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Coats's disease: clinical, angiographic, histopathological findings and clinical management

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SUMMARY Twenty-four consecutive patients with Coats's disease are reported. In 9 cases the blind eye was enucleated because of total exudative detachment of the retina or because of untreatable secondary glaucoma. No treatment had been given. Fifteen patients between 22 months and 52 years of age were managed by argon laser or xenon photocoagulation and/or cryocoagulation. The number of treatment sessions varied from 1 to 9 per patient and totalled 49 sessions. Fluorescein angiography was performed in all cases. In most eyes the arterial system seemed to be more damaged than the venous side. Many arterioles ended in macroaneurysm-like dilatations surrounded by avascular areas or complete vascular closure in more advanced cases. In 11 out of 14 eyes the vision improved after treatment or remained unchanged. The follow-up varied from 1 to 8 years. Persistent and aggressive long-term treatment of Coats's disease is recommended because the prognosis without treatment is poor.

There is considerable confusion in the literature regarding retinal disease with abnormal vasculature and exudation. Coats[1] originally described 3 types of retinal disease with massive exudation: type 1, without vascular change; type 2, with vascular changes; and type 3, with arteriovenous communication. Later type 3 has been regarded as a retinal angioma. Three of Coats's cases of type 2 were later considered to be a separate entity by Leber,[2] who described similar cases characterised by multiple miliary aneurysms especially in the smaller arteries of the retina. This form has been named 'Leber's multiple miliary aneurysms with retinal degeneration.'[3] A posterior variety of the localised telangiectasis was recently described by Gass and Oyakawa[4] as 'idiopathic juxtafoveolar retinal telangiectasis.' In this paper we report the clinical and histopathological findings and the management of the various forms and stages of retinal telangiectasis or Coats's disease.

Patients and methods

Twenty-four consecutive patients representing various stages of Coats's disease are reported. In 9 cases (group A) the blind eye was enucleated because of total exudative detachment of the retina or because of untreatable secondary glaucoma. No treatment had been given. In 15 cases (group B) the lesion occupied part of the retina; most of these eyes proved treatable.

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Group A. The age of the patients ranged from 2 to 59 years. All except one were males. The 3 youngest patients (cases 1–3) represented leucocoria and exotropia. Four patients (cases 4, 7, 8, 9) had pain in the eye which had been blind since childhood. Two patients (cases 5 and 6) had noticed gradual deterioration in vision in one eye during the last 6 to 18 months. In these cases the routine eye examination carried out by the school nurse 2 years earlier had not revealed any abnormalities in their vision. One patient suffered from epilepsy, in the other cases the general history was unremarkable. The other eye proved healthy in 7 cases, 1 patient had bilateral Coats's disease, and 1 showed uveal coloboma and myopia of −7·0 D in the other eye.

Group B. In 2 female and 13 male patients the lesion was less advanced. The mean age of the patients was 25·0 years (range 22 months to 52 years). Nine patients complained of deterioration in vision, 1 had pain in the eye, 3 were discovered at routine eye examination, and in 2 cases the parents had noticed 'something abnormal in the look of the child.'

The management of patients in group B consisted of photocoagulation and/or cryotherapy except in one eye which was regarded untreatable. The posterior lesions were treated with argon laser or xenon photocoagulation. In most cases more than one treatment was needed to occlude all the leaking

Table 1 *Clinical and histopathological data on patients in group A*

Case no.	*Age/sex*	*Eye*	*Symptoms*	*Visual acuity*	*Intraocular pressure*	*Lens*	*Iris*	*Chamber angle*
1	1 M	L	Leucocoria, exotropia	NPL	—	Clear	Inflammatory infiltrates	Inflammatory infiltrates
2	2 M	L	Leucocoria, exotropia	PL?	—	Clear	No rubeosis	Occluded
3	4 M	R	Exotropia since 1 yr of age	NPL	—	Dense cataract	No rubeosis	Occluded
4	36 M	R	Blind right eye since childhood	PL	55	Dense cataract	Rubeosis	Inflammatory infiltrates
5	13 M	R	Vision decreased for 6 months	PL	8	Dense cataract	No rubeosis	Occluded
6	16 M	L	Vision decreased for 2 yr	NPL	—	Dense cataract	No rubeosis	Occluded
7	48 M	L	Left eye blind since childhood	NPL	7	Dense cataract	Neovascul. + cyclitic membrane	Occluded
8	59 M	R	Poor vision in right eye since childhood; pain	NPL	42	Dense cataract	Rubeosis	Open
9	47 F	R	Poor vision in both eyes since childhood; left eye blind since 17 yr of age	CF 1 m				
		L		NPL		Dense cataract	Dense membrane	Occluded

NPL=no perception of light; PL=perception of light; CF=counting fingers.

vessels. The peripheral lesions were treated with photo- or cryocoagulation or a combination of both depending on the amount of subretinal exudate and detachment of the retina. In small children and in the most peripheral lesions the cryotherapy, under the visual control of indirect ophthalmoscopy, was preferred. The number of treatment sessions varied from 1 to 9 per patient, and the total of 14 patients had altogether 49 treatment sessions. The follow-up periods varied from 1 to 8 years.

