Disc damage likelihood scale

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Glaucoma is a disease characterised by apoptotic ganglion cell death related, at least in part, to intraocular pressure. Any examination for glaucoma must therefore include some comment upon the health of these cells. Unfortunately, it is difficult to examine the ganglion cell layer ophthalmoscopically and so ophthalmologists typically rely on an analysis of a more visible ocular structure—the optic nerve. Ganglion cell death that causes a characteristic optic nerve change—namely, progressive narrowing or loss of the neuroretinal rim, is the hallmark of glaucoma. As scientists and clinicians, ophthalmologists need to both qualitatively and quantitatively describe their ophthalmoscopic impression of the optic nerve both for diagnosis and to establish a baseline so that change may be detected by serial examination. How then should ophthalmologists record their impression of the optic nerve?

Since the late 1960s, the most commonly used quantitative classification of the optic nerve has been Armaly's cup/disc ratio.¹ This staging scale describes the disc using cup diameter as a percentage of overall disc diameter. The cup/disc ratio, especially the vertical cup/disc ratio, represented a significant advance in quantifying glaucomatous optic neuropathy. Its advantages, namely ease of use and lack of magnification artefacts—are appealing; however the cup/disc ratio has two significant problems that limit its accuracy.

As Danesh-Meyer *et al* point out in this issue of the *BJO* (p 437), the two principal limitations of the cup/disc ratio staging system are the fact that the system does not account for disc size and that focal narrowing of the neuroretinal rim is not adequately highlighted. These issues combine to limit the usefulness of the cup/disc ratio for diagnostic accuracy. The effect of disc size is critical when understanding the expected appearance of the optic nerve. It is well known that the size of the nerve is widely variable among individuals,² while the neuroretinal rim area is similar.³ If the rim area is roughly constant, the cup area is directly proportional to disc area.⁴ The effects of disc

size on cup size are apparent in figure 1. Here, I have drawn the theoretical amount of neuroretinal rim for a given cup/disc ratio for three sizes of optic nerve. If one fixes the rim area at about 0.75 mm², it is apparent that for small nerves (1 mm disc diameter) the cup/disc ratio will be about 0.2. The same rim area for a large optic nerve will result in a cup/disc ratio of about 0.85. If cup/disc ratio alone is used as a criterion for damage then it is possible that large optic nerves will incorrectly be called glaucomatous,⁵ and small optic nerves incorrectly will be called normal.

The second issue is that focal changes in the neuroretinal rim that are so characteristic of glaucoma are not readily detected by the cup/disc ratio. For example, in figure 2, the two optic nerve drawings contain identically sized cups and discs. The cup/disc ratio is the same. Even the vertical cup/disc ratio is the same. Yet no one would consider them identical because there is an unequal rim appearance. Ophthalmologists recognise that changes in the rim are often the earliest findings in glaucoma so it stands to reason that a disc interpretation system should highlight the rim, not the cup, as a unit of measure.

A hopefully more accurate way of describing the optic nerve has been created.^{6,7} This scale is based on the neuroretinal rim width for a given disc diameter. The most recent version of this scale is seen in table 1. In the current study, the authors have further investigated the utility of the disc damage likelihood scale (DDLS) and found it to be superior to cup/disc ratio

Figure 1 Theoretical cup/disc ratio by rim area for different optic nerve diameters.

and the HRT-2 for distinguishing between normal and glaucoma or glaucoma suspects. This is an important finding because it bolsters the notion that a glaucoma staging scale based on rim is valid. Some might contend that this study is little more than an exercise in circular reasoning. After all, if the knowledgeable physician uses rim damage as the gold standard, of course a staging system based on rim will be more accurate than one that is based on cup. But that is just the point. The DDLS does nothing more than give physicians a new way to record the observations they were already making. The current study findings, in combination with its excellent reliability,⁷ appear to show the DDLS to be superior to the cup/disc ratio as a way to describe the glaucomatous optic nerve.

There is no such thing as a perfect staging system and the DDLS is no exception. As the authors note, the DDLS is not able to describe tilted optic nerves very well. It is not able to readily detect new areas of rim damage if another area already has more damage. It is susceptible to magnification artefacts. Despite these issues, the majority of nerves will be more accurately described using a rim based scale the accounts for the effects of disc size.

How should the clinician use this information? While the DDLS as a staging scale is a work in progress, the concept of rim width and disc size is immediately applicable. I recommend that physicians measure the diameter of the disc and draw the nerve in the chart. Additional items of interest are then noted on the drawing. I do not recommend determining the cup/disc ratio unless accompanied by a drawing. Least helpful would be recording an isolated cup/disc ratio. For reasons noted above, such a measurement is not worthless, but is essentially useless, as a means of describing the optic nerve or communicating the information to others. Ophthalmologists are already examining the neuroretinal rim to diagnose glaucoma and understand the concept of physiological cupping. Now, by using a combination of disc size and rim width, a more accurate recording of this information is possible.

Figure 2 Two optic nerve drawings with identical cup/disc ratios but with unequal rim width.

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

Br J Ophthalmol 2006;**90**:395–396.
doi: 10.1136/bjo.2005.083360

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Choroidal neovascularisation

Progressive RPE atrophy around disciform scars

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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