Br Ir Orthopt J 2010; **7:** 8–13

What does that mean? A review of some aspects of orthoptic terminology

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Abstract

Aim: **To highlight some areas where there is potential for confusion regarding terminology within orthoptics and to discuss the evidence surrounding these topics.**
Methods: **A literature-based review was performed. Relevant material was identified using Google, PubMed, and an orthoptic journals/conference transactions search facility. A forward citation search was also performed using Web of Knowledge.**
Results: **Ambiguous terms have been highlighted in the areas of strabismus surgery, microtropia, abnormal retinal correspondence and critical periods for visual development. In strabismus surgery, the terms 'functional' and 'cosmetic' have double meaning and can even be inaccurate. Microtropia has undergone numerous name changes over the years – some geographical, and others due to its multiple clinical presentations. In addition, abnormal retinal correspondence is a term that has become ambiguous due to the advent of macular translocation procedures which physically move the retinal points. Lastly, 'critical period' is frequently used in the singular and applied to amblyopia treatment. However, evidence has demonstrated there are three critical periods applied and that the timing of these varies according to which aspect of visual function is being evaluated.**
Conclusion: **Regardless of how ambiguities in orthoptic terminology arise, many issues can be resolved by simply updating the terms used. However, when conducting a review of the literature, the evolution of orthoptic terminology must be accounted for, to ensure accurate evaluation.**

Key words: Abnormal retinal correspondence, Critical periods, Microtropia, Strabismus surgery, Terminology

Introduction

Terminology in medicine is continually changing and developing. There are many examples of words which were once in common use but are now discarded. For example the clinical terms previously used to describe people with reduced IQ were moron (if they had an IQ of 50–69), imbecile (IQ 20–49) or idiot (IQ less than 20).[1] Other words, including retarded, stupid and feeble-minded, were considered valid descriptors in the psychological community but have all now passed into common slang use, purely in a derogatory context.

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Introduction

Within orthoptics, as research expands our knowledge about conditions being treated on a daily basis, what we say may no longer necessarily be what we mean. For example, in 1907 Maddox[2] used the phrase 'newly-acquired field of fixation' to refer to the condition we now call abnormal retinal correspondence. Terms can also have double meaning: a binocular visual acuity result may be interpreted as the acuity tested with both eyes opened,[3] as opposed to the maximum visual acuity obtainable while maintaining binocular single vision.[4] To try to eliminate this confusion, the term 'controlled binocular acuity'[5] is now used to describe the latter type of assessment.

There are a number of implications of these changes in terminology, in particular in education (helping students to interpret information from a variety of sources) where there is a large focus on evidence-based practice and research. With journals from all over the world easily accessible, it can be difficult to evaluate outcomes between studies when the terms are not defined in the same way. In fact, achieving full coverage of the literature can be difficult without being fully aware of the array of alternative search terms. Thus a literature-based review has been performed, with relevant material identified using Google, PubMed and an orthoptic journals/conference transactions search facility. In addition a forward citation search was done, using Web of Knowledge. The purpose of this paper is to highlight some areas where there is potential for confusion regarding terminology within orthoptics and to discuss the evidence surrounding these topics.

Strabismus surgery

When discussing the surgical treatment options for strabismus, the terms 'functional' and 'cosmetic' are frequently used by clinicians to differentiate between those who have binocular vision or the potential to develop it, and those who do not. However, these terms could be considered misleading or even incorrect. Cosmetic, in the medical sense, refers to the correction of a defect;[6] thus its application to strabismus repair is correct. However, a patient may interpret the word as meaning 'decorative' or 'appearance-enhancing'. There is substantial evidence that strabismus surgery does not just enhance appearance.[7–11]

For example, analysis of questionnaire responses administered to patients before and after strabismus surgery demonstrated that they experienced statistically significant improvements in a number of aspects. These included specific health problems such as diplopia and headaches, daily tasks including driving and sporting activity, social interaction, self-image, and job prospects.[7] Although objective evaluations of the cosmesis of patients with strabismus are to be regarded with caution, as subjective opinion may differ between patients, one study reported that 92.5% of dating agencies judged that strabismic subjects would have greater difficulty finding a partner.[12] This is not an unreasonable supposition given that adults with strabismus describe problems with eye contact and their appearance to others. Thus the benefit gained from strabismus surgery amounts to more than just the mere straightening of the deviating eye – and if 61% of all patients with strabismus are willing to trade part of their life expectancy for being rid of the affliction and its associated effects,[8] does use of the term 'cosmetic surgery' trivialise the impact of this condition upon the patient?