Results

The lesion was unilateral in 23 cases out of 24, and the right and left eye were equally often affected (Table 1). In group A all enucleated eyes were blind without

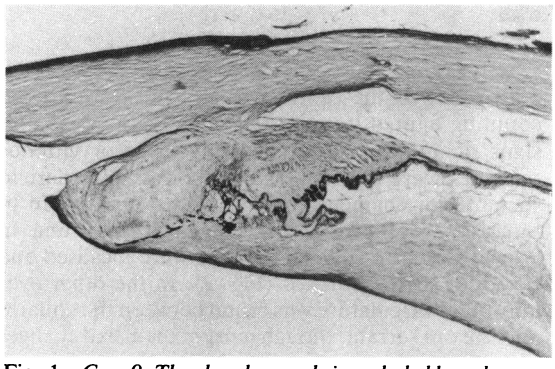

Fig. 1 *Case 9. The chamber angle is occluded by a dense cyclitic membrane growing over the pupillary border. (Haematoxylin and eosin, ×26).*

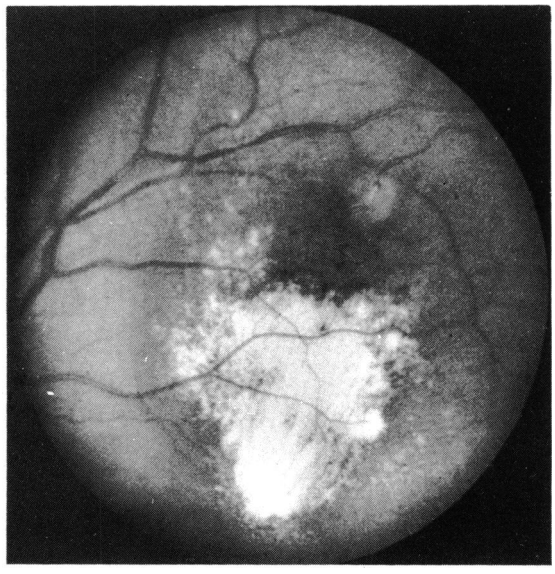

Fig. 2A

Fig. 2 *Case 10, left eye.* (A) *A copy of the colour photograph showing abnormalities in the superior temporal artery and vein with considerable collection of lipid exudates down to the fovea.* (B) *Fluorescein angiogram shows narrowing and localised dilatations in the artery surrounded by coarse capillary network, capillary microaneurysms, and leakage. The corresponding vein shows fewer irregularities. No capillary closure or leakage in the macula.* (C-D) *Same eye 6 years after photocoagulation with argon laser. No exudates or leaking vessels visible, mild pigmentary disturbances in the fovea.*

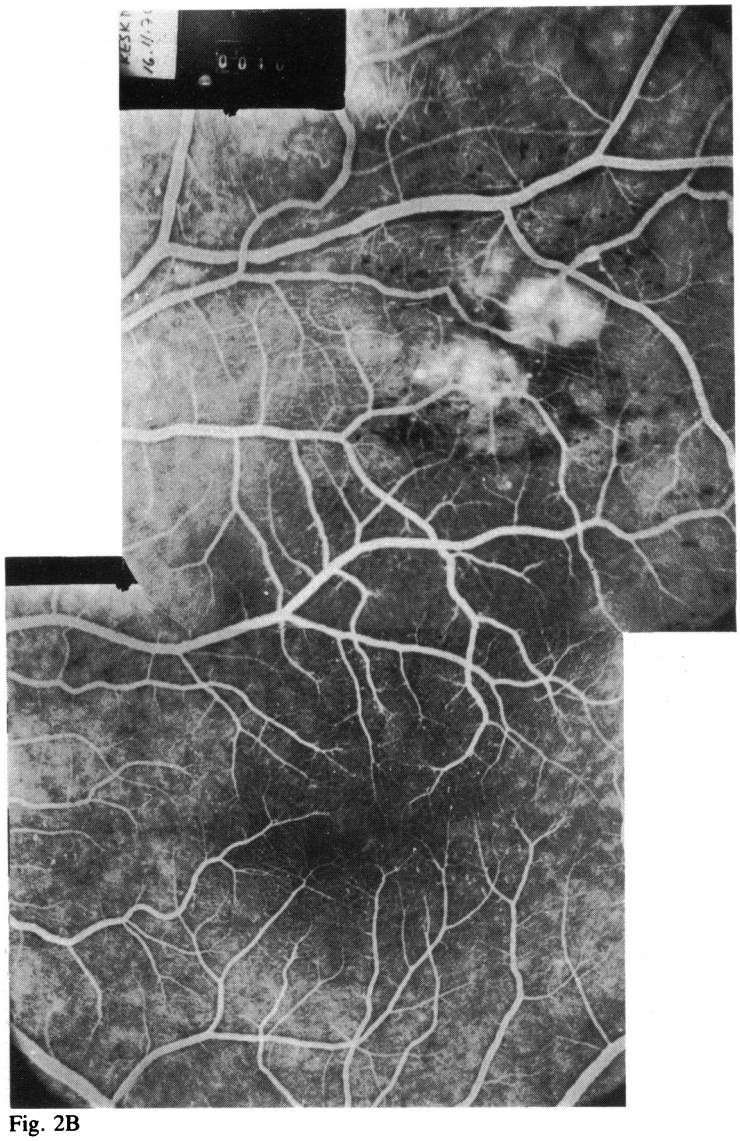

Fig. 2B

projection of light. Two cases (4, 8) showed rubeosis iridis and secondary glaucoma. The others were quiet. The lenses showed a dense cataract in 7 of the 9 cases. The anterior segment of the eyes showed occluded chamber angle (Fig. 1) and 2 eyes inflammatory changes of the iris. The posterior segments of the eyes revealed bullous total retinal detachment with deep and subretinal exudates, massive gliosis and retinal disorganisation, fresh and/or old haemorrhages, ghost cells, and cholesterol chrystals. All showed also retinal vascular changes: dilatation and hyalinisation, while the choroid and the sclera appeared mostly normal.

