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The importance of contrast sensitivity testing in children

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Abstract

Aim: To discuss the information contrast sensitivity (CS) testing can provide over visual acuity testing, and review the literature relating to CS deficits in children to determine whether there is an optimum test available.

Methods: A literature search of databases available through the University of Liverpool library was performed. All searches related to the importance of CS in children, paediatric conditions affecting CS and current clinical tests available for the assessment of CS.

Results: Many paediatric conditions exist where CS is defective, often despite 'normal' visual acuity (VA): for example, optic pathway gliomas, myopia and primary congenital glaucoma. The finding of a loss of CS has been found to be more prominent and disturbing to an individual than a loss of VA, emphasising the importance of CS assessments in children. Therefore, the clinical assessment of CS in children is valuable in terms of strategies to support the child and establishing the individual's functional level of vision. Unfortunately, current paediatric clinical tests of CS have failed to demonstrate the same standards and repeatability and reliability as adult tests for the assessment of CS.

Conclusions: The range of functional deficits that accompany paediatric ocular disease require more assessments for full evaluation of visual function than standard VA tests. However, reliable assessments for paediatric CS are limited. A new paediatric CS test may be of clinical value.

Key words: Contrast sensitivity, Paediatric, Visual impairment

Introduction

The ability to test visual acuity (VA) is of great clinical value, and affects diagnostic and management decisions. However, measuring an individual's ability to detect minor changes in luminance is also of importance and is often referred to as contrast sensitivity (CS). Today in

clinical practice often only VA is assessed, by means of an optotype chart in Snellen or logMAR format in high contrast. Although VA measurements are essential in determining whether a patient is suffering from a visual disturbance, patients can often experience visual disturbances in the presence of normal VA.

The aims of this review are to discuss the information CS testing can provide over visual acuity testing, and to review the literature relating to CS deficits in children to determine whether there is an optimum test available. To achieve these aims we will evaluate available evidence relating to CS testing in children. The following points will be addressed:

- the limitations of the assessment of VA in isolation;
- the role of the assessment of CS;
- normal development and paediatric conditions affecting CS;
- clinical assessments of CS.

The search of the literature utilised the following databases available through the University of Liverpool library: Medline, Scopus, Web of knowledge, and the Cochrane library. Key words searched for included: paediatric, visual impairment, contrast sensitivity, clinical tests, functional vision, and visual acuity. Boolean logic enabled an effective search strategy to be employed.¹

Limitations of visual acuity testing

VA is a measure of spatial vision which is defined as the finest element that can be resolved at a fixed distance^{2–4} and is the most frequently used indicator of spatial vision in both clinical studies and clinical practice.³ The measurement of VA in children is often utilised to detect refractive errors, amblyopia and other ocular anomalies as well as to monitor improvement of vision with ongoing treatments.⁴ While there are a wide range of VA tests available, ensuring that it can be assessed in all patients irrespective of cognitive ability, VA only measures an individual's recognition of the smallest high-contrast pattern visible.^{5,6} Although the measure of VA provides important clinical information, the ability to measure CS in a clinical environment is also of importance in all ages and is a highly recommended as part of a functional visual assessment.^{2–3,7} Studies have demonstrated high levels of correlation between VA and other spatial vision measures such as CS, but there is

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considerable variation, with the correlation being as low as 0.27 in low vision patients, and up to 0.79 in patients with normal VA.^{3,8,9} The variability in the correlation emphasises the limitations of VA results. Such a large variability is in part attributable to differences in the population characteristics between studies, but it is also known that factors such as the method of assessment of CS, the luminance and data collection differences all contribute to variation in both VA and CS.^{9,10} Nonetheless, there is also a possibility that participant(s) may have been suffering from a condition in which a normal level of VA is demonstrable despite a sensory visual impairment causing a reduced CS threshold.