In addition, given that patients frequently possess an expanded binocular field of view[10,11] or peripheral fusion[13] following surgical correction of long-standing esotropia, it could be argued that correction of strabismus in individuals deemed to have no binocular potential has a functional purpose after all. Though these studies were conducted with patients possessing mild amblyopia, development of peripheral fusion appears possible even in those with higher degrees of amblyopia.[14] This also raises the question of whether the tests used to determine post-operative binocularity are sensitive enough for orthoptists to discriminate between those with binocular potential and those without. Khan *et al.*[15] reported that in those undergoing prismatic post-operative diplopia testing, only 7% of those testing positive for diplopia experienced it in free space following botulinum toxin administration. This demonstrates that clinical tests are not entirely predictive of treatment outcomes. Binocular potential in patients with a long-standing esotropia is an area that would benefit from further research.

The term 'functional' can also have a double meaning. In the context of strabismus surgery, it refers to the potential for binocular vision to be restored post-operatively, but it would seem to imply that the patient was not 'functioning' before the surgery. The term is also used to describe vision loss related to psychological factors,[16] and amblyopia too has been referred to as 'functional'.[17,18]

Based on this body of evidence demonstrating the problems with the terms 'functional' and 'cosmetic', these expressions do seem to be outdated and invalid. But what would be a suitable alternative? The American Association for Pediatric Ophthalmology and Strabismus has suggested that the term 'cosmetic' be replaced with 'reconstructive'.[19] The additional benefit of this statement is to support the case for strabismus surgery in adults: coding it as a cosmetic procedure has implications for insurance coverage, as many insurers do not pay for purely cosmetic procedures. Another option would be to simply define the aim of the surgery as with or without binocular potential, although, as mentioned above, sometimes patients classified as having no binocular potential develop it spontaneously later on – for example, if they control to a microtropia after surgery.

Lastly, strabismus correction frequently requires more than one surgical procedure over time, but by labelling such procedures as a 'reoperation' or 'redo', does this imply that the result from the first procedure was inaccurate? In many cases further surgery is anticipated or planned, so should we take a leaf out of the refractive surgeon's book and refer to it as an enhancement procedure?

Microtropia

Microtropia is another term that would benefit from clarification. In the example above, of a reduction in angle following surgery, is it correct to use the term 'microtropia', or would 'residual heterotropia' be more appropriate? Even the intrinsic nature of microtropia is sometimes misunderstood.

The condition currently referred to as microtropia, in its guises of with/without identity and with/without a latent component, has undergone at least 10 different name changes in the last 50 years. These include esophoria with retinal slip, esophoria with fixation disparity, strabismus spurious, microtropia unilateralis anomalo-fusionalis, flick eso, microstrabismus, monofixational phoria, monofixation syndrome, small angle squint and microtropia.[20–22] Despite having the core elements of a $<10^{\Delta}$ heterotropia in association with unequal visual acuity, foveal suppression of the affected eye and abnormal binocular single vision,[23] the condition has a varied presentation, and clinicians were initially uncertain as to whether its incarnations were separate clinical identities or part of a single 'syndrome'[21] – which explains the array of names. This is best demonstrated by volume 26 of the *British Orthoptic Journal*, in which there are three articles each referring to microtropia differently.[20,24,25]

Pratt-Johnson[24] refers to the 'small-angle esotropia with binocular vision', describing all the associated core microtropia characteristics. But later he refers to 'suppression associated with anisometropic amblyopia', featuring amblyopia, a suppression scotoma and reduced depth perception. Although he states there is no manifest deviation, no mention is made of the state of fixation; thus this term could refer either to microtropia with identity, or to von Noorden's concept of subnormal binocular vision,[26] in which a suppression scotoma is present with central fixation in the absence of a manifest deviation and amblyopia.

Meanwhile Crone[25] describes a 'manifest ministrabismus with harmonious abnormal retinal correspondence' but refers to it as an esophoria – a definition born of his argument that any type of heterophoric fixation disparity was in fact a manifest strabismus. This confusion between microtropia and fixation disparity had already persisted for some time.[20] Fixation disparity is in fact a physiological process under binocular viewing conditions, involving the fusion of disparate retinal elements within Panum's fusional space to maintain binocular single vision.[27] While the disparity is generated by a slight under- or over-convergence of the visual axes, if it were considered to be a manifest strabismus then by definition the vast majority of the population would possess abnormal retinal correspondence. Lastly, there is Lang's article,[20] presenting the definitions as they are known today: microtropia with or without identity.

In America and Canada, the preferred term is monofixation syndrome[21] – perhaps a confusing name given that the central suppression scotoma described only occurs under binocular viewing conditions. However, the term references the lack of bifoveal fixation in the condition. This is still preferable to its previous name of monofixational phoria.[20] Though this referred to the superadded heterophoria present in some individuals with the condition, it implied no manifest deviation was present. Lang[28] argued that 'monofixation syndrome' was still an inappropriate term because it also encompassed the suppression scotoma that is sometimes associated with anisometropic amblyopia, fully accommodative esotropia or surgically corrected intermittent exotropia, all of which he defined as 'anomalies of binocular vision in the orthoposition'. However, it is now accepted that microtropia frequently occurs in these conditions, and may possibly even precede their onset;[23,29] thus the term was perhaps appropriately applied after all.

