Table 1 The disc damage likelihood scale

New DDLS stage	Narrowest width of rim (rim/disc ratio)			Old DDLS stage
	For small disc <1.50 mm	For average size disc 1.50–2.00 mm	For large disc >2.00 mm	
1	0.5 or more	0.4 or more	0.3 or more	0a
2	0.4 to 0.39	0.3 to 0.29	0.2 to 0.29	0b
3	0.3 to 0.29	0.2 to 0.29	0.1 to 0.19	1
4	0.2 to 0.29	0.1 to 0.19	less than 0.1	2
5	0.1 to 0.19	less than 0.1	0 for less than 45°	3
6	less than 0.1	0 for less than 45°	0 for 46° to 90°	4
7	0 for less than 45°	0 for 46° to 90°	0 for 91° to 180°	5
8	0 for 46° to 90°	0 for 91° to 180°	0 for 181° to 270°	6
9	0 for 91° to 180°	0 for 181° to 270°	0 for more than 270°	7a
10	0 for more than 180°	0 for more than 270°		7b

The DDLS is based on the radial width of the neuroretinal rim measured at its thinnest point. The unit of measurement is the rim/disc ratio—that is, the radial width of the rim compared to the diameter of the disc in the same axis. When there is no rim remaining the rim/disc ratio is 0. The circumferential extent of rim absence (0 rim/disc ratio) is measured in degrees. Caution must be taken to differentiate the actual absence of rim from sloping of the rim as, for example, can occur temporally in some patients with myopia. A sloping rim is not an absent rim. Because rim width is a function of disc size, disc size must be evaluated before attributing a DDLS stage. This is done with a 60D–90D lens with appropriate corrective factors. The Volk 66D lens minimally underestimates the disc size. Corrective factors for other lenses are: Volk 60D×0.88, 78D×1.2, 90D×1.33. Nikon 60D×1.03, 90D×1.63.⁸

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Choroidal neovascularisation

Progressive RPE atrophy around disciform scars

M A Zarbin

Steal syndrome versus aberrant wound healing?

In this issue of the *BJO*, Sarks and co-workers (p 442) report that progressive retinal pigment epithelium (RPE) atrophy develops around the perimeter of disciform scars in patients with age related macular degeneration (AMD). As noted by the authors, the progressive RPE atrophy seems to be caused by the presence of the disciform scar and seems to be distinct from AMD associated geographic atrophy (see, for example, table 2 in their paper). Sarks and co-workers postulate the existence of a “steal” syndrome in which: (1) active choroidal neovascularisation induces remodelling of the adjacent choroidal circulation with reduced blood flow to smaller choroidal vessels; and (2) secondary attenuation and, ultimately, atrophy of the RPE occurs adjacent to disciform scars.

The study was executed carefully. On the basis of fundus photographs alone, it is difficult to judge the presence of RPE atrophy versus RPE depigmentation

with reduced but not completely atrophic overlying photoreceptors.¹ Thus, as the authors recognise, there may be some degree of overestimation of the extent of atrophy in those 18 patients in whom pathological material was not studied.

Is choriocapillaris degeneration and RPE atrophy adjacent to disciform scars caused by the “steal” syndrome postulated by the authors or secondary to other factors?

Is choriocapillaris degeneration and RPE atrophy adjacent to disciform scars caused by the “steal” syndrome postulated by the authors or secondary to other factors? Two additional hypotheses seem worth considering.

Firstly, the processes that incited choroidal neovascularisation might also be responsible for the RPE and choroidal degeneration. Sarks's group and others have noted that choroidal new vessels

(CNVs) often are located adjacent to areas of choriocapillaris degeneration/non-perfusion, which may be a manifestation of RPE dysfunction.^{2–5} Thus, choriocapillaris degeneration and CNV formation may be linked. Grunwald and co-workers' observations on decreased choroidal blood flow in AMD eyes are consistent with this hypothesis.⁶ As Sarks and co-workers suggest, AMD associated RPE degeneration might induce the progressive choroidal changes adjacent to disciform scars through loss of RPE derived trophic factors that help maintain normal choriocapillaris physiology. The authors noted, however, that the development of progressive atrophy did not appear to be influenced by the presence of drusen or pigment changes, which may not be consistent with this explanation.

A second hypothesis is that progressive RPE atrophy in this setting is a manifestation of an abnormal wound healing response. This notion is consistent with two observations mentioned by Sarks and co-workers: (1) RPE atrophy progresses even after the size of the disciform scar has stabilised, and (2) a similar process of progressive RPE atrophy can develop in eyes undergoing laser photocoagulation for diabetic macular oedema without concomitant presence of choroidal neovascularisation.⁷

Although mammalian epithelial cells exhibit stereotyped responses to tissue damage, there is considerable variation, depending on the tissue involved, the

developmental stage of the tissue, and the nature of the injury. (Thus, wound healing responses range from scarless fetal epidermal wound healing to chronic non-healing foot ulcers in diabetic patients.) Epithelial wound healing involves cellular migration across the site of injury as well as fibroblast ingrowth, inflammatory cell recruitment, and changes in the blood vessels of the underlying stroma.⁸⁻⁹ In epidermal wound healing, activated fibroblasts, macrophages, and vascular endothelial cells form granulation tissue that facilitates re-epithelialisation and is remodelled, ultimately, to form a scar.¹⁰⁻¹¹ The molecular signals modulating these responses are being identified.¹² We have observed similar processes (for example, RPE reorientation at the wound margin, RPE detachment from its basement membrane, RPE migration with lamellipodia formation, and RPE proliferation) in organ culture studies of RPE wound healing on aged submacular human Bruch's membrane.¹³ Although RPE cells utilise some of the molecular signals that regulate wound healing in other systems (for example, c-JUN NH₂ terminal kinase),¹⁴ the precise signals involved in the CNV associated wound healing response in AMD eyes are unknown. Maturation of epithelial wounds is characterised by complete re-epithelialisation of the injury site (with cessation of epithelial migration and proliferation), removal of debris, and re-establishment of a new basal lamina and normal permeability barriers.

Sarks's group has shown that in areas of early CNV formation, macrophages and foreign body giant cells are present beneath thinned segments of Bruch's membrane.² In one specimen, a CNV was associated with activated pericytes. Other histological and immunohistochemical studies are also consistent with the notion that AMD and AMD induced CNVs are associated with

chronic injury to the RPE and choriocapillaris.¹⁵⁻¹⁸ Based on results in other wound healing paradigms, one might predict that an RPE and choroidal injury response will not be quiescent until the wound is mature. To the extent that the signals initiating a wound healing response do not result in re-establishment of normal tissue architecture, there may be no downregulation in their expression, and an ongoing process of cell activation may be established.

The authors' suggestion that patients with disciform scars may not be visually stable (at least for a period of time) is plausible. The visual consequences of progressive RPE and choroidal degeneration adjacent to disciform scars remain to be explored in greater detail. In the meantime, the authors recommend that one should take into account ongoing scotoma enlargement when planning management and assessing treatment outcomes in patients with AMD associated CNVs, which seems wise.

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