Functional importance of the assessment of contrast sensitivity

CS is a measure of the ability to detect slight changes in luminance; it can provide an evaluation of the detection of objects of varying spatial frequencies and/or variable contrast.¹¹ The contrast threshold is the reciprocal of CS, and defined as the level of contrast at which a light and dark pattern is first discriminated. The measurement of contrast thresholds for sine-wave gratings across a range of spatial frequencies determines an individual's contrast sensitivity function (CSF).¹¹ While the assessment of the complete CSF provides a detailed evaluation of an individual's CS, it is the contrast threshold at one specific spatial frequency which is more commonly assessed during a clinical assessment, primarily due to time constraints. For example, using the Pelli-Robson test assesses individuals contrast thresholds at a specific spatial frequency at the recommended testing distance of 1 metre.⁹

The assessment of CS increases our understanding of how individuals perceive the world, as it provides a more complete and detailed assessment of visual function related to a 'real visual environment'.¹² The measurement of CS has been shown to contribute additional information to that provided by VA alone, becoming the most comprehensive single means of evaluating the visual system's response to pattern information, particularly in patients with visual impairments.^{2,3,5,13,14} CS plays an important role in mobility performance, the ability to recognise faces and the ability to undertake daily living tasks in individuals with visual impairments.⁹ The ability of an adult patient with a severe vision impairment to resolve details in daily living, such as reading, is assessed by measures of VA, whereas the ability of a person to detect objects relative to their background, for example when walking down the stairs, is measured by CS.¹⁵ There was no evidence found in the literature suggesting that this does not apply to a child's visual system. Evaluating remaining functional vision such as CS and VA in both children and adults with severe vision loss is key when determining rehabilitative strategies and quality of life. Overall, studies have shown that the assessment of other spatial vision measures contributes additional information to that provided by acuity alone.^{2-3,5,12-14}

Paediatric disease and conditions with defective contrast sensitivity

Numerous conditions can occur in children which ultimately affect the individual's functional vision (a wide range of examples are included in Table 1). Assessing CS and finding a defective contrast threshold in many of these patients could provide an explanation as to why children may be demonstrating behaviour compatible with a sensory visual impairment despite normal VA.¹⁶ Assessing CS in a clinical setting may also prevent patients with a visual function disorder that is not apparent on standard VA testing from being considered as having 'normal vision' and being discharged.

Table 1 clearly demonstrates that children with disabilities are likely to have visual disorders with an increased risk of reduced visual function, including reduced CS.^{6,13,17-28} The correlation between the individual's VA and CS can be variable, and therefore a quick, reliable and repeatable assessment of both CS and VA is desirable, particularly for children with or without disabilities.^{3,8,9} Good *et al.* and Fazzi *et al.* demonstrated that over 60% of children with cerebral palsy have cerebral visual impairment, resulting in a substantial loss of CS (see Table 1).^{25,29} In addition, children with Down syndrome are at risk of a variety of ocular anomalies such as cataracts, high refractive errors, strabismus and congenital glaucoma, which can all have an adverse effect on CS.¹⁴ In these children it is vital to be able to establish their functional level of vision, i.e. their CS threshold, to acknowledge what treatment or support may be required to ensure the child has the best possible outcome.

Studies have demonstrated a reduction of CS in patients with developmental delay (10.5%).^{30,31} These children are also more likely to suffer from strabismus and refractive errors such as hyperopia, astigmatism and anisometropia, all which can affect an individual's CS threshold.^{30,31} Therefore, a reduced CS threshold in patients with Down syndrome or developmental delay may not be directly attributable to the primary disorder, but possibly due to the associated ocular abnormalities that are prominent in these patients. Although this has been supported in the literature, it is not apparent that the individual associated ocular abnormalities have been specifically examined, but rather the effect of the overall disability on CS.^{30,31}

The growing population of children born prematurely is also known to have a high prevalence of ophthalmological and neuro-developmental disorders in which the retina or posterior visual pathways may be disturbed, thus affecting CS.^{32,33} Previous studies have shown prematurely born children had statistically significant lower CS at all spatial frequencies when compared with full-term children. Although results were of statistical significance, mean differences in logarithmic CS (Log CS) were only slight: for example, a mean difference of 0.03 Log CS (1.5 cpd) and 0.09 Log CS (3 cpd) were shown.³² One may suggest that the reduced CS in these children is not directly attributable to the prematurity but the associated ophthalmological and neuro-developmental disorders that occur as a result of the prematurity. In support of this, Larsson *et al.* found premature children

