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6.1 The Burden of Pathologic Myopia

As described more specifically in the earlier chapters, pathologic myopia, also known as ‘malignant myopia’ or ‘degenerative myopia’, is a major growing public health problem worldwide [1, 2]. The condition is particularly prevalent in Asian and Middle Eastern countries [3]. Clinically, pathologic myopia is associated with the progressive and excessive elongation of the globe, which may be accompanied by degenerative changes in the sclera, choroid, Bruch’s membrane, retinal pigment epithelium and neural retina [4, 5]. The elongation of the eyeball leads to the development of myopic refractive error

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and related complications [6]. Recently studies have shown that the eyeball continues to elongate even in the fourth decade of life in individuals with pathologic myopia [6]. From a public health perspective, pathologic myopia is associated with vision deterioration, vision-specific disability and a worsening in specific domains on quality of life (QoL) [7], i.e. decreased work productivity, reduced mobility and restricted activities of daily living [8]. In addition, the condition is also associated with legally blinding ocular complications, such as glaucoma, retinal detachment, myopic maculopathy, myopic retinopathy and premature cataracts [2]. Pathologic myopia is naturally associated with a high dependence of contact lens wear which leads to a higher prevalence of contact lens complications [9]. Despite a plethora of information on myopia, very little evidence-based data are currently available on pathologic myopia specifically. Where necessary, the authors have extrapolated and/or estimated what is known about myopia (especially high myopia) for pathologic myopia.

6.2 Social Determinants of Pathologic Myopia

The social determinants of health (SDH) are defined by the World Health Organization (WHO) Commission on the Social Determinants of Health as ‘the conditions in

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Table 6.1 The social determinants of pathologic myopia

Social determinants	Description
Race and ethnicity	Highest prevalence among Asians, i.e. Taiwanese [11], Japanese [7], Singaporeans [12] and Chinese [13] Compared to Asians lower prevalence in African and Pacific Island groups [14] When compared to African Americans and/or Mexican Americans, higher prevalence found in Whites [15]
Age	Clinically significant pathologic changes have also been found in patients who are middle-aged (working life) or younger [16]. The incidence and severity of pathologic signs increases with age. For instance, the visual acuity of high myopes decreases significantly as individuals age, which may be the result of complications including lacquer cracker, submacular haemorrhage, Fuchs spots and chorioretinal atrophy [17]
Gender	Higher prevalence in women than men [15, 17]
Social group	Higher prevalence in young (particularly Asian) children [18, 19] and young and professional working adults [20, 21]
Geography	Higher prevalence in industrialised/developed nations [22, 23] Within nations there are rural–urban differences, i.e. inner-city urban areas have higher odds of the condition than outer suburban areas [20]
Lifestyle	Associated with amount of time spent outdoors, i.e. total time spent outdoors was associated with less myopia, independent of indoor activity, reading and engagement in sports [24, 25]
Education	High prevalence in individuals with high level of education/academic achievement [21, 26, 27]
Occupation	Associated with near work indoors [28]. For example, people whose profession entails substantial reading during either training or performance of the occupation (e.g. lawyers, physicians, microscopists and editors) have higher degrees of myopia [29–33]
Familial inheritance (parental refraction)	Heritable myopia susceptibility – there is a positive correlation between parental myopia and myopia in their children [34–37], particularly if both parents are myopic [19]

which people are born, grow, live, work and age’. These conditions or circumstances are shaped by families, communities and the distribution of money, power and resources at worldwide, national and local levels and affected by policy choices at each level [10]. The public health impact of pathologic myopia is also shaped by the underlying SDH. Table 6.1 summarises the key SDH shaping the prevalence and incidence of pathologic myopia.

6.3 Public Health Impact of Pathologic Myopia

6.3.1 Population Impact

The prevalence and severity rates of myopia are increasing [38]. Pathologic myopia is now one of the leading causes of legal blindness in many developed countries [5, 39–43]. The worldwide distribution of those who have degenerative myopia is less clear. In a survey of 15 countries more than 50 years ago by Fuchs [44], the prevalence of progressive pathologic myopia was as low as 0.2 % in Egypt and as high as 9.6 % in Spain [44].

Recent studies have continued to reflect the wide variations in estimates of pathologic myopia prevalence but suggest that the condition affects a significant proportion of the population in numerous countries [11, 12, 45–48]. The wide variation in prevalence rates is suggestive of a geographic and/or ethnicity influence: the Blue Mountains Eye Study in Australia reported a 1–3 % prevalence rate [49]. However, the condition has been found to be especially common in Asia where prevalence rates can be as high as 5 % [42, 48, 50]. In the United States, of those with high myopia, approximately 27–33 % of all myopic eyes have the pathologic form, which subsequently corresponds to a prevalence of 1.7–2 % [47].

