Amblyopia: An overview

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Introduction

Amblyopia, or commonly referred as lazy eye, can be defined as a reduction in vision with no demonstrable abnormality of the visual pathway that is not immediately resolved by refractive correction.[1] With a prevalence of 3-5%, amblyopia represents a major public health problem. In fact, amblyopia is the leading cause of monocular vision loss in the United States in people younger than 40 years.[2] It develops during maturation of the visual pathway and is largely reversible during the first seven to eight years of life. This is known as the 'critical period'. In some situations the critical period may be extended.[3]

Causes

Amblyopia may develop when the images coming into one or both eyes are either blurred or obscured. There is an abnormal binocular cortical interaction and results in a loss of acuity, contrast sensitivity and/or positional disorder.[4] Amblyopia is usually classified by cause.[1] However, it is not uncommon for the types to co-exist. It can be grouped into three major categories.

(1) Strabismic amblyopia: when it is due to the presence of a squint
(2) Refractive amblyopia: it can be subdivided into
a) Anisometropic amblyopia: where there is a large difference in refractive error between the two eyes
b) Meridional amblyopia: where there is a significant degree of astigmatism
c) Ametropic amblyopia: where the refractive error is so significant that neither eye receives a good quality images.
(3) Stimulus deprivation amblyopia: where, for example, a cataract or ptosis obscures the visual axis

Strabismus means the eyes are not looking in the same visual direction and is not necessarily giving rise to amblyopia, especially in intermittent type with freely alternating fixation. In Hong Kong, intermittent exotropia (divergent squint) is the most common type of squint which is less commonly associated with amblyopia. However, if the condition becomes constant and/or has a strong fixation preference, amblyopia may arise while esotopia (convergent squint) is more commonly associated with amblyopia.

Refractive amblyopia is the result of a blurred visual image being formed on the retina. Among the three subtypes, anisometropic amblyopia is the most common one and hypermetropic (long-sightedness) anisometropia is more amblyogneic. A difference of more than +1.0 D is generally considered to be of significance and should be corrected with spectacles. Myopic anisometropia is less commonly resulting in amblyopic since they can see clear at some point in the near fixation. Meridional amblyopia is due to the presence of high astigmatism which means the refractive power along the two perpendicular axes has a significant difference. There is no general consensus as to how much astigmatism is amblyogneic. The presence of 1.5 to 2.0 D of astigmatism will generally require spectacle correction. Ametropic amblyopic usually refers to the condition that there are high degrees of refractive error in either one or both eyes.

Stimulation deprivation amblyopia is due to the presence of structurally abnormality preventing the formation of clear image in the visual pathway. The most commonly cause is ptosis that obscuring the visual axis. Other causes include cataract, lid or orbital masses.

Screening

Amblyopia is a deficit of vision that has to be treated within the critical period. Effective treatment depends on early detection, and a broad consensus of professional opinion supports vision screening of infants and young children. Screening programmes have been set up to detect this largely asymptomatic condition and refer children for treatment. There are several issues concerning screening that have not yet been fully resolved and these include when to screen, how to screen and who should do the screening. Currently there are a variety of recommendations for vision screening programmes and a number of different approaches to providing the services. The battery of tests carried out usually includes monocular visual acuity testing with an age-appropriate test with or without assessment of extraocular muscle function, binocular status, and colour vision assessment. No single method of screening has been demonstrated to be superior in detecting amblyopia and all methods have significant limitations.

Most screening programmes to detect strabismus and amblyopia, or their precursors, have been with children between 3 and 6 years.[5-9] There has been a great deal of debate regarding the optimum time for visual screening. Amblyopia screening as such is not generally practical in infancy. However, an alternative approach is to screen for early strabismus and strabismogenic factors that are precursors and predictors.[10]

Vision screening for preschool children has employed traditional visual acuity - based methods to evaluate visual functions.[11] These tests typically involve reading optotypes - such as Allen Symbols, LEA figures, HOTV Letters, and Snellen Acuity - using numbers or letters. The advantage of these optotype visual acuity-based techniques for preschool vision screening is that they provide a direct measure of visual function. However, successful screening by means of optotype visual acuity testing requires an older and more cooperative child and is less effective in younger children who have limited attention spans. Results of these testings are also highly dependent on the skills and experience of the examiners. Newer technologies such as photoscreening and automated refraction have prompted a re-evaluation of visual screening. However, they detect problems associated with the development of amblyopia, i.e. amblyogenic factors, instead of detecting amblyopia directly. While the natural history of many amblyogenic factors is not well understood, not all patients with amblyogenic factors will develop amblyopia. However, the risk of developing amblyopia appears to increase along with the magnitude of amblyopic factors.