Table 1. Paediatric conditions and their effect on VA and CS^{6,17–28}

| Condition | n | Effect on VA | Effect on CS |
|---|---|---|---|
| Myopia ¹⁷ | 105 myopic eyes 28 control eyes | Corrected acuities 6/6 Snellen or better | Loss at higher spatial frequencies for moderate myopes, all spatial frequencies for high myopes |
| Unilateral congenital cataract ¹⁸ | 15 15 controls | Grating acuity reduced on average by 0.6 log units | 100% had losses in spatial CS in deprived eye which increased with increasing spatial frequency |
| Primary congenital glaucoma (PCG) ¹⁹ | 10 9 controls | Significant pattern electroretinogram (PERG) deficits in PCG patients. PERG almost reached level of controls at high temporal frequency | In both groups, mean CS peaked at 6 cpd, but significantly lower in patients with PCG at all spatial frequencies |
| Retinopathy of prematurity (ROP) ²⁰ | 291, severe threshold ROP in 1/both eyes + cryotherapy 10 low birth weight controls (–ROP) | 63% showed VA and CS correlated well post-treatment. 30.4% had better CS than VA | 70.7% ‘normal’ CS compared with 74.4% for the controls |
| Retinitis pigmentosa (RP) ²¹ | 30 Mean age 37.8 ± 17.6 years | VA varies in RP patients due to the stages of disease. CS and VA significantly correlated when VA >20/50 | CS values 15–170 at 12 cpd in patients with VA better than 20/25. Reduced CS found at both high and low spatial frequencies |
| Optic neuritis ²² | 30 | Statistically significant difference between VA of the affected and unaffected eyes | CS showed the most significant difference between the affected and unaffected eyes |
| Septo-optic dysplasia (SOD) ²³ | 17 | VA significantly decreased in all children, 7 had perception of light only | 10 had CS measured, normal in 1 |
| Optic pathway glioma (OPG) ²⁴ | 16 patients 16 controls | Corrected VA of 6/9 or better | Significant reduction in CS in OPG patients who had normal VA |
| Cerebral visual impairment (CVI) ²⁵ | 34 months to 5 years 16 controls | Grating acuity thresholds decreased by a factor of 2.3 compared with controls (median acuity: 0.49 logMAR) | CS decreased by a factor of 30 compared with controls |
| Infantile nystagmus syndrome (INS) ^{26,27} | 84 infants | Mildly reduced VA early on, gradual maturation with age paralleling a normal VA curve | Significantly reduced at all spatial frequencies |
| Amblyopia ^{6,28} | 22 aged 3–6 years | VA varied and depended on severity of amblyopia | Reduced at high spatial frequencies and other areas of CSF. Non-amblyopic eye showed reduced CS threshold despite a ‘normal’ VA |

without associated neurological conditions still demonstrated reduced CS compared with full-term children.³² It is the loss of CS that can be more disturbing, and the CS that is more severely reduced, as premature children can demonstrate normal levels of VA with defective CS.^{2,15,32} Although children with poor VA were excluded from Larsson *et al.* study, premature children had significantly lower CS in the mid to low range of spatial frequencies, highlighting visual problems despite normal VA.³²

CS measurements can monitor or influence treatment plans in conditions such as retinitis pigmentosa or amblyopia. In retinitis pigmentosa the assessment of CS can reveal subtle improvements in patients undergoing new drug treatment and therefore necessitating detailed evaluation, as high-contrast figures are insufficient to evaluate mild damage to foveal cones.^{21,34} Additionally, CS measurements can contribute to the treatment plans of other conditions such as amblyopia. Both CS and VA are reduced in the amblyopic eye in amblyopia patients and should both be measured to monitor functional vision in the amblyopic eye during occlusion treatment. However, CS in the non-amblyopic eye in these patients should also be monitored as studies have shown that despite normal VA the eye which is considered ‘normal’ does actually demonstrate a defective CS threshold.²⁸

In addition to influencing management, CS may aid in the diagnosis of some conditions. During the early stages of optic neuritis VA can remain at a normal level whilst low- and mid-range spatial frequencies of CS are

reduced.^{22,35} CS measures often provide the initial indications of visual compromise, even prior to changes in the fundus appearance.¹⁷ In young children infectious and post-infectious causes of optic nerve impairment should be considered as alternatives to optic neuritis. Therefore, early recognition of symptoms is important, which could be detected through CS testing.³⁵ Additionally, the assessment of CS in some cases can provide a much quicker diagnosis than currently used tests. For example, the current standard assessments to accurately diagnose optic pathway gliomas (OPGs) are often lengthy and require young children to be co-operative, or are extremely expensive and involve additional risks (e.g. neuro-imaging). Standard VA assessments are not reliable enough as they can fail to detect optic nerve compression.^{22,35} However, sweep visual evoked potentials (sVEPs) have been suggested as an effective screening test to assess an individual’s functional vision, using grating and CS stimuli.²⁴ These findings suggest that a repeatable, reliable, quicker and more cost-effective method of assessing CS in children would be valuable to use in a clinical setting to aid in the diagnosis of OPGs.