Pathologic myopia is also associated with higher risk of other ocular complications. For example, the risks of developing macular choroidal neovascularisation are nine times higher in those with pathologic myopia compared to only two times higher in mild myopic individuals [49, 51, 52]. Similarly, in the Blue Mountains Eye Study, glaucoma was present in 4.4 % of pathologic myopic eyes compared to eyes without myopia [53]. In addition, in those with pathologic myopia, the yearly incidence of retinal detachment has been estimated to be 3.2 % [54–56]. The prevalence of pathologic myopia and its complications are discussed in more detail (see Chap. 3).

6.3.2 The Impact of Pathologic Myopia on Health-Related Quality of Life

Health-related quality of life (HRQoL) is a multidimensional concept and has been defined by the World Health Organization (WHO) QoL group as:

An individual’s perception of her/his position in life in the context of the culture and value systems in which she/he lives and in relation to goals, expectation, standards and concerns. It is a broad ranging concept affected in a complex way by the person’s physical health, psychological state, level of independence, social relationships, personal beliefs and her/his relationship to salient features of the environment [57].

The HRQoL and economic burden of pathologic myopia is not well understood, as few reports are available relating *specifically* to pathologic myopia and recent data are lacking. Furthermore, the HRQoL is likely to be different in individuals with high myopia without complications, as opposed to individuals with high myopia with complications [58]. As complications tend to develop later in life for individuals with high myopia, the HRQoL and ocular conditions will vary at different stages of life [58].

Using a 32-item questionnaire (instrument not specified by study authors) [59], Takashima and colleagues found that HRQoL in Japanese patients with pathologic myopia was reduced compared with control subjects with no ocular disease and minimal refractive error [60]. In particular, they found that those with pathologic myopia had significantly lower scores in ‘vision-related daily tasks’, ‘social impact’, ‘eye satisfaction’ and ‘life satisfaction’ and higher scores in ‘understanding of eye disease’ compared to control subjects. However, there were no differences between ‘emotional well-being’, ‘leisure and support’ and the General Well-Being Schedule (GWBS) between those with and without the disease. Participants with pathologic myopia had significantly lower mean scores in ‘role limitations and social life’ and ‘disability’ and significantly higher scores in ‘support’ compared to controls. In summary, using a vision-specific QoL questionnaire and two global measures of eye and life satisfaction, pathologic myopia patients reported a significant impact on HRQoL.

Similarly, in a UK study, patients with high myopia (defined as ≥ -10.00 dioptres (D) in the worse eye and ≥ -8.00 D in the better eye) had significantly worse visual functioning (assessed by the Visual Function Index, VF-14 scale [61]) and QoL (assessed by the Vision-related QoL questionnaire, VQOL [62]) compared to patients with moderate (-4.00 to -9.75 D in the worse eye) and low myopia (-1.50 to -3.75 D in the worse eye and at least -1.00 D in the better eye) [63]. Moreover, there were no significant differences in visual functioning, and QoL between high myopia and keratoconus patients suggesting the magnitude of the patient-centred impact is similar in these groups [63]. This study also explored the HRQoL impact of myopia using qualitative interviews. The evidence suggests that high myopes have psychologic, cosmetic, practical and financial impacts. Patients felt that high myopia dominated life from early age, resulting in a lack of self-confidence, social isolation and difficulty forming relationships and difficulties with sports participation especially swimming. Dependence on optical correction for functioning was a constant daily concern for patients with high myopia, who felt that wearing thick spectacles was an unsightly social handicap. Patients with high myopia also reported discomfort wearing contact lenses [63]. Finally, in a large study of Chinese adolescents,

higher levels of myopia were significantly associated with worse visual function [64]. Children with refractive error ≥ -0.5 D had a mean self-reported visual function of 82.6 ± 13.9 , which declined monotonically to 57.6 ± 15.5 for children with higher levels of myopia < -5.5 D [64].

Given that pathologic myopia and its disabling complications frequently result in low vision, the vision-related QoL impact of the disease is likely to be similar to that found in low-vision patients. Low vision is defined by WHO as visual acuity $\leq 6/18$ that cannot be corrected to normal level with conventional spectacles, contact lenses or surgery, thus excluding people who can have vision restored or corrected to within normal limits [65, 66]. Overall, low vision is significantly associated with decreased functioning and QoL and increased emotional distress [67]. In a cross-sectional study of low-vision patients (mainly with age-related macular degeneration, glaucoma and diabetic retinopathy), visual functioning was extremely low using the VF-14 questionnaire [68]. Similarly, using the 51-item field test version of the National Eye Institute Visual Function Questionnaire, patients reported low mean scores in QoL domains of general health, general vision, near activities, distance activities, vision-specific expectations, vision-specific role difficulties, driving and peripheral vision [68]. Moreover, patients with low vision scored significantly worse than normal-sighted patients aged over 75 years, patients with congestive heart failure and patients with clinical depression in the SF-36 (Short Form 36 Health Survey) HRQoL domains of physical functioning, role limitations caused by physical health problems and role limitations caused by emotional problems [68].

