Grapheme-colour synaesthesia and the orthoptist

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Abstract

Aim: To give an overview of the condition known as grapheme-colour synaesthesia, and examine whether there is a need for orthoptists to be aware of its existence and potential impact when it coexists with other conditions, such as specific learning difficulty and Meares-Irlen syndrome.

Methods: A literature-based review was performed. Relevant material was identified using the University of Liverpool library catalogue, Google, PubMed and Web of Knowledge. The focus was on relevant research published within the last 15 years.

Results: Evaluation of the literature shows that grapheme-colour synaesthesia is thought to arise from increased structural connectivity between brain areas involved in the processing of colour and visual word forms. The grapheme-colour linkages are very specific and long-standing, possibly congenital, and the condition is probably genetic. It is more common than previously thought, and could potentially coexist with visual perceptual difficulties such as dyslexia and Meares-Irlen syndrome. However, due to a lack of awareness of the condition it is rarely diagnosed. As grapheme-colour synaesthesia appears to have an impact on reading and mathematical ability, it is relevant to the orthoptist’s extended role in assessing children with these conditions.

Conclusion: Grapheme-colour synaesthesia is more common than previously thought, and although more research is needed to establish the true impact of grapheme-colour synaesthesia on other visual conditions, the orthoptist may wish to consider including the condition in the list of differential diagnoses in cases of specific learning difficulty and Meares-Irlen syndrome.

Key words: Grapheme-colour synaesthesia, Functional brain imaging, Meares-Irlen syndrome, Specific learning difficulty

Introduction

People commonly speak of ‘blue Mondays’ or ‘tasting victory’, but for some individuals this goes beyond the realms of literary metaphor and into sensory reality. They taste chocolate whenever the word ‘oysters’ is mentioned, or see the colour red when viewing the letter A. These people have a neuropsychological condition known as synaesthesia. It is unusual in being associated with an extra perception rather than the absence of one, as in achromatopsia or agnosia.

When perceiving a stimulus in one sensory modality, those with synaesthesia experience an extra, simultaneous perception within that modality – or from another sensory modality entirely. This occurs in the absence of input to that extra modality. For example in the modality of vision, an additional colour is perceived when viewing a grapheme (alphanumeric character) that is not coloured (i.e. black). The first stimulus perceived is known as the inducer of the synaesthetic sensation; the extra perception is known as the concurrent.

Synaesthesia has a profound impact upon perception, but is often undiagnosed because it is rarely associated with other neurological problems. The most common concurrent is coloured visual phenomena, or photisms, making it of relevance to eye care professionals such as orthoptists. Synaesthetes must go about their daily lives unable to ‘switch off’ their unusual perception due to its automatic nature, and are often unaware their experiences have a clinical identity until later in life. Given that other defects of visual perception such as colour blindness and Meares-Irlen syndrome are assessed within orthoptic practice, it could be argued that orthoptists should also be aware of the signs and symptoms of synaesthesia.

This literature review aims to describe and promote awareness of the condition. A literature search was done on PubMed and Web of Knowledge databases using the search terms ‘synaesthesia’, ‘synaesthete’, ‘synaesthetic’, and the American spellings of these words. Some non-peer-reviewed papers were identified using the Google search engine, and relevant books obtained by searching the University of Liverpool library catalogue. The review focuses primarily on grapheme-colour synaesthesia, thus papers focusing on other types were not included in the review unless they had specific relevance. Only papers published in the last 15 years were included in the study.

Features of synaesthesia

Synaesthesia is a complex condition, with over 53 known inducer-concurrent variants. Some common pairs are shown in Table 1. Despite this, the different types of synaesthesia share...
common features. This review focuses on grapheme-colour synaesthesia, in which photisms are induced by combinations of letters, numbers and calendar days/months. It is the most heavily studied type and is relevant to orthoptic practice, given our heavy use of letters as fixation targets and in visual acuity testing. More orthoptists are also becoming involved in the assessment of those with specific learning difficulties, including problems with reading. This type of synaesthesia can also affect reading and mathematical ability.

In grapheme-colour synaesthesia, as in most other types, the concurrent is automatic, arising involuntarily. The inducer-concurrent associations remain highly consistent over time, and the majority of synaesthetes report having their condition ‘for as long as they can remember’. This has been confirmed via sibling reports as far back as the age of 4 years, though it is difficult to prove such instances at younger ages due to limitations in the child’s ability to report their synaesthesia. This indicates synaesthesia is a lifelong condition, perhaps congenital. The similarities between the different variants would suggest they share a common aetiology.

The point prevalence of grapheme-colour synaesthesia amongst adults in the United Kingdom is estimated to be approximately 2% – higher than previously reported. It appears less common in children, with a prevalence of about 1.3% in 6- to 7-year-olds, although this may be an underestimate. There is some debate over whether the condition affects females more than males. A skewed gender ratio towards females has been found in many epidemiological studies. However, many of these studies used newspapers to recruit their participants – a method that tends to produce a female:male gender bias. One study not using this recruitment method found no significant gender bias, but as the number of synaesthetes diagnosed was small (n = 35) the sample lacked statistical power.

