article and I conclude in like manner.

In his second and, I should think, least read publication, concerning etiologic factors, Coats stated, "It seems certain therefore that ophthalmoscopically recognizable vascular changes are not a necessary precursor of the disease. It may be that changes are indeed present but they are too subtle to be detected with the ophthalmoscope or even with the microscope."

Whatever these changes may be, they certainly constitute "a very peculiar form of vascular disease" (Coats, 1908, p. 440), which in conjunction with "an extensive mass of exudation" (Coats, 1908, p. 440), comprise an entity in which "all these things are in the highest degree characteristic of this disease and form a picture not to be mistaken for any other." (Coats, 1911, p. 296).

## SUMMARY AND CONCLUSIONS

1. The literature on Coats' disease was reviewed.
2. The ophthalmoscopic picture of Coats' disease was described.
3. Coats' original cases were analyzed. His comprehensive discussion on all aspects of his cases was explored.
4. A series of 47 cases, followed and treated by this writer, was presented. Certain features in common with this group and with Coats' original cases and other cases in the literature were examined and discussed.
5. In agreement with previous reports, the basic lesion in Coats' disease was considered to be a congenital anomaly of the vasculature of the retina and optic nerve, manifest ophthalmoscopically as telangiectasia.
6. The telangiectasia may involve the superficial or deep retinal vessels, separately or together. Hemorrhage occurs from the deep telangiectasia, producing subretinal mounds.
7. Details of the ophthalmoscopic picture were correlated with the reported pathologic findings.
8. Long-term follow-up shows that many cases, even those quite advanced, may regress spontaneously.
9. Treatment of cases with subretinal mounds is likely to result in failure.

10. Retinal telangiectasia is frequently bilateral.

11. There was a high incidence of recurrence following treatment. Treated cases should be followed indefinitely.

12. The possible relationship of retinal telangiectasia to similar lesions elsewhere in the body was discussed. The probability of an intracranial counterpart of the retinal lesion was pointed out.

13. For the first time, the retinal telangiectasia was shown to be a familial disorder. Three incomplete pedigrees were presented.

14. The writer believes that Coats' disease and von Hippel's disease together with retinal arteriovenous malformations, including racemose aneurysm, form a broad spectrum of retinal vascular anomalies. An effort was made to substantiate this hypothesis from the cases presented. It was suggested that this group of disorders be referred to as congenital vascular retinopathy.

15. Since the time of Coats, little of real significance has been added to our knowledge of the etiology of the condition bearing his name.

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