In group B the central visual acuity varied from counting figures 1 m to 1·0 (Table 2). In 14 eyes no signs of anterior segment involvement or vitreous opacities were found. One eye showed some flare in the anterior chamber and cells in the vitreous. In 14 out of 15 eyes the lesion was in the temporal retina. In 3 eyes (cases 10–12) the changes were localised and posterior to the equator (Fig. 2). In the other eyes abnormal vasculature was found between the equator and the ora serrata, though most of them had changes posterior to the equator as well (Fig. 3).

Ophthalmoscopically the affected retina was greyish and more or less detached (Figs. 4–6).

Table 2 *Clinical data on patients in group B*

Case no.	*Age/Sex*	*Eye*	*Symptoms*	*Localisation and extent of abnormal vasculature*	*Visual acuity pretr./latest*		*Treatment; number of treatment sessions*	*Follow-up (years)*
10	26 M	L	Decreased vision	Superotemporal, MP	0·25	0·8	Argon×6	5
11	43 M	L	No	Superonasal, MP	1·0	1·0	Argon×1	1·5
12	7 M	L	Decreased vision	Temporal, P	0·7	0·5	Xenon×3	8
13	21 F	R	Decreased vision	Temporal, P+MP	0·7	1·0	Argon×3 Cryo×1	2
14	9 M	R	No	Temporal, P+MP	0·8	0·8	Argon×7 Xenon×1 Cryo×1	4
15	2 M	R	Suspicion of squint	Temporal, P+MP	—	1·0	Xenon×1 Cryo×3	2
16	16 M	L	Decreased vision	Inferotemporal, P+C	0·1	0·2	Cryo×1	2
17	51 M	L	Decreased vision	Temporal, P+MP	0·1	C.F.	Cryo×1 encirclement×1	2
18	52 M	R	Decreased vision	Temporal, P+C	0·1	0·1	Argon×2	1
19	48 M	R	Redness, pain	Inferotemporal, P	1·0	1·0	Argon×2	2
20	19 M	R	Decreased vision	Superotemporal, P+C	0·1	—	—	—
21	37 M	L	Decreased vision	Temporal, P+MP	0·7	0·05	Cryo×2	2
22	22 M	L	No	Temporal, P+MP	0·7	0·05	Argon×3 Cryo×2	5
23	12 F	R	Suspicion of squint	Temporal, P	1·0	1·0	Cryo×2	1
24	10 M	R	Decreased vision	Temporal, P+MP	C.F.	C.F.	Argon×3 Cryo×2	1

MP=midperipheral, P=peripheral, C=central. CF=counting fingers. Pretr.=pretreatment.

Irregular dilated vessels as well as intra- and subretinal lipid exudates were seen in all cases. Macular changes were found in 9 eyes. Six eyes (cases 10, 12, 13, 16, 18, 24) showed hard exudates in or close to the fovea (Figs. 2A, 5C), 2 eyes (cases 17 and 20) had cystoid macular oedema (Fig. 6), and in 1 eye (case 21) preretinal fibrosis was discovered.

Fluorescein angiography was performed in all cases in group B. Some amount of capillary drop-out was seen in all cases, and in the more advanced cases large areas of complete vascular closure were seen, particularly in the periphery (Fig. 4B). The rest of the capillary bed in the affected area was coarse and dilated. Large microaneurysms were seen in the arteries and capillaries, less often in the veins. The microaneurysms and other localised vessel dilatations leaked fluorescein, but many of the abnormal looking coarse capillaries did not leak (Figs. 3B, 7C).