Synaesthesia tends to run in families. Forty-three per cent of children born to parents with the condition are synaesthetes themselves, indicating a genetic component. However, a pair of male monozygotic twins were discovered of whom only one twin had synaesthesia, showing that environmental influences also contribute to the formation of grapheme-colour linkages. There have been documented incidents of such pairings being acquired from coloured alphabet fridge magnets and jigsaws, and of colours migrating to orthographically similar letters in Greek/Cyrillic languages acquired in later life. In light of this it has been suggested that a ‘susceptibility gene’ is inherited, which predisposes an individual to synaesthesia.

This is plausible given that some common grapheme-colour linkages exhibited by synaesthetes (e.g. A = red) have also been found in non-synaesthetes. Perhaps possession of the susceptibility gene causes such linkages to be consciously perceived. The genetic studies required to confirm this are still in progress.

The neurophysiology of grapheme-colour synaesthesia

Given that grapheme-colour linkages also happen in non-synaesthetes, what causes the conscious perception of these colours in those who do have synaesthesia? Features of the synaesthetic experience suggest the concurrent perception is generated through cortical channels. For example, synaesthetes have difficulty naming the actual (surface) colour of a letter when the colour of the associated photism activates the same colour-opponent retinal channel (e.g. when viewing a green letter that produces a red photism). Several cortical models have been generated to explain how the synaesthetic perception comes about. The core model is cross-activation, hypothesising that in synaesthetes there is increased cortical connectivity between brain regions involved in processing colour and grapheme form. Stimulation of the colour area leads to activation in the grapheme area via these connections, generating the concurrent photism alongside the perception of the inducing grapheme. Extra connections were suggested at two particular sites in the brain (Fig. 1). In the fusiform gyrus, increased connectivity was proposed between extrastriate colour region V4 and the adjacent Visual Word Form Area (VWFA). At the second site in the parieto-temporal region, connections were thought to be present between the angular gyrus (believed to process abstract letter and number concepts), and the nearby superior temporal gyrus (the higher processing centre for colour). Psychophysical experiments provide strong evidence for the involvement of these regions, showing that synaesthetic colours are modulated by factors known to be processed within these brain areas.
In order to establish whether these sites are actually involved in the generation of the synaesthetic experience, many imaging studies have been carried out. However, due to errors in classifying the experimental groups many of the findings conflict, and should be interpreted with caution. These errors originated from attempts to classify grapheme-colour synaesthetes into subtypes; for it was clear they are not a homogeneous group. Two classification systems were developed, the first being based on how synaesthetes responded to different types of inducer. ‘Lower’ synaesthetes had photisms induced by the visual appearance of the grapheme, due to cross-activation between V4 and the VWFA. ‘Higher’ synaesthetes had photisms induced by the grapheme’s meaning or numerosity (number concept), due to cross-activation at a later processing level within the angular and superior temporal gyri.\textsuperscript{31}

The second classification was based on how the photism was perceived. ‘Projector’ synaesthetes saw the photism as being projected on top of the grapheme, while ‘associator’ synaesthetes felt they experienced an associated impression of colour in the mind’s eye. These two classifications were thought to overlap: projector synaesthetes were also lower synaesthetes, associator synaesthetes were also higher synaesthetes.\textsuperscript{34} Thus, experimental samples were separated based on this classification, as it fitted in with the characteristics of synaesthetes being studied.

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Subsequently, it was discovered that one could be a higher synaesthete but also have projected photisms\textsuperscript{35} – the two classification systems were in fact separate. By this point, however, most studies had classified groups based on projector/associator type, as this was easier to determine. Therefore higher and lower synaesthetes, who have a different neurological site for cross-activation, were probably mixed throughout the two groups. This would reduce the chance of a significant activation site being found during signal analysis.

Despite this, careful examination of the findings of imaging studies does show robust V4 activation in many synaesthetes in the absence of grapheme surface colour,\textsuperscript{36–38} though their status as lower synaesthetes is unconfirmed in most cases. In another study,\textsuperscript{39} a confirmed higher synaesthete was found to have activation of the angular gyrus region during the mental addition of dice dots – which was not present in the control subject.

Diffusion tensor imaging, a technique specifically designed to identify patterns of connectivity between brain regions, also provides support for the cross-activation model. This imaging method has revealed that the brains of grapheme-colour synaesthetes do possess increased structural connectivity, relative to non-synaesthetes, in the regions described above.\textsuperscript{40} However, as Kadosh and Walsh\textsuperscript{41} have pointed out, it is unclear whether these extra connections are the cause or the effect of synaesthesia. Ramachandran and Hubbard\textsuperscript{29} have suggested that such connectivity is present from birth, or at least early infancy – though this hypothesis would be difficult to test. Further research needs to be done on synaesthetes who have been classified appropriately to gain more information on the neurological foundations of the condition.

