MYOPIA

How blinding is pathological myopia?

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The risks of visual loss in myopia are sufficiently high to warrant measures to prevent pathological myopia

Myopia may not be a fatal disease, but the economic, social and medical costs of myopia should not be ignored. In east Asian cities, myopia is very common and appears to be rising in some parts of the world.[1 2] Vision in myopia may be restored using optical devices such as spectacles and contact lenses, but high myopia is closely linked to potentially visually disabling eye diseases. An extensive literature has documented a myriad of complications including cataract, glaucoma, myopic macular degeneration, retinal holes, and choroidal neovascularisation.[3]

Although the prevalence rates, natural history, and management of pathological myopia have been addressed in numerous clinical and population based studies, many questions remain unanswered. Firstly, the definition and grading of pathological al myopia is not uniform. There are several other terms also used to describe pathological myopia such as ''degenerative myopia'' and ''malignant myopia.''[4] Duke-Elder defined pathological myopia as myopia with degenerative changes especially in the posterior segment.[5] Tokoro defined pathological myopia as myopia caused by pathological al axial elongation.[6] A more specific definition, myopic retinopathy, refers to the degeneration of chorioretinal tissue associated with axial elongation of the eye.[7] In the Blue Mountains Eye Study, myopic retinopathy included the presence of staphyloma, lacquer cracks, Fuchs' spot, myopic chorioretinal thinning or atrophy, β peripapillary atrophy, cytotorsion or tilting of the optic disc, and the T sign found in central retinal vessels. Shih and co-authors used a grading system for myopic macular chorioretinopathy.[8] MO indicated a normal posterior pole with no tessellation pattern in the macular area; M1 indicated tessellation and choroidal pallor pattern in the macular area; M2 indicated choroidal pallor and tessellation, and the border of an ectasia posteriorly was visualised: M3 indicated pallor and tessellation with several yellowish lacquer cracks in Bruch's membrane and posterior staphyloma; M4 showed choroidal pallor and tessellation, with lacquer cracks with posterior staphyloma and focal areas of deep choroidal atrophy, M5 indicated choroidal pallor and tessellation with lacquer cracks, posterior staphyloma, geographic areas of atrophy of retinal pigment epithelium and choroids, and choroidal neovascularisation were visualised. M3 or greater was defined by Shih *et al* in this issue of *BJO* as ''with maculopathy.''

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Secondly, the level of severity of refractive error associated with specific pathology is not well determined. More severe myopia and longer axial lengths have been linked to specific pathologies such as cataract, glaucoma, or lattice degeneration.[9–11] Current evidence is emerging primarily from cross sectional studies, but there are well conducted cohort studies documenting a temporal relation between myopia at baseline and the development of specific myopia related pathologies during follow up.[12–14] Ideally, population based studies with appropriate sample size, accurate measures of refractive error and biometry at baseline, high retention rate, and detailed documentation of the presence and severity of pathologies during the follow up period should be conducted.

The exact nature of the relation is still unknown: perhaps the association is linear in nature with increased risks of pathology with each unit increment increase in spherical equivalent (SE) in dioptres (D), or unit increase in axial length in millimetres (mm). Another school of thought is that there may be a threshold effect, and the risks of pathology increase exponentially beyond a certain level of refractive error. In the Blue Mountains Eye Study of 3654 adults aged 49 years or older, myopic retinopathy increased from 0.3% for very low myopia of SE >-1.0D, 0.7% for SE -1.00 to -2.99D, 3.0% for SE -3.00 to -4.99D, 11.4% for SE -5.00 to -6.99D, 28.6% for SE -7.00 to -8.99D, to 52.4% for SE at least -9.0D.[7] Common definitions of high myopia used in previous studies include -5.0D, -6.0D, -8.0D, -10.0D, and -12.0D, but these definitions are arbitrary, and there is no uniform consensus on the most clinically useful definition. If there is a threshold effect, future population based studies of the risks of specific pathologies at different levels of myopia could provide new insights on how to set limits for high myopia.

