Distortion of Vision

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Metamorphopsia, or distortion of vision, is an important visual symptom, particularly if it is the chief reason for a patient seeking optometric assessment. It often indicates serious underlying pathology, and it always demands thorough history-taking and examination. There are many causes of visual distortion, but in most cases with underlying ocular pathology the problem is in the macula. This article discusses the more common causes of such symptoms, including epiretinal membrane, macular hole, age-related macular degeneration (AMD), non-AMD choroidal neovascular membranes, central serous retinopathy (CSR) and vascular maculopathies.

Epiretinal Membrane

An epiretinal membrane (ERM) may be thought of as a delicate sheet of glial “scar” tissue that has formed on the surface of the retina in response to some sort of retinal upset. By far the most common cause is posterior vitreous detachment (PVD), which probably causes slight trauma of the retinal surface, not in itself sufficient to lead to symptoms. A healing response ensues, with the formation of a glial membrane. Like any scar tissue this then contracts, leading to a wrinkling of the retinal surface. This anatomical distortion of the retina, usually involving the macula, may then produce distortion of vision. In some cases the anatomical change is associated with a pathophysiological effect leading to associated water-logging, or oedema, of the macula with further worsening of vision. Sometimes an ERM may be a sequel to previous retinal vascular occlusion, or retinal laser treatment or surgery.

Macular Hole

For reasons that have yet to be established, macular hole is much more common in females than in males. Unlike ERM, most are not associated with PVD. Indeed, it is the non-separation of a contracting vitreous face, leading to tangential traction across the fovea that is thought to be the cause of the condition. Initially this traction produces a detachment of the fovea – known by the misnomer stage I macular hole. The vitreous may separate at this stage to give a PVD, but if not, then the fovea may dehisce to give a small hole – the stage II full-thickness macular hole (FTMH). Over time this hole enlarges, and there is often seepage of fluid beneath the rim of the hole to give the classic appearance of the “punched-out” FTMH (stage III) (Figure 1). Eventually the vitreous may separate to give a PVD, a stage known as stage IV FTMH. Note that there is no loss of retinal tissue, and the hole hardly ever leads to retinal detachment (but can in high myopes). Central vision usually settles at around 3/60-6/60 acuity in an untreated case, and in 10% of people the condition is bilateral.

Age-Related Macular Degeneration (AMD)

Referral refinement of AMD has been discussed previously in this series (see OT May 6 2011 and May 20 2011) and it is probably the most important of the causes of distortion because it is increasingly common and can often be treated. The condition is more common with advancing age. In its “dry” form it is a degeneration affecting Bruch’s membrane and the retinal pigment epithelium (RPE), with secondary changes in the retina. The subject may have noticed a preceding slight reduction in vision, with difficulty in making out detail, and poor reading vision in conditions of reduced illumination. These symptoms

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relate to the “dry” degenerative changes. As Bruch’s membrane is compromised, blood vessels may grow through from the choroid to directly underlie the retina, particularly beneath the central macula. These vessels are fenestrated, and thus leak fluid, which pools beneath the macula and penetrates into it. This is “wet” AMD. As the macula is lifted the vision becomes distorted. The new vessels proliferate beneath the elevated retina and spread out, gradually being replaced by “scar” tissue. The vessels may bleed, and this may result not only in sub- and intra-retinal haemorrhage, but occasionally in breakthrough bleeding into the vitreous.

Increasingly, those affected by AMD are becoming aware that distortion of vision may herald the onset of the wet form of the condition. Note, however, that dry AMD may also cause some distortion. This is usually mild, and fairly static, unlike wet AMD where the distortion worsens almost day-by-day.