6.3.3 Economic Impact of Pathologic Myopia

Data on the economic impact of pathologic myopia are scarce although some for myopia exist. In 1990, the general cost associated with myopia was estimated at US\$4.6 billion globally [69]. In Australia, the health costs imposed by myopia on individuals and the community are considerable, running into hundreds of millions of dollars a year for spectacles, contact lenses and refractive surgery alone [70]. In Singapore, it has been estimated that myopic persons spend \$US90 million annually on spectacles alone [38]. The direct cost of myopia for Singaporean teenagers is estimated to be US\$25 million annually [71]. These economic estimates do not include indirect costs such as lost workdays, restricted activity days, caregiver costs, cost of suffering associated with untreated myopia and ongoing medical research into myopia. Moreover, these calculations do not include other medical costs, such as those associated with morbidity arising from myopia, especially pathologic myopia, such as retinal

detachment, glaucoma and cataract, associated visual disability and blindness, and complications arising from the use of contact lenses such as potential corneal infections and scarring [38, 71]. Treatment for myopia-related complications has been estimated at \$2–2.5 million annually in Singapore. About 300 retinal detachments are operated on each year (although not all are attributable to myopia) [72], and around 950 contact lens complications over a 2-year period are treated at public hospitals in Singapore [73]. Further research is required to determine the direct and indirect costs associated specifically with pathologic myopia and its associated ocular complications.

6.4 Public Health Strategies to Minimise the Impact of Pathologic Myopia

Though most individuals with myopia will only develop low to moderate levels of the condition, some will progress to pathologic myopia [74]. From a clinical perspective, interventions to stem progression include optical correction by spectacles, contact lenses (i.e. overnight orthokeratology contact lenses) [75], and refractive surgery; scleral strengthening [76]; and pharmacological interventions [76, 77]. However, the effectiveness of these interventions has been inconsistent. For example, the protective effects of optical devices disappear with long-term use [77]. Refractive surgery merely changes the shape of the anterior ocular surface but does not alter the shape of the elongated eyeball and the associated risk for retinal complications [77]. Anticholinergics (e.g. atropine), a substance that blocks the neurotransmitter acetylcholine in the central and peripheral nervous systems and has papillary dilation properties in the eyes, has been applied in combination with bifocals in an attempt to decrease the accommodative requirements of the eyes and stem the progression of myopia. However, results from recent trials have been disappointing – with most demonstrating only marginal clinically significant reductions in myopia progression [74, 78]. For all these interventions, further validation and analyses are still required [77].

Currently, the exact pathogenesis of pathologic myopia remains unclear, and both environmental and genetic factors appear to contribute to the development of the condition [78, 79]. As a result, research efforts for an effective intervention to slow progression have been hampered [78, 80]. However, evidence suggests that even minimal reductions in progression might provide substantial benefits to the individual non-clinically [77]: Several studies have reported HRQoL improvements following treatments. For example, Leong and colleagues found that using the Quality of Life Impact of Refractive Correction (QIRC) questionnaire, HRQoL was significantly better in patients who had received implantable Collamer Lens compared to contact lens wear, particularly in

the areas of freedom from reliance on refractive correction on waking, during travel and during sports [81]. Similarly, patients receiving bilateral anterior chamber angle-fixated phakic intraocular lens (pIOL) implantation for high myopia reported significant improvements in several areas of QoL (measured using the Refractive Status and Vision Profile (RSVP) questionnaire), including concern with vision, driving, glare, optical problems, physical/social functioning, problems with corrective lenses and ocular symptoms [82]. LASIK surgery for patients with high myopia also appears to result in important functional and QoL gains [83].

In the interim, whilst the exact pathogenesis of pathologic myopia is still being investigated, the main objectives of public health strategies should be to reduce the incidence of myopia and the progression to pathologic myopia. Public health approaches should also aim to prevent, minimise, maintain or improve the well-being of individuals already affected. We therefore recommend the following five strategies to counteract the negative public health impact of pathologic myopia:

6.4.1 Strategy 1: Health Behaviour Intervention Programmes

A recent systematic review and meta-analysis of 23 studies in children and adolescents found that time spent outdoors may have a protective effect in reducing the prevalence of myopia and therefore may slow the progression to its pathologic form [84]. Health behaviour intervention programmes could therefore focus on encouraging both young adults and children to achieve a healthy balance of outdoor physical activity and near work, such as reading [74, 78, 80]. There are currently two trials undertaken in China (NCT00848900) and Singapore (NCT01388205) which are investigating the effect of additional time spent outdoors on minimising occurrence and progression of myopia during school hours and incorporating family engagements in outdoor activities, respectively. Outcomes of these trials are expected in either later 2013 or 2014. It is important to note that though these studies and trials targeted myopia rather than pathologic myopia per se, early intervention, i.e. before an individual develops pathologic myopia, may reduce progression to the pathologic stage of the disease.