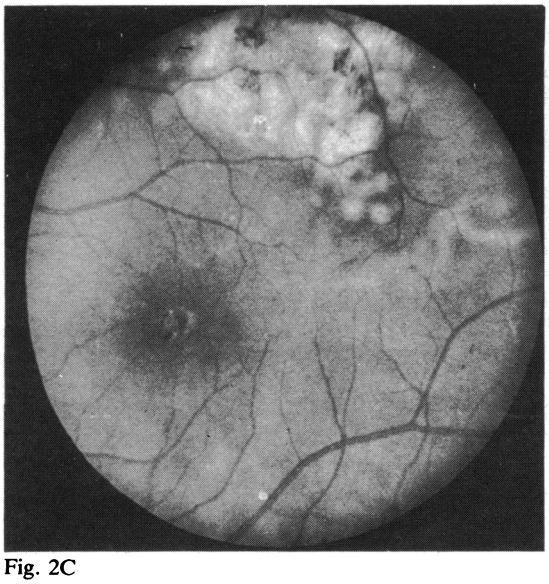

Fig. 2C

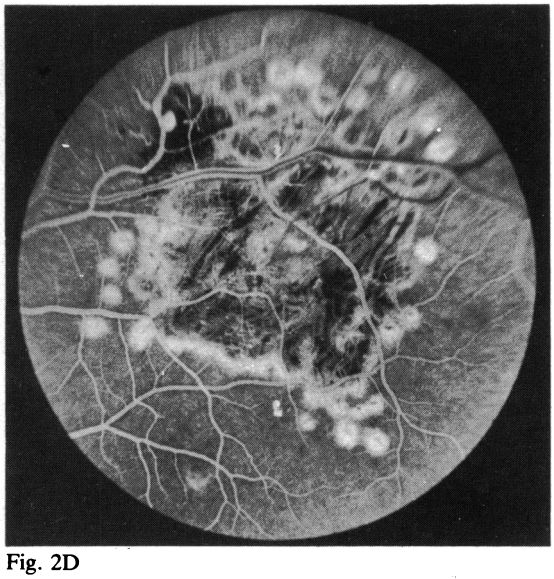

Fig. 2D

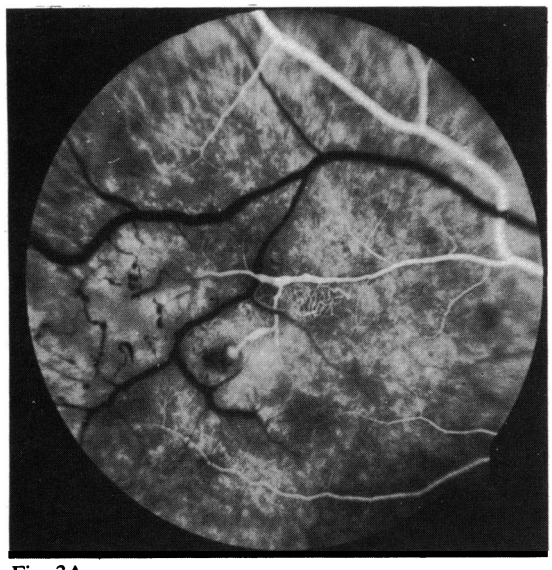

Fig. 3A

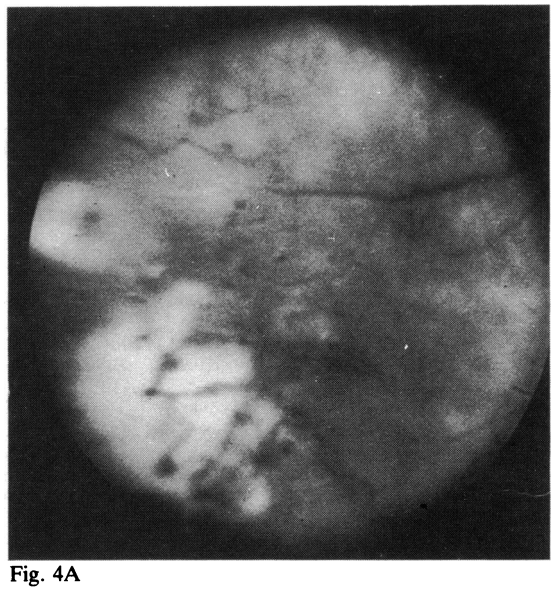

Fig. 4A

Fig. 4 *Case 14, right eye.* (A) *A copy of the colour photograph showing peripheral vascular changes and exudation.* (B) *Fluorescein angiogram of the same area shows extensive capillary closure and large shunt vessels and microaneurysms which leak fluorescein.* (C-D) *Another area of the same fundus before* (C) *and after* (D) *cryocoagulation.*

In most eyes, in the less advanced cases in particular, the arterial side seemed to be more damaged than the venous side (Figs. 2, 3). Many arterioles ended into macroaneurysm-like dilatations surrounded by avascular areas (Fig. 3). Filling of the affected capillary bed was delayed. The corresponding veins were slightly dilated and some of them showed localised dilatations and staining of the wall. In the more advanced cases large shunt vessels from an artery to a vein or from one vein to another were seen (Fig. 4).

Two eyes with perifoveal lipid exudates (cases 12

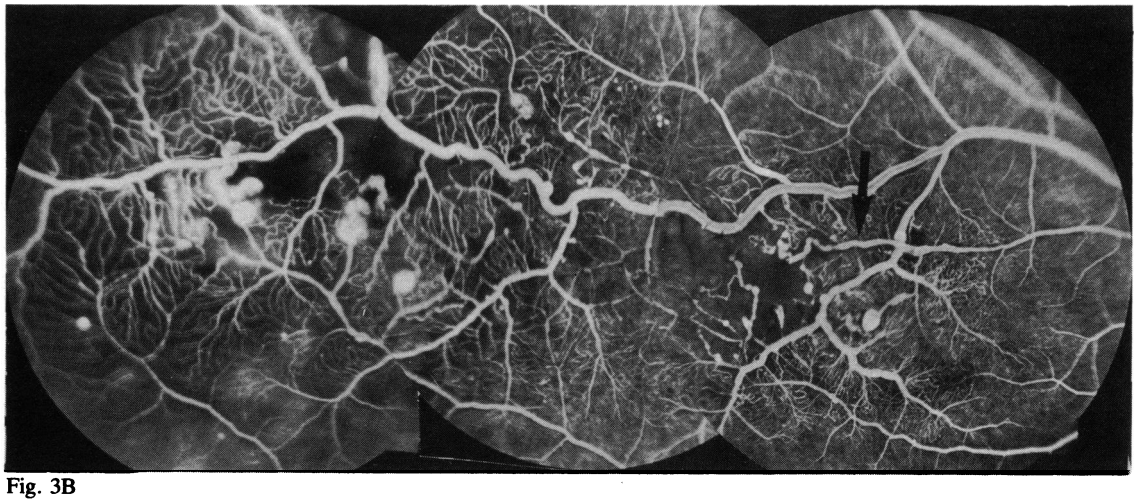

Fig. 3B

Fig. 3 *Case 13, right eye.* (A) *An early phase of the angiogram showing narrowing and slow filling of the affected artery.* (B) *In the venous phase the same artery (arrow) and some other arteries in the periphery end in macroaneurysm-like dilatations which leak fluorescein. The capillary bed is prominent with areas of capillary closure. The corresponding vein shows some engorgement and localised dilatations.*