Thirdly, the contribution of pathological myopia towards visual impairment has been underestimated. Myopia related visual impairment may affect the productivity, mobility, quality of life and activities of daily living of individuals. Potentially blinding myopia related pathologies are often irreversible in nature, especially if diagnosed late. In summary, the risks of visual loss in myopia are sufficiently high to warrant measures to prevent pathological myopia. In the Shihpai Eye Study of elderly Taiwanese Chinese aged 65 years or older, myopic macular degeneration was the second leading contributing cause for visual impairment (12.5%), after cataract (41.7%).[15] Choroidal neovascularisation is one of the most common causes of vision loss in pathological myopia.[16] In the 10 year follow up study by Shih and colleagues (p 546) in Taiwan, of 552 adults aged 40 years and above we have high myopia defined as -6.0D or axial length greater or above 25.5 mm. Follow up studies allow the clear temporal documentation of predictive factors and natural history of disease. After 10 years of follow up, 15.9% of adults aged 60–69 years and 26.2% of adults aged 70 years and above had final visual acuity of 20/200 or worse. In addition, the visual acuity deteriorated by at least two lines on the Snellen chart for 62.6% of patients with myopic maculopathy (M3 or greater). The visual prognosis of different types of myopic pathologies could also be evaluated in greater detail. In the study by Shih and colleagues, patchy atrophy and choroidal neovascularisation had poorer visual outcomes compared with lacquer cracks.

While refractive surgery may provide optimal vision correction, there is no evidence that the risks of any of the myopic pathologies, such as chorioretinal degeneration, would be favourably influenced by the surgery. A greater appreciation of pathological myopia by eye care practitioners would facilitate a better understanding of approaches for screening and management of children and adults at risk of pathological myopia. Because of the ''epidemic'' and high rates of myopia even in childhood in east Asian cities, the impact of early

high and pathological myopia in teenagers and young adults should not be overlooked. New evolving technologies for the early diagnosis, medical, and surgical treatment of specific myopia related pathologies, such as laser photocoagulation for choroidal neovascularisation, will improve the prognosis of patients at risks of severe visually incapacitating complications. If the risks of pathological myopia are proportional to the severity of myopia, measures to prevent the early onset and rapid progression of myopia in childhood will eliminate or reduce pathological myopia later in life.

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Optic neuritis

The approach to bilateral simultaneous isolated optic neuritis

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These patients require keen attention from the first ophthalmologist to contact them

In this issue of *BJO* (p 551), de la Cruz and Kupersmith describe a series of 15 adult patients who presented to a neuro-ophthalmic referral practice over a 5 year period with bilateral simultaneous optic neuritis. Patients were not included in this series if they had either evidence of multiple sclerosis or myelopathy, a known systemic disease or medication that is associated with optic neuropathy, or cancer. They were evaluated for systemic disease by a battery of testing including lumbar puncture, which was abnormal in two patients. They indicate that in one case, their patient was ultimately shown to have sarcoidosis, the others did not have a specific diagnosis made (although three (20%) were thought to be post-viral). Their data show that most of these cases have a good outcome when treated with their recommended corticosteroid regimen of 3–5 days of intravenous methylprednisolone 1 g daily followed by a taper of oral prednisone tailored to the clinical course.

Fortunately, the presentation with bilateral simultaneous optic neuritis in adults is unusual, yet because of the devastating implications for the patient, possibly including occult yet active systemic disease, these patients require keen attention from the first ophthalmologist to contact them.

> The key to applying the data in this paper to a patient you might encounter is to know if the entry criteria apply

The key to applying the data in this paper to a patient you might encounter is to know if the entry criteria apply. Certainly, a history and examination to rule out occult demyelinating disease is a given, and if a drug that causes optic neuropathy is being used, it will immediately be a suspect cause. If the patient has a known neoplasm, then one will carefully search for a process such as carcinomatosis, which could affect each optic nerve, or for a metastasis that involves the chiasm. But the protocol proposed by de la Cruz and Kupersmith does not apply if the patient has a known systemic disease. Thus, a major issue is just how far does one go in such a patient to search for an occult underlying process that could cause bilateral optic neuropathy because, if it were found, not only would the lessons of this series not apply, but the systemic process might require urgent specific therapy.

One should first take a detailed history looking for undiagnosed chronic systemic disease or a recent systemic illness or heraldic event. In a patient who had received a vaccination 2 weeks earlier, one will strongly consider post-vaccinial optic neuritis. If there was a significant viral syndrome just before onset, one will consider post-viral optic neuritis. We also typically inquire about symptoms that would be consistent with collagen-vascular disease, vasculitis, or occult infection (for example, lues) or inflammations (for example, sarcoidosis). Thus, we ask about symptoms such as arthralgia, myalgia, haematuria, haemoptysis, chronic fevers, rashes, parotid/facial swelling, mucosal ulcerations. As an example, the presence of asthma or chronic cough of relatively recent onset makes us think of processes such as sarcoid, vasculitis, or tuberculosis. Mucosal membrane ulceration makes us think of infections such as syphilis or Behçet's disease. Recurrent thrombosis or early fetal wasting make us consider anti-phospholipid antibody syndrome.