Recommendations regarding who should carry out screening for amblyopia vary. In America practice varies from state to state and preschool vision screening is carried out by a variety of professional, volunteer and lay professional, volunteer and lay personnel.[12,13] In Sweden nurses carry out a vision screen as part of other healthcare surveillance checks.[14] In some places trial of home vision screening kits for parents have been conducted.[13,15] In the UK orthoptists have been shown to compare favourably with other screening personnel. In a comparative trial in Newcastle estimates of 100% sensitivity and 97.1% specificity were calculated for orthoptists undertaking vision screening in three year olds. Health visitors achieved better specificity at 100% but managed a sensitivity of only 50%.[16] Nurse and lay screeners can achieve similar sensitivity, when specificity is set at 0.90, for detecting preschool children in need of a comprehensive eye examination.[17]

In an attempt to provide standards for which conditions should be detected, the American Association for Pediatric Ophthalmology and Strabismus (AAPOS) Vision Screening Committee has reviewed the literature and introduces a set of standard risk factors that should be detected with preschool vision screening. These risk factors are derived from consensus, reviews of surveys of paediatric ophthalmologists, evaluation of "gold-standard" criteria from large vision screening programmes.[11, 18-20]

Vision in Preschoolers Study Group[21] also defined their targeted groups with slightly different criteria. Children were referred to specialists in Singapore according to their local guidelines.[22]

Chan et al suggested referral criteria for Hong Kong children should be set as hyperopia of ≥+2.00 D, myopia of ≥-1.00 D, astigmatism of ≥1.00 D and anisometropia of ≥ 1.25 D.[23] Sensitivity using only these criteria for abnormal refraction in identifying children with amblyopia, esotropia, exotropia and subnormal vision (<6/12) was respectively 100%, 84.6%, 45.2% and 95.7%. The overall sensitivity for the identification of visual problems was 86.1% and the overall specificity was 76.0%.

Treatment

Currently, the most commonly employed treatments of amblyopia include correction of the refractive error with spectacles, patching of the better eye and penalisation of the better eye with atropine, along with treatment of other associated underlying disorders.

For anisometropic amblyopia, constant wear of a pair of corrective spectacles alone can improve visual acuity pair of (VA) in many cases with moderate amblyopia.[24] This can be combined with patching at the beginning or when the VA shows no further improvement after wearing glasses alone. This will depends on the preference of the ophthalmologist, the density of the amblyopia and also the compliance of spectacles. In case of meridional and ametropic amblyopia, there are still no well designed studies but the same treatment principles are usually adopted.

However, in children without documented amblyopia but having just mild to moderate refractive error that may be amblyogenic, there is still some controversy as to when spectacles should be recommended. In our experience, a simple rule of 1-2-4 can be considered. When there is hypermetropic anisometropia of +1.0 D, astigmatism of 2.0 D or myopia of -2.0 D or a hypermetropia of +4.0 D, spectacles can be considered even though there may not be amblyopia. Spectacles are not a must in these conditions if the uncorrected visual acuity is satisfactory. However, in the presence of 2.0 D astigmatism or myopia, the uncorrected vision may not be good enough for learning, especially Chinese characters as they are more complex in structure.

Patching conceptually forces the patient to use the amblyopic eye by occluding the better eye. However, the regimen of patching is not well established. Some will prefer aggressive treatment with full time patching that run a risk of occlusion amblyopia, i.e. development of amblyopia in the sound eye because of occlusion depriving it of visual stimulation. More commonly, it is done in a dosing method. A general rule is the hours of patching daily equal the years of age of the patient and this is then stepped up or down according to the response of the patient.

In the recent reports by the Pediatric Eye Disease Investigator Group, the regimen may be simplified further. In their study group of patients aged between 3 and 7 with moderate amblyopia, defined as visual acuity ranging from 20/40 (6/12) to 20/100 (6/30), patching is slightly more effective than atropinisation and a 10 or more hours of patching showed a more rapid response than lesser hours of patching for patients in the range of 20/80 (6/24) to 20/100 (6/30).[25] Although by 6 months, the difference in improvement between the two treatment groups is not significant.[25] For patients with moderate amblyopia, 2 hours of daily patching is equally effective as 6 hours of daily patching.[26] For severe amblyopia with a range of 20/100 (6/30) to 20/400

(6/120), 6 hours of daily patching has similar effect as full-time patching.[27] Moreover, in other reports by the same group, patching is also effective in treating some amblyopic patients aged between 7 and 18 years old.[28, 29]

Although patching and atropinization is effective in treating amblyopia, the vision still may not be fully restored. A two-year follow up of the moderate amblyopia group treated either by patching or atropine showed that the amblyopic eye was still 2 lines worse than the sound eye.[30]

Another issue regarding treatment of amblyopia is recurrence after cessation of treatment. Nearly one fourth of successfully treated amblyopic children had a recurrence within the first year off treatment.[31] The risk factors include patients treated with 6 or more hours of daily patching and then stopped abruptly without weaning to 2 hours of patching, better visual acuity at the time of cessation of treatment, a greater number of lines improved during the previous treatment and a history of recurrence.[31, 32] These patients may need to be followed up more carefully.

The other issues that are still not answered in these studies include the treatment protocol for patients below the age of 3 years and the effect of different treatment methods for severe amblyopia. In young toddlers, long patching hours may have a higher risk of occlusion amblyopia.

Summary

Amblyopia can usually be treated effectively with either glasses or occlusion therapy or both. This depends on early detection by personnels with relevant experience since they are usually asymptomatic in the case of refractive amblyopia. Patients with strabismic amblyopia may be brought to attention earlier because of the squint condition may lead parents to seek for medical advice. Only through effective screening and a high index of suspicion, we can salvage the vision of our children.

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