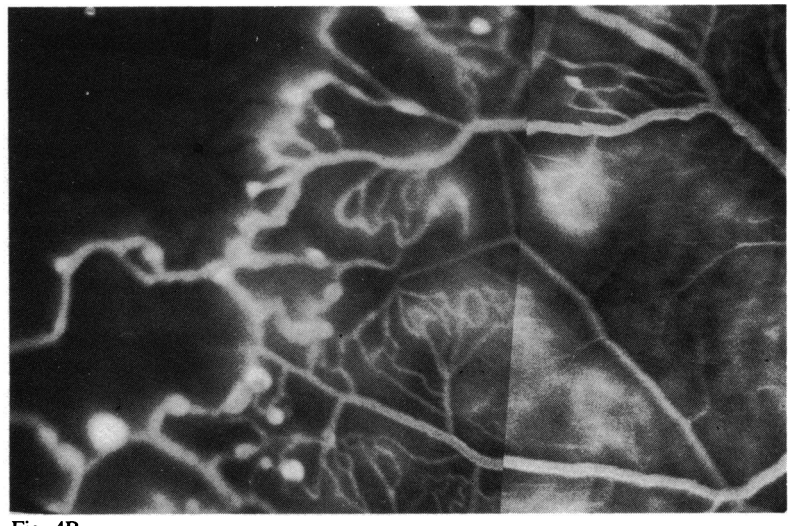

Fig. 4B

and 18) showed capillary closure, abnormal dilated capillaries, and microaneurysms in the macula temporal to fovea (Fig. 7). In the 4 eyes with heavy macular exudates (cases 10, 13, 16, 24) the macular capillaries were normal and did not leak fluorescein (Figs. 2, 5). The 3 eyes with cystoid macular oedema and preretinal fibrosis (cases 17, 20, 21) showed capillary dilatation and leakage in the macula (Fig. 6).

The treatment resulted in an atrophic scar (Figs. 2, 4, 5). Gradually the lipid exudates disappeared also from the macula (Figs. 2, 5). In the more advanced lesions the cryotherapy proved more effective than photocoagulation. After disappearance of the macular exudates some pigmentary disturbance in the fovea remained. In one eye with heavy exudates (case 10, Fig. 2) the vision recovered to 0·8, whereas in the other (case 16) it remained at 0·2 because of the macular scar (Fig. 5). Altogether in 11 out of 14 treated eyes the vision improved or remained unchanged. In 2 eyes the central visual acuity deteriorated owing to persistent cystoid macular oedema, and in one eye owing to posterior subcapsular cataract which developed after the otherwise successful treatment. In one eye retinal detachment progressed after cryotherapy; it was successfully treated by an encirclement procedure.