Visual function, including CS, in children with neurological impairments can often be difficult to quantify. Studies have proven that a visual skills inventory completed by parents or a structured history taken by a clinician is often useful in determining whether a child is likely to have reduced visual function.³⁶ Although the assessment of CS is desirable, it is not always realistic or

Table 2. Normative CS thresholds assessed with the Pelli-Robson chart tested at 1 m

| Author | Age at test (years) | Average result (Log CS) |
|---------------------------------------|---------------------|--|
| Haragadon <i>et al.</i> ⁴⁴ | 6 | Mean \pm SD, RE: 1.63 \pm 0.12, LE: 1.65 \pm 0.06 |
| O'Connor <i>et al.</i> ³³ | 11 | Median (IQR), RE: 1.65 (1.575, 1.65), LE: 1.65 (1.60, 1.65), BE: 1.90 (1.80, 1.95) |
| Leat and Wegmann ¹³ | 6 to <8 | Mean (95% CI based on SD), monocular results: 1.68 (1.57) |
| Leat and Wegmann ¹³ | 23 to 37 | Monocular results: 1.79 (1.59) |
| Mantjaryvi and Laitinen ⁴² | 6 to 9 | Mean \pm SD, RE: 1.72 \pm 0.08, LE: 1.76 \pm 0.07, BE: 1.91 \pm 0.07 |
| Mantjaryvi and Laitinen ⁴² | 10 to 19 | Mean \pm SD, RE: 1.73 \pm 0.08, LE: 1.76 \pm 0.07, BE: 1.91 \pm 0.07 |

RE, right eye; LE, left eye; BE, both eyes.

possible to routinely assess it in all patients. Using the visual skills inventory has been proven to be a good way of identifying potential CS deficits necessitating further visual function assessments such as CS.³⁶

Development of contrast sensitivity

In visually normal children CS develops rapidly over the first 6 months of life. There is rapid neural development of the visual system as a whole over the first 3 months of life with CS reaching adult levels for at least low spatial frequencies by 3 months old.^{37,38} Norcia *et al.* stated that between 4 and 9 weeks CS improves significantly at all spatial frequencies. However, after 9 weeks of age maximum CS thresholds increase by a smaller amount while sensitivity at higher spatial frequencies continues to increase dramatically.³⁹

The time during which neural plasticity makes the visual system vulnerable to abnormal experiences and interruptions in normal visual development is defined as the sensitive period.³⁹ For VA, the sensitive period extends to approximately 8 years of age, but the sensitive period for CS has not been clearly defined. The CSF is known to continue to develop beyond the age of 8 years, not reaching a maximum until approximately 20 years of age, suggesting that the sensitive period for CS may extend beyond the age of 8 years.^{11,31,38-41} Leat and Wegmann supported this evidence by demonstrating a statistically significant difference in CS thresholds between two age groups: 6 to <8 years and 23 to 37 years ($p < 0.001$) (data in Table 2).¹³ Therefore, it is important to compare CS values with appropriate age-related norms, due to the variability of CS with age (see Table 2). For example, mean CS thresholds in children aged between 6 and 12 years old have been found with the Pelli-Robson CS chart to be between 1.63/1.65 Log CS (SD: 0.12/0.06 Log CS) and 1.72/1.76 Log CS (SD: 0.08/0.07 Log CS) for the right and left eye, respectively.^{13,42} The significant variation between mean thresholds may be due to the development of CS thresholds/function, age variations, the scoring methods used and the luminance used during assessment.¹¹

Mean CS thresholds for adults between the ages of 20 and 40 years have shown to be 1.84 \pm 0.12 Log CS. Over the age of 60 years mean CS thresholds decrease significantly to around 1.72 \pm 0.08 Log CS.⁴² However,

regardless of the variation in scores across the different age categories, studies have stated a Pelli-Robson score of at least 1.05 Log CS is required for fluent reading.^{13,43} Indicating a score below 1.05 Log CS with the Pelli-Robson chart could identify a visual impairment which may have an impact on an individual's daily routine.