6.4.2 Strategy 2: Health Promotion Programmes and Screening to Improve Awareness

People with high myopia (spherical equivalent of at least -6.0 D/ -8.0 / -10.0) are often not aware that their impairment could progress into a more complicated form

and/or other ocular complications [2, 77]. Therefore, public health promotion campaigns at the national, regional, or community level should attempt to improve this awareness via public health messages to the public. Moreover, health-care professionals should inform and educate their patients to detect early signs of their myopia progressing towards its pathologic state, as well as ensuring timely referrals for further ophthalmic assessments are made [77].

6.4.3 Strategy 3: Regular Monitoring of Vision Status and Vision-Related Quality of Life

Individuals at high risk of pathological changes, i.e. those with high myopia and presenting with the one or more of the social determinants of having the condition (Table 6.1), could undergo regular clinical screening and vision-specific functioning monitoring with their general health practitioner, optometrist and/or ophthalmologist. Individual advice should be sought from the attending eye health professional on the ideal interval between regular check-ups. Screening activities should include fundus photography to detect macular and retinal lesions as well as optical coherence tomography (OCT) measures to provide a more detailed understanding and sensitive detection [85]. Patients who develop choroidal neovascular membrane (CNV) should then be treated [85, 86]. Patients' vision-related functioning and QoL should also be regularly monitored by their health practitioners to assess for any reductions in daily living activities, emotional and functional well-being [87]. Several functioning and QoL instruments such as the VF-14 [88] and the IVI [87], respectively, are currently available and have been comprehensively validated using modern psychometric theory such as Rasch analysis.

6.4.4 Strategy 4: Low-Vision Care and Rehabilitation

Pathologic myopia and its ocular complications (such as glaucoma, cataract, degenerative changes in the sclera, choroid and retinal pigment epithelium) almost always result in the patient developing low vision [39]. For example, in 90 % of patients who develop CNV, the majority has visual acuity $\leq 6/60$ in the affected eye 5 years after onset [85, 86]. Thus for those affected, low-vision care and rehabilitation are highly recommended as these have been shown to result in significant improvements [67, 68, 89]. Low-vision services include clinical, rehabilitation services and the use of adaptive technologies. Clinical low-vision care involves a comprehensive examination of the eyes and vision by an ophthalmologist and/or an optometrist, including an assessment of visual

function. Rehabilitation services refer to assistance with activities of daily living, counselling, orientation and mobility training, peer support groups, community and social services, advocacy (support groups and organisations) and education and employment and training [89].

6.4.5 Strategy 5: Implantation of Public Health Research Initiatives

Finally, investments into innovative public health-related pathologic myopia research are required. The current literature is limited on practical interventions to prevent and stem the progression of pathologic myopia. Future research could include investigating cost-effective behavioural interventions and health promotion activities in the primary care and community setting. Studies are also required to elucidate the gene-environment interactions in the pathogenesis of pathologic myopia and to determine the impact of SDH on shaping the pattern of disease. Understanding the SDH in pathologic myopia patients will assist in better risk profiling of individuals who could benefit from interventions to prevent progression to high myopia.

Conclusion

Pathologic myopia and its ocular complications can cause severe vision deterioration and substantial reductions in vision-specific QoL. The public health impact of the condition is substantial and is shaped by certain social determinants of health (SDHs). As the prevalence and severity of myopia continue to rise, especially among Asians, pathologic myopia will become one of the leading causes of blindness for many developed countries. Studies have established a significant negative impact of high myopia on QoL, in the areas of activity limitation, and emotional and social well-being. Often the consequence of developing the condition is low vision. Though no data currently exist (and therefore further research in this area is warranted), one can anticipate the substantial direct and indirect economic impact of pathologic myopia by extrapolating the cost data available on myopia in general. Whilst the long-term effectiveness of current medical treatment are being investigated and established, the main goals of public health strategies should be to reduce the incidence of myopia, particularly high myopia, and the progression to pathologic myopia. We have proposed five public health specific strategies to achieve these goals. They include the following: to develop and implement health behaviour intervention programmes, execute health promotion programmes and screening initiative to improve community awareness, conduct regular monitoring of vision status and vision-related QoL, deliver quality low-vision care and rehabilitation and invest in public

health research initiatives to further enhance and improve the first four strategies. The public health problem of pathologic myopia is growing, but some of this challenge can be met with the implementation of evidence-based public health interventions.

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