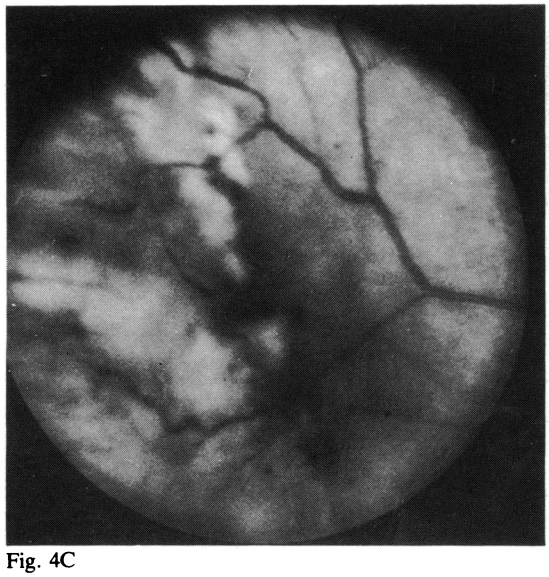

Fig. 4C

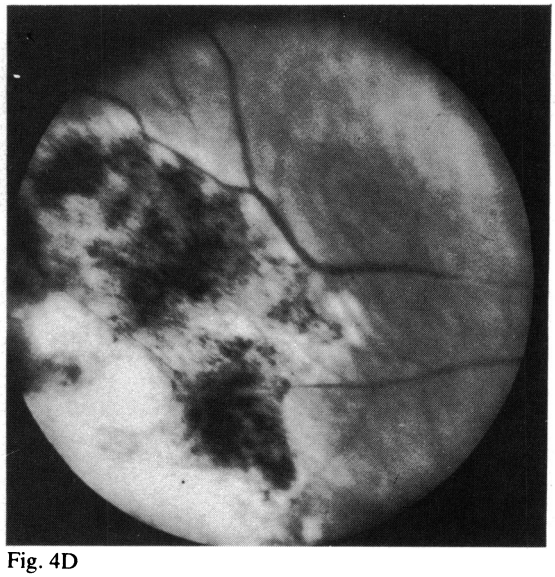

Fig. 4D

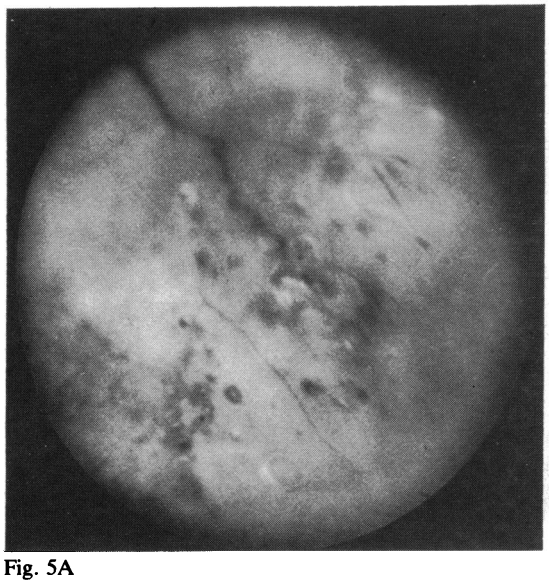

Fig. 5A

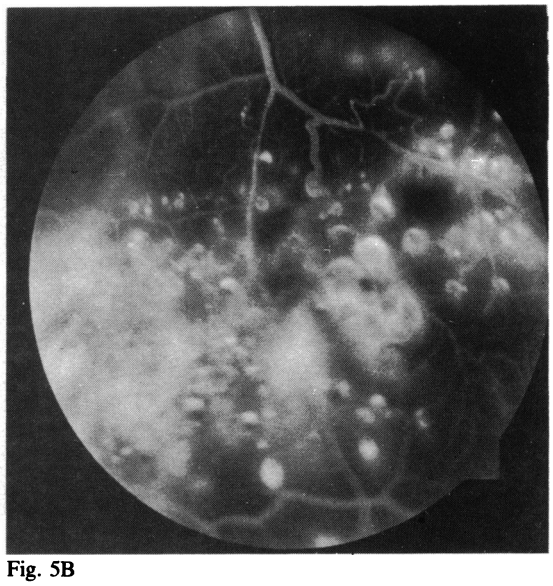

Fig. 5B

Fig. 5 *Case 16, left eye* (A) *A copy of the colour photograph showing abnormal vasculature, some exudates, and detachment of the retina in the lower temporal periphery.* (B) *Same area in the angiogram.* (C) *Considerable collection of exudates at the posterior pole.* (D) *No abnormalities in the macular capillaries visible.* (E-F) *Same fundus 2 years after cryocoagulation showing a residual macular scar* (E) *and atrophy of the treated area* (F).

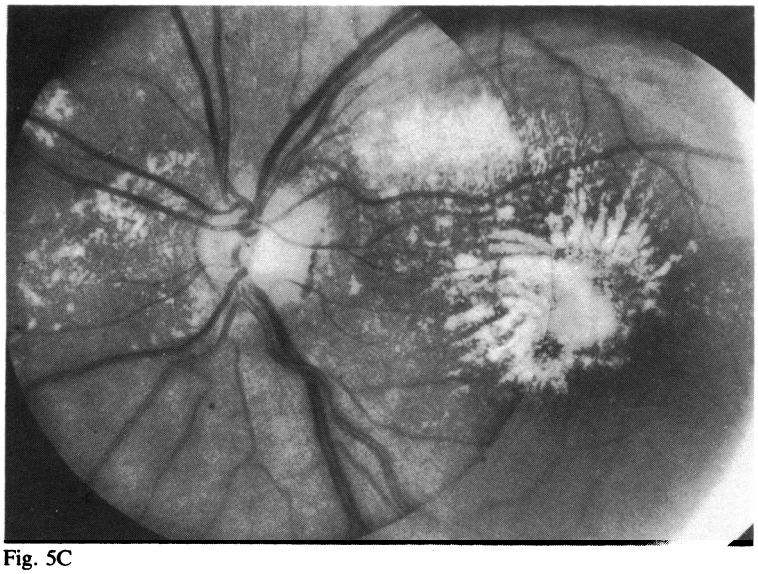

Fig. 5C

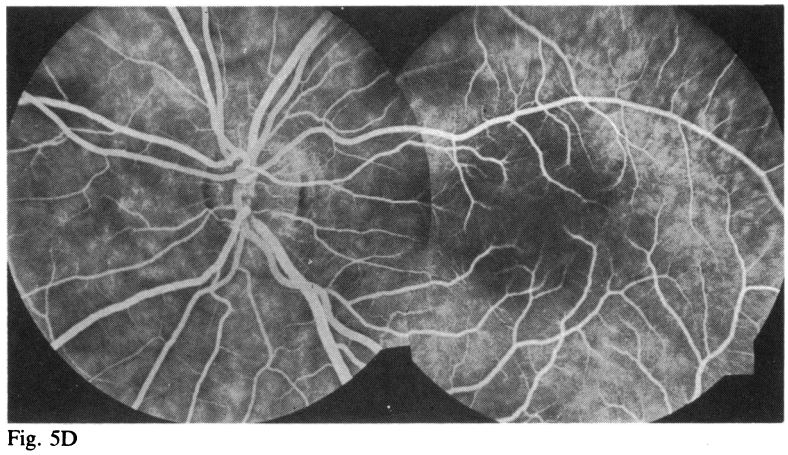

Fig. 5D

Discussion

In the present cases no definite ophthalmoscopic, angiographic, or histopathological differences could be seen between the juvenile and the adult forms of Coats's disease. All stages from localised areas of telangiectatic vessels and exudation to total exudative detachment of the retina were found both in children and in adults. Earlier a hypothesis has been presented according to which the presence of hypercholesterolaemia and an inflammatory process in the uvea might have some role in the pathogenesis of the adult forms of Coats's disease.[5] Our patients had no previous ocular diseases or signs of posterior uveal inflammation. These cases speak in favour of the same basic pathogenesis and the congenital origin of the various forms of Coats's disease,[6] as even the histology does not reveal any differences between the juvenile and adult forms.[7] It has been suggested that the pathological changes may result from a functional or structural break-down of the blood-retina barrier giving rise to mural disorganisation, aneurysmal dilatations, and telangiectasis.[8]