But although such confusions were an artefact of the still-evolving knowledge base on microtropia, even today the terms are still confused. A recent article on monovision[30] stated that 'monofixation syndrome does not require misalignment of the visual axes', demonstrating that there is still some ambiguity regarding the concept. 'Monofixation syndrome' has also been used interchangeably with the term 'subnormal binocular vision',[31] despite the two being separate conditions.

The different terms used both historically and geographically make it difficult to conduct a comprehensive literature search on the topic. In volume 26 of the *British Orthoptic Journal*, three articles dealing specifically with the microtropia phenomenon were titled 'Microstrabismus',[20] 'Small angle squints – theory and practice',[22] and 'From orthophoria to microtropia'.[25] Small-angle squint is a particularly vague term of choice, for other authors[32] have used this term to describe heterotropia of up to 20^{Δ}.

In addition, the definitions for the different descriptions of microtropia do not necessarily overlap. For example, in the monofixation syndrome it is argued that normal retinal correspondence (NRC) is present, maintained by an enlarged Panum's fusional area,[21] whereas Lang's definition of microtropia states that harmonious abnormal retinal correspondence (ARC) is present.[20] It has been argued that both types of retinal correspondence can be present in microtropia, with ARC centrally and NRC peripherally.[33] This has been contested,[34] but as Panum's fusional space is larger in the peripheral regions, the point-to-point correspondence present in central vision expands to point-to-area correspondence peripherally, making this concept possible even in the presence of central ARC. Retinal correspondence, too, is a term subject to some ambiguity, and is therefore discussed below.

Retinal correspondence

Retinal correspondence occurs when retinal elements of one eye correspond with those of the other eye and have a common visual direction. Thus in NRC the foveae have a common visual direction, with the nasal retinal points in one eye corresponding with temporal retinal points in the contralateral eye.[23] Bearing this in mind, would we regard patients with age-related macular degeneration (AMD) who have had macular translocation as having NRC, given that the actual retinal points in question have physically moved and no longer share a common visual direction with the other eye? This is a grey area as a small proportion of such patients can still achieve peripheral fusion and gross stereopsis,[35,36] yet it is uncertain whether there is utilization of a pseudofovea, thus not fulfilling the definition of ARC.[23] Though the presence of central scotomata in AMD means that the acquisition of a pseudofovea is possible, the frequency of diplopia following macular translocation would argue against this.[35,37]

The presence of such ambiguity would highlight that the definition of retinal correspondence is a little lacking. However, consider the possibility that retinal correspondence is a cortical process, as opposed to taking place at the retinal level.[38] Though this idea is based on animal studies and has yet to be confirmed in humans, such a concept would eliminate this terminology issue: the physical retinal points may have moved in macular translocation, but the cortical connections at the ocular dominance column level would not have changed. Therefore retinocortical correspondence would be a more appropriate term to use in these circumstances, and perhaps in general, as employed by Cleary *et al*.[34]

This discussion regarding retinal correspondence highlights how the development of knowledge through research may necessitate a change in terminology. However, there are other expressions used regularly within orthoptics where the term itself may be appropriate but the way in which it is used may encompass a number of meanings, and therefore further qualification of the term is required. One such example is the phrase 'the critical period'.

Critical periods

'*The* critical period' is a term used frequently, usually referring to the time period within which the visual acuity in the amblyopic eye improves in response to treatment. However, when considering the definition of a critical period in the context of the visual system,

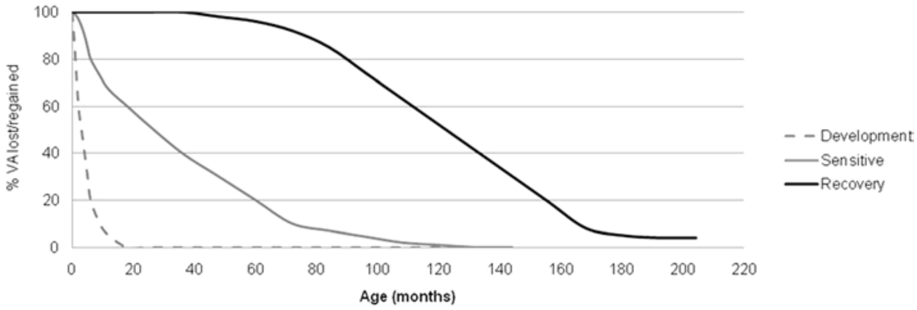

Fig. 1. An evidence-based schematic representation of the three critical periods affecting visual acuity. The *y*-axis represents the percentage of visual acuity (VA) that can be lost or gained during the three critical periods.

namely 'a period during which some property of the system can be changed',[39] use of the term in the singular is not appropriate.