Contrast sensitivity tests

Clinical assessment of CS involves the use of either sine/square-wave gratings or optotype charts. The Pelli-Robson CS letter chart is one of the most readily available methods for assessing CS thresholds in adult patients (Fig. 1). The Pelli-Robson is an optotype CS chart of a fixed spatial frequency and reducing contrast. CS charts using optotypes adopt a specific psychophysical method of identification. In the case of the Pelli-Robson, it has a 26-alternative forced-choice design (the subject is 'forced' to give a response at threshold rather than just indicating the letter cannot be seen), ensuring a more accurate threshold is reached. The test uses 10 Sloan letters which are comparable to the Landolt C. Each letter has a recognition score to ensure each triplet is of the same difficulty in terms of recognition. Forced-choice procedures have been shown to yield more reliable results as they minimise methodological biases more than non-forced-choice testing designs, which can be influenced by patient criterion differences.⁹ The Pelli-Robson chart has also been shown to have good test-retest reliability. Although research has shown there was a slight increase in mean CS scores on retest of 0.025 Log CS, this was not considered clinically significant. In addition, the coefficient of repeatability of all the test-retest CS scores was 0.15 Log CS.⁹

While the Pelli-Robson chart is a suitable test for adults, it is not suitable for use in young children, due to the stimuli consisting of letters and the increased number of contrast levels on the chart. At present the most readily available clinical tests for assessing CS in children are the LEA low contrast symbols fixed CS charts, the LEA grating test and the Hiding Heidi CS test.^{13,45} Additional tests include the Vistech system and the Cambridge CS test.^{46,47} Although each test has its individual clinical value, problems have been identified such as the overestimation of the CS threshold in children. This results in the levels of CS in the tests not being sufficiently low to measure a true threshold, and thus of limited use for children with normal or near normal VA.^{13,47} For example, although the Hiding Heidi test has demonstrated a positive correlation with the Pelli-Robson chart, Hiding Heidi has been shown on average to measure Log CS 0.23 units higher than the Pelli-Robson chart and can also be subjective, resulting in bias.^{13,48} Although it has not directly been compared with the Pelli-Robson chart, the Cambridge CS test has been shown to have low test-retest reliability; reliability was shown to improve if the same examiner assessed the individual, but not by a significant value.^{46,47} There are currently no paediatric optotype tests of CS which are directly comparable with the Pelli-Robson CS test.¹³



Fig. 1. Pelli-Robson chart.⁴⁹

Optimum paediatric contrast sensitivity test design

Arden stated that the failure to assess or measure CS is a defect in our ability to diagnose and monitor its progress.² Therefore, the development of a new paediatric CS test which is quick, reliable and as repeatable as the adult Pelli-Robson CS test is desirable. A picture optotype test of a fixed spatial frequency and reducing contrast, with 16 levels comparable to the Pelli-Robson chart is preferable. This will ensure there is a degree of standardisation between adult and paediatric testing.⁹ Having 16 contrast levels will minimise the risk of a ceiling effect, therefore increasing the reliability. The Pelli-Robson chart has shown good test reliability; therefore a new paediatric test of CS would need to aim for the same standards.⁹ Each contrast level on the paediatric test would need to consist of a triplet of optotypes which equally have a good recognition level with children, so the child can give the answer verbally or through matching to avoid bias. The new paediatric test would be able to assess CS at either a 3 m or a 1 m distance and, due to the test being the same format as the Pelli-Robson chart, individual optotypes would be able to be scored at a value of 0.05 Log CS.^{42,50} Patients would be required to read the symbols, starting with those of the highest contrast, until two or three optotypes in a triplet were read incorrectly.

Conclusion

This review highlights the value of testing CS in certain situations and conditions affecting a paediatric popula-

tion, such as prematurity, children with Down syndrome and developmental delay, and children with ocular conditions such as primary congenital glaucoma, optic pathway gliomas and optic neuritis. The range of functional deficits that accompanies paediatric ocular disease requires assessments for full evaluation of visual function in addition to standard VA tests. However, reliable assessments for currently existing CS tests suitable for paediatric use are limited. A new paediatric CS test may be of clinical value. The test should be able to provide an accurate measure of functional vision in children with and without a visual impairment. Such a test will allow clinicians to establish an individual's functional level of vision and aid with rehabilitative strategies if required.

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