An increase in the area of abnormal vasculature with age was not found, although several patients gave a history of gradual deterioration in vision. In advanced cases the vision was lost due to exudative detachment of the retina. In less advanced cases the main causes of loss of central vision were accumulation of lipid exudates or development of cystoid oedema in the macula. Thus even localised lesions may lead to blindness. In some eyes lipid collections at the macula were due to abnormal leaking capillaries in or close to the macula. In 2 eyes with heavy exudates in the macula, however, no vascular changes were found at the posterior pole. Cystoid oedema and preretinal fibrosis of the macula were related to dilatation and leakage of the macular and paramacular capillaries. In some eyes capillary changes at the macula seemed to be part of the basic

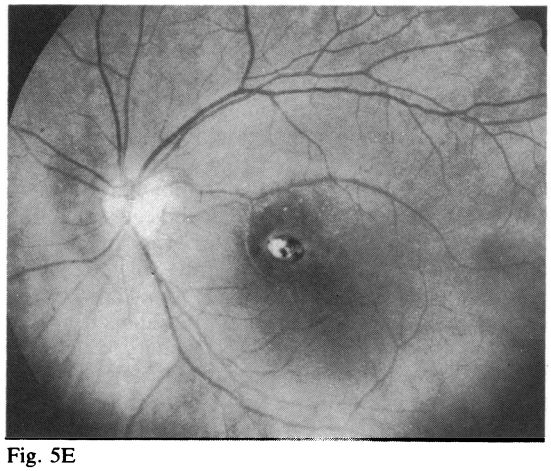

Fig. 5E

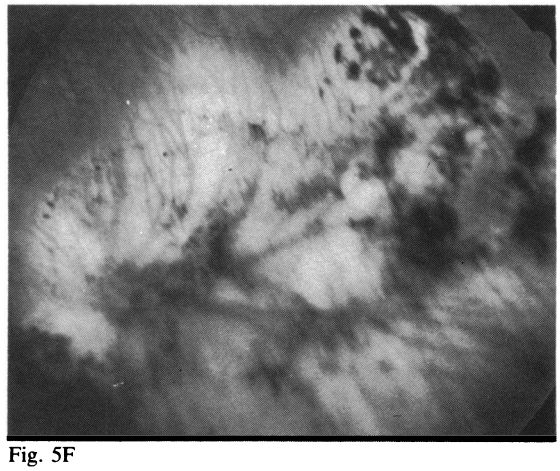

Fig. 5F

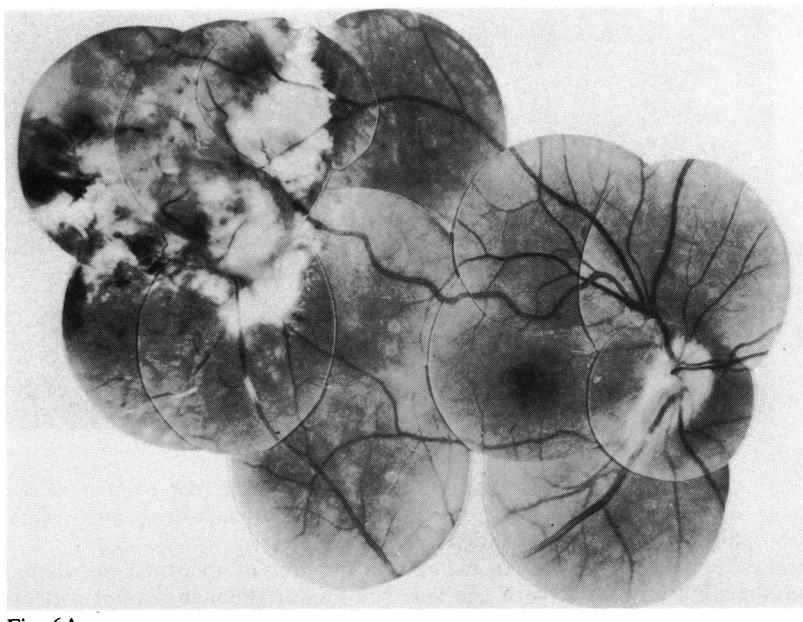

Fig. 6A

Fig. 6 *Case 20, right eye.* (A) *Peripheral Coats's disease with cystoid oedema of the macula.* (B) *Fluorescein angiogram of the macula showing leakage of the macular capillaries and accumulation of the dye in the cystoid spaces.*

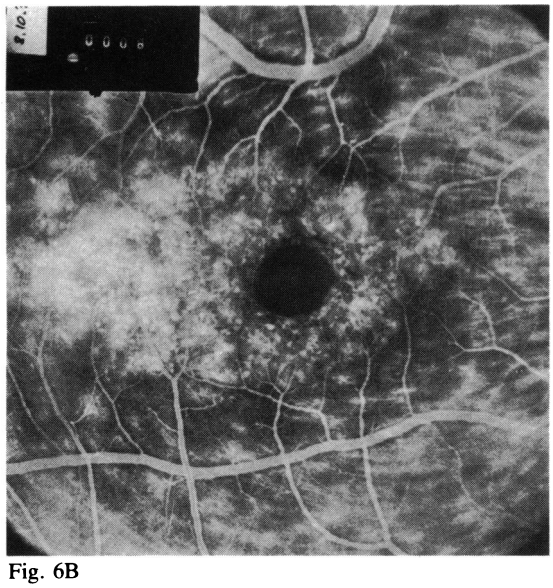

Fig. 6B

vascular abnormality, whereas in the others macular oedema was more probably secondary to peripheral vascular changes.