Non-AMD Choroidal Neovascular Membranes

Although AMD is by far the most common cause of choroidal new vessels (CNV), there are many other underlying aetiologies that may lead to this same end-point:

- Myopia (Figure 2)
- Sub-retinal inflammatory conditions e.g. presumed ocular histoplasmosis syndrome (POHS) (Figure 3), punctate inner choroidopathy (PIC)
- Trauma
- Angioid streaks (Figure 4)
- Idiopathic CNV

The common factor with these conditions is compromise of Bruch’s membrane and this is what allows new vessels to grow into a sub-retinal position. In some myopes, breaks in Bruch’s membrane may be seen on fundoscopy as “lacquer cracks”. In POHS and PIC scattered foci of inflammation occur, resulting in the characteristic appearance of these conditions, with sub-retinal hypo-and hyper-pigmented lesions (Figure 3). Blunt ocular trauma may lead to large, curvilinear breaks in Bruch’s membrane, which may be complicated at a later date by the growth of CNV. Angioid streaks is a hereditary condition resulting in a degeneration of Bruch’s membrane and the RPE. It is named thus because the resulting breaks in Bruch’s membrane resemble blood vessels on cursory examination (Figure 4).

Central Serous Retinopathy (CSR)

In stark contrast to macular hole, this condition is much more common (20x) in males than females. It typically affects a younger age group, most commonly in the third and fourth decades. Those affected by CSR may have several or many episodes, so the subject may report previous episodes of distortion. The pathophysiology of the condition is not fully understood, but it seems to be related to a temporary breakdown in those mechanisms that control fluid flow across the retina, such that fluid pools beneath the retina. There are usually associated pigmentary changes in the RPE (Figure 5), and subtle RPE abnormalities in the fellow eye may indicate bilaterality even when there have been no subjective symptoms from that eye.

Vascular Maculopathies

There are many vascular abnormalities of the fundus that may affect the macula, but those most often seen in optometric practice are:

- Diabetic maculopathy
- Branch retinal vein occlusion
- Central retinal vein occlusion

Diabetic maculopathy is one of the chief causes of visual loss in people with diabetes and a common cause of sight impairment registration. Retinal vein occlusions are a relatively common cause of sudden visual loss in one eye. In all of these conditions macular oedema may ensue. In the case of diabetic maculopathy there is usually evidence of retinopathy, in the form of microaneurysms, flame-shaped haemorrhages, so-called cotton wool spots, and exudates. In the case of retinal vein occlusion there are scattered flame-shaped haemorrhages, and larger blot haemorrhages, sometimes with cotton wool spots. The distribution of the haemorrhage is in one quadrant of the fundus, in one hemisphere, or in all quadrants, for branch, hemi- and central retinal vein occlusions respectively.

It is unusual for distortion to be the presenting symptom for these
maculopathies, even when macular oedema is demonstrable, and indeed distortion may be a minor symptom for the patient, if present at all. It is not uncommon, however, for patients with macular oedema to complain of micropsia. This occurs because the macular oedema effectively spreads the photoreceptors further apart, and the image is hence seen as smaller. The opposite effect sometimes occurs with the surface-wrinkling maculopathy of ERM.

Taking the history
Not surprisingly, the symptoms of distortion and reduced VA tend to go together. If there is distortion, then that is often described by the patient as their chief complaint. Sometimes, however, it is only mentioned as a symptom after some prodding. If the symptom is not offered by the patient, but they complain of reduced central vision or other central visual abnormality, then you should ask if their vision is distorted. If there is a scotoma, is it absolute, or can the patient see “through” it?

It is important to ask about the duration of the symptom: distortion that has been present for twelve months is unlikely to require same-day assessment. It is also important to determine the degree of recent change in the symptom. As mentioned above, fairly static distortion of a mild degree is not infrequently present in dry AMD, whereas the wet form of the condition, in the acute stage, often produces distortion which can be seen to worsen day by day. This is also true of CNV of other aetiologies. The distortion of macular hole and of ERM usually reaches a steady state, with VA of about 6/60-3/60 in macular hole, and better than this in ERM. Is there a history of previous visual distortion? If so, particularly in a young to middle-aged male, then this is strongly suggestive of CSR.

Examination
The VA may give some clue to the aetiology in stable distortion, with macular hole typically around 6/60 to 3/60, and ERM 6/9 to 6/24 (there are exceptions, of course, and note that VA in ERM is likely to steadily worsen before stabilising); note whether the patient uses eccentric fixation, or has to “search” for the letters. Is the patient myopic?