**Synaesthesia in the orthoptic clinic**

It has been argued that synaesthesia does not have a negative impact on daily life.\textsuperscript{42} Further, because of its perceptual nature it is ‘invisible’, unlike strabismus. It also appears to be untreatable. However, might its diagnosis be of relevance in orthoptic practice in the same way as colour-blindness, which also cannot be diagnosed formally? Grapheme-colour synaesthesia was relatively unstudied. Rich et al.\textsuperscript{19} reported that 35% of their 136-strong sample confused words with similar synaesthetic colours, but they did not screen their sample formally for dyslexia, and excluded four participants whose questionnaire responses were indicative of the condition. In one non-peer-reviewed report, Stephan\textsuperscript{45} examined a cohort of 92 dyslexic students and 87 non-dyslexic students, failing to find a significant difference in synaesthesia incidence between the two groups. However, males significantly outnumbered females (103:76). Though increased incidence of synaesthesia in females has not been conclusively proved, there is the possibility that this gender imbalance may have affected the results.

In contrast, another non-peer-reviewed report\textsuperscript{46} suggested a significant incidence of grapheme-colour and number-form synaesthesia in adults referred for SLD assessment (1:24, out of 378 referrals). Synaesthesia was most common in those with ADD/ADHD, though a small percentage of dyslexics and dyspraxics were also affected. One critical fault with this report was the failure to verify the synaesthetic experiences of the patients using a recognised test, such as the Test of Genuineness.\textsuperscript{11} A larger-scale study of synaesthesia in those with SLDs is needed, with careful testing to confirm the authenticity of reported synaesthetic experiences, before drawing firm conclusions about the need to identify grapheme-colour synaesthesia in this patient group.

**Synaesthesia and Meares-Irlen syndrome**

Meares-Irlen syndrome is another condition associated with reading difficulties that can occur independently of dyslexia.\textsuperscript{47} Orthoptists are expected to carry out Meares-Irlen screening as part of the assessment of children referred for SLDs.\textsuperscript{45} One common symptom associated with this syndrome is ‘illusions of colour’, defined as auras around letters/words or blobs of colour on the page that obscure or distract from the word being read.\textsuperscript{48} Some synaesthetes do experience photisms that blot out print,\textsuperscript{49} and sometimes the colours can interfere with reading.

As some of the symptoms of grapheme-colour synaesthesia and Meares-Irlen syndrome overlap, the differential diagnosis could be important. Grapheme-colour synaesthetes are unlikely to benefit from using coloured overlays if their reading difficulties originate purely from the distracting colours. As the form and meaning of the grapheme can still be appreciated through an overlay, the photism will still be generated.

Grapheme-colour synaesthesia could be potentially identified during the case history by asking how long the colour symptoms have been present and whether they...
vary distinctly between letters/words. Making the patient aware of the origins of their photisms will prevent raised hopes at the prospect of an overal curing their synaesthesia.

Conclusion
Grapheme-colour synaesthesia appears to be a congenital condition, with a strong familial component possibly caused by inherited susceptibility genes. The mechanism by which the concurrent photism is generated is not fully established due to the impact of individual differences on the results of imaging studies. However, synaesthetes possess definite alterations in the level of structural connectivity between brain regions involved in colour- and word-processing. This suggests certain brain regions could be ‘cross-wired’ in those with synaesthesia, producing the concurrent colour perception. Further research needs to be done on subjects that have been classified correctly in order to gather more information on the neurological foundations of the condition.

Further research is also required into the incidence of synaesthesia in patients with other conditions presenting to the orthoptic clinic. There are no chronicled instances of synaesthesia coexisting with ophthalmological or orthoptic problems, but as many clinicians are unaware of the condition, such a combination could be possible, yet undiagnosed. Equally, synaesthetes are often unaware that their condition has a clinical identity; therefore, they may not even volunteer information about their synaesthesia unless specifically asked (e.g. during Meares-Irlen screening).

The impact of grapheme-colour synaesthesia on other visual conditions has been relatively unstudied. Some articles point out difficulties in reading and mathematics because of the distracting photisms. If subsequent research supports the possibility that such distractions contribute to reading difficulties, including grapheme-colour synaesthesia in the list of conditions to identify during visual assessment of a poor reader could be beneficial. For example, some of the symptoms of grapheme-colour synaesthesia and Meares-Irlen syndrome overlap. Grapheme-colour synaesthetes are unlikely to benefit from colour-coded overlays unless they have Meares-Irlen syndrome as well, so the two conditions should be differentially diagnosed.

Though it appears to be untreatable, raising awareness of grapheme-colour synaesthesia is important. Many synaesthetes are unaware until adulthood that their experiences have a name, and feel somewhat alone in their perceptions. A simple explanation of their synaesthesia’s origins, and reassurance that others see as they do, could help these individuals feel less isolated and more able to talk about what they see. Alerting educators to the presence of the condition can also help ensure those with difficulties with reading or mathematics receive appropriate assistance for their educational needs.

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