Fluorescein angiographic studies of the less advanced cases indicated that the arterial side of the vasculature was initially more damaged than the venous side. The affected arteries were attenuated but showed localised dilatations. Some of them filled slowly and terminated in macroaneurysm-like dilatations surrounded by areas of capillary closure. The findings agree with those reported by Spitznas *et al.*[9] but are in contrast with the histopathological findings of Wise and Horava,[10] who found hyalinisation and sclerosis of arteries only in advanced stages of Coats's disease. Reese[11] also stated that 'the lesion seems to be on the venous rather than the arterial side of the vascular tree.' The predominance of arterial changes in many of these eyes was, however, described by Leber,[2] and recently similar observations have been made in fluorescein angiographic studies.[12] The capillary net in the affected area was coarse and the individual capillaries were dilated, but many of them did not leak fluorescein, which indicates that the endothelium in these capillaries was intact. The draining veins were usually slightly dilated, which may be secondary to decreased blood flow in the arterial side of the capillary bed, and shunts from one vessel to another seemed to be secondary to capillary closure. Preretinal or disc neovascularisation was not seen.

Both macular and peripheral exudates absorbed after treatment of the peripheral changes, as has been

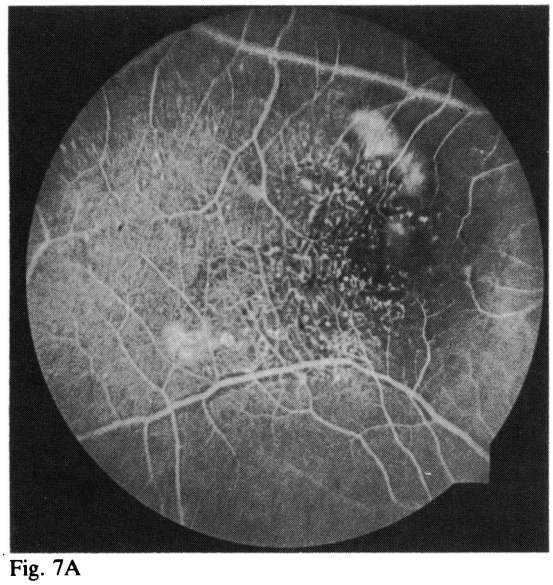

Fig. 7A

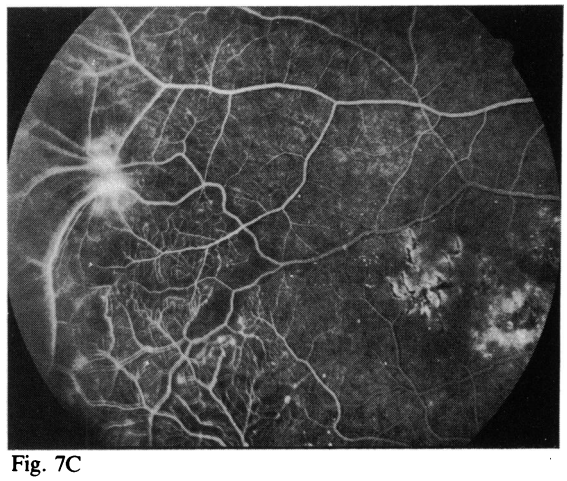

Fig. 7C

Fig. 7 *Case 18, right eye.* (A-B) *Abnormal capillaries and capillary closure in the macula* (A) *with late leakage* (B). (C) *Most of the abnormal capillaries in the periphery do not leak. The latter angiogram was taken after the first treatment of the macular changes with argon laser.*

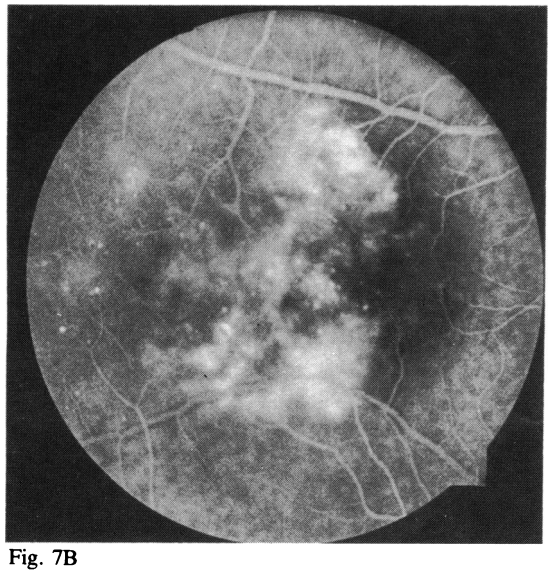

Fig. 7B

described.[13–15] Longstanding exudation in the macula resulted, however, in permanent macular scar and poor central vision. In 79% of the treated eyes the vision either improved or was stabilised at pre-treatment level, which corresponds with previous results.[9 15–17] In most eyes, however, several sessions of treatment were needed.

In the earlier reports photocoagulation has been used more often than cryotherapy. But in this series, especially in the more advanced cases and in the far periphery, cryotherapy proved to be more effective than photocoagulation. We observed no complications in eyes treated with photocoagulation. In one eye extensive cryotherapy resulted in posterior subcapsular cataract, and in another eye exudative detachment of the retina progressed after treatment. This detachment was cured by an encirclement procedure. In spite of these complications treatment of localised forms of Coats's disease with cryo- or photocoagulation is highly recommended, because the prognosis without treatment is almost invariably poor. It has to be emphasised that the number of treatment sessions per patient in our series varied from one to 9. The aggressive attitude towards the treatment of this condition has been previously emphasised.[18–19] Only a few of the reported cases have been shown to have regressed spontaneously.[20] The treatment may not only save the eye but retain useful vision as well.

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