Looking at visual acuity as an example, its development (development period), disruption by amblyogenic factors (sensitive period) and restoration by treatment (recovery period) are all changes to a particular property of the visual system.[40,41] They each have a critical period, with a similar start but differing end points. Fig. 1 is a schematic demonstrating the time span of the different critical periods for the visual system, based on evidence. Although the approximate start and end points for each period are known (with the exception of the recovery period – reports of recovery from amblyopia in adulthood would suggest this is yet to be accurately pinpointed), the incline of the slopes is not known and is therefore interpolated.

In the development and sensitive periods, visual acuity is lost at the point where an amblyogenic factor appears, with earlier onset causing a greater loss; but in the recovery period, visual acuity is gained at the point where amblyopia treatment is initiated, with earlier treatment allowing the recovery of more visual acuity. The recovery curve never reaches zero, indicating potential for recovery beyond the age span covered by the graph.

Although logically the sensitive and recovery periods start at the same time, amblyopia treatment has been shown to be successful in children older than the posited upper age limit for amblyopia development,[42] which has been estimated at 6–8 years;[39,43] thus the two critical periods do not end at the same time as shown in Fig. 1. Equally, Daw[40] argues that visual acuity reaches adult levels by age 3–5 years, which is less than the proposed upper age limit for amblyopia development,[39,43] so this would suggest that the development and sensitive periods do not end at the same time either. However, such acuities were obtained with uncrowded tests, whereas testing with crowded optotypes indicates that visual acuity continues to improve significantly after this age.[44,45] Grating acuity and contrast sensitivity are also shown to still be developing up until age 6 years,[46] so not all aspects of visual acuity development are necessarily complete at the ages stated by Daw.[40]

The starting points for the development and sensitive periods, however, are slightly more disparate. For example, astigmatism does not appear to induce meridional amblyopia in children younger than 6 months of age.[47] In the same fashion, neonates who experienced a brief period of strabismus or form deprivation within the first 6 weeks of life have been shown to develop normal vision and binocularity at follow-up assessment[48] – though this is based on preferential looking techniques and therefore mild amblyopia could go undetected. Based on such evidence it has been proposed that there is a period during visual acuity development within which the presence of an amblyogenic factor does not appear to cause gross amblyopia,[49,50] and therefore the peak for the sensitive period of visual acuity in Fig. 1 would be shifted slightly to the right. Although, of course, longer/more severe periods of deprivation are known to have an adverse effect on visual development in the affected eye,[40] and such clinical factors would also create some variability in the position of the sensitivity line in Fig. 1.

Having established the existence of three separate critical periods in the course of amblyopia onset and treatment, how would this affect clinical practice and research? Firstly, recommendations for amblyopia treatment should be consistent. The Cochrane Review on vision screening for amblyopia in childhood[51] states that amblyopia 'needs to be treated within the critical period for visual development in order for treatment to be effective', whereas the review on interventions for strabismic amblyopia[52] states that treatment is thought to be effective only during the critical period for amblyopia onset – but as argued above, many aspects of vision including visual acuity and contrast sensitivity are at adult levels by age 6/7 years and yet children still respond to treatment past this age.

It is also troublesome when research papers refer to the critical period in the singular with regard to the timing of amblyopia treatment, as this implies a single timeframe for all treatment methods, when in fact treatment can be tailored by targeting the critical periods. For example, removing congenital cataracts in the first 6 weeks of life, early in the recovery period, is shown to have the best visual outcome.[50] To add to the confusion, literature on treatment of congenital cataract also refers to the 'critical period' for cataract surgery – the time at which the surgery has the best visual acuity outcome.[49] In addition, evidence suggests that the sensitive and recovery periods for stereo-acuity differ significantly from those for visual acuity,[53] which reinforces the need to specify which critical period and which visual function is being referred to.

As can be seen, critical periods in visual development could have more of an impact on treatment decisions than perhaps were previously considered. Further research into critical periods for other aspects of vision, such as the development of binocularity, could potentially influence orthoptic treatment decisions for the future – for example knowing when to intervene with glasses in cases of infants with refractive error.

Conclusion

This article has highlighted a few areas of orthoptics where terminology used has either a double meaning or could generate misconceptions when interpreted incorrectly. Some of the confusion originates from changes in terminology over time, as knowledge and understanding has increased. In other cases the evolution of new treatment methods has highlighted the grey areas in long-accepted names and definitions. Regardless of how such ambiguities arise, many issues can be resolved by simply updating the terminology, as has been done recently with the renaming of binocular visual acuity to controlled binocular acuity. However, when conducting an extensive review of the literature, the evolution of medical terminology must be borne in mind, to ensure accurate evaluation.

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