There may be no clinical signs in the anterior segment, although a history of ocular trauma may be corroborated by pupillary sphincter ruptures, lens subluxation, or phacodonesis. In people with diabetes, and those with suspected previous vascular occlusion, you should examine the iris carefully for evidence of ruberosis iridis prior to dilatation of the pupil.

It is unwise to venture an opinion on the macula without first dilating the pupil. Before focussing on the macula however, examine the vitreous (see OT October 28 2011). A PVD is likely in ERM and unlikely in macular hole.

Examination of the macula at the slit-lamp with the hand-held condensing lens (e.g. 90D or 78D) allows stereoscopic assessment of macular elevation. Look for the signs of degenerative AMD (drusen and pigmentary changes). Can you discern a sub-retinal membrane? Is there sub-retinal blood? Look for lacquer cracks in a myopic patient; these can be very subtle (Figure 2). Can you see the sub-retinal, vessel-like abnormalities radiating from the disc that are typical of angiod streaks (Figure 3)? With this condition, the RPE often has a stippled or granular appearance. Can you see the scattered, well-demarcated, hypo- and hyperpigmented lesions of PIC or POHS (Figure 4)? In the latter the lesions are often scattered in the peripheral fundus, particularly inferiorly. Can you see the classic “punched-out” appearance of a macular hole, which is usually surrounded by a “halo” of slightly elevated retina (Figure 1)? Does the macula have the sheen of an ERM, possibly with corrugations of the retinal surface? Do you see the scattered flame haemorrhages of a retinal vein occlusion (Figure 6), or the microaneurysms, flame-shaped haemorrhages, cotton-wool spots and exudates of diabetic retinopathy?

Optical coherence tomography (OCT) is a useful ancillary investigation in these cases. It is non-invasive, and provides a cross-sectional “picture” of the macula, which aids in confirming the presence of an abnormality and in making the diagnosis.

Management
Of the causes of metamorphopsia discussed above, it is CNV that demands the most urgent management. The drugs Lucentis and Avastin have proved very...
effective in the treatment of CNV with various underlying aetiologies, but if good vision is to be salvaged or retained, then the CNV must be treated in its early stages. This is because the membrane usually grows rapidly, and as it does so it tends to form a layer of sub-retinal scar tissue which permanently separates the overlying retina from the underlying RPE and choroid. Furthermore, the passage of fluid into the retinal substance causes a disruption of its normal cellular architecture which has physiological consequences. Hence, patients with CNV should be referred urgently for assessment and treatment. Same-day assessment via an ophthalmic emergency is not usually necessary, provided the optometrist has established a rapid written referral route (e.g. via fax) to a rapid-access macula or retinal clinic. Since referrals to secondary care may be screened by non-medical personnel, you should state clearly on the referral that the patient has distortion of vision and that you suspect CNV, or wet AMD. If you are unable to refer directly to a clinic providing rapid access for these cases, then referral to the ophthalmic emergency service may be necessary to ensure prompt treatment.

If the diagnosis seems to be CSR (e.g. distortion with moderately reduced vision, with no evidence of CNV or sub-retinal blood, and no pre-disposing condition for CNV, in a young or middle-aged patient) then there is little urgency. This condition is often self-limiting, and generally the first step in management is to allow a period of several months for this to occur, before active treatment is considered. Of course, if there is doubt about this diagnosis, and you feel CNV could be present, then you should refer urgently as above. Macular oedema secondary to diabetic retinopathy or retinal vein occlusion should be treated promptly. These patients should be referred urgently, but same-day assessment is not necessary. Again, it helps to indicate on the referral that diabetic or retinal vascular disease is present.

Macular holes require a surgical treatment, and it is known that the shorter the history, the greater the chance of a successful outcome from surgery. However, on the scale of vitreo-retinal urgency these patients would generally be placed in the “moderate” category, with a wait of weeks to months for surgery on the NHS. There is little to be gained, therefore, from same-day ophthalmological assessment. They should be referred as urgent, with the referral indicating that the diagnosis appears to be macular hole. Epiretinal membranes pass through stages in their development: they form as delicate glial sheets then go through a contractile phase before remaining fairly static, although macular oedema may evolve. The vast majority of ERMs seen in optometric practice are in this third stage: they have stabilized. Hence, when you are counselling patients, you can tell them that it is very likely that their symptoms will not worsen significantly. If you are confident of the diagnosis then referral need not be urgent. State on the referral that the diagnosis is ERM because, as for macular hole, this will help to ensure that the patient is directed to a vitreo-retinal clinic.

When patients are counselled regarding macular conditions some will baulk at the idea of surgery, particularly if they are elderly. It should be pointed out to them that age is rarely a contraindication to vitreo-retinal surgery, which is increasingly carried out under local anaesthetic on a day-surgery basis. The patient may still have many years ahead of them, and they cannot be sure that the “good” eye will remain in good health: a time may come when they rely on the currently “bad” eye. Furthermore, the retina is relatively intolerant of anatomical and physiological disturbance, and with retinal conditions in general, the sooner they are treated the greater the chance of a satisfactory outcome. The results of surgery for ERM, and particularly for macular hole, are very good nowadays, but early intervention is important. It is no good for the patient if they decline referral for surgery and then change their mind five years later because the “bad” eye has become the best prospect for useful vision!

**Figure 6**

Central retinal vein occlusion

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**About the author**

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**References**

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### Module questions

**Course code: C-17277 O/AS/SP/IP**

1. Which of the following statements is **FALSE**? IP optometrists:
   a) Must only prescribe within their level of experience and competence
   b) Must take responsibility for clinical assessment, diagnosis and management, prescribing where necessary
   c) Must contractually adhere to the College of Optometrists' Clinical Management Guidelines
   d) Must lodge an interest in acute eye care, glaucoma or both

2. Considering the factors that affect treatment of bacterial keratitis, which one of the following lists is **INCORRECT**?
   a) Patient age, immunocompetence, general health
   b) Steroid use, co-existing corneal pathology, gender
   c) Size of the lesion, promptness of presentation, location of lesion
   d) Hospital vs. community infection, therapeutic dosage level, patient compliance

3. Which of the following statements regarding the classification criteria for Sjogren's Syndrome is **TRUE**?
   a) Item I or II plus two from Items III, IV, V, and VI indicates secondary Sjogren's
   b) Any 4 items, as long as IV or VI are included, indicates primary Sjogren's
   c) Sensitivity & specificity of referral can be improved if optometrists assess 4 of the 6 items
   d) Phenol red thread is quicker but less comfortable than Schirmer strips

4. Central serous retinopathy:
   a) Usually have some pigmentary changes in the RPE
   b) Only occurs in males
   c) Requires urgent treatment
   d) Is now usually treated with Lucentis or Avastin

5. Choroidal new vessels (CNV):
   a) Resemble lacquer cracks
   b) Can give rise to presumed ocular histoplasmosis syndrome (POHS)
   c) Are the chief cause of angiod streaks
   d) May result from compromise of Bruch's membrane in a variety of conditions

6. Which of the following statements about distortion of vision is **TRUE**?
   a) It often denotes serious underlying eye disease
   b) It often demands urgent assessment in a secondary care setting
   c) It may be best managed surgically, even in patients of advanced age
   d) All of the above

**Course code: C-17573 O**

1. Epiretinal membrane:
   a) May be caused by previous retinal vein occlusion
   b) Causes continuous worsening of vision due to distortion
   c) Is always present when a PVD has occurred
   d) Requires treatment in the vast majority of cases

2. Which of the following statements about a full-thickness macular hole is **TRUE**?
   a) It can be excluded if a PVD is present
   b) It does not occur in males
   c) It does not lead to retinal detachment
   d) It may reduce central vision to 3/60

3. Age-related macular degeneration:
   a) Is always in the "wet" form if distortion is present
   b) Is characterized by retinal wrinkling
   c) Requires urgent assessment if distortion has been present for 2 weeks
   d) Can always be treated with Lucentis or Avastin

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