Low-Contrast Acuity Measurement: Does It Add Value in the Visual Assessment of Down Syndrome and Cerebral Palsy Populations?

Julie-Anne Little,1,2 Sara McCullough,1 Julie McClelland,1 A. Jonathan Jackson,2,5 and Kathryn J. Saunders1

PURPOSE. Children with Down syndrome (DS) and cerebral palsy (CP) often have reduced visual acuity (VA). This study assessed VA and low-contrast acuity (LCA) with Lea symbols in DS and CP populations to explore whether LCA measures provide useful additional information about visual performance. VA and LCA were also measured in a large group of typically developing young people.

METHODS. High-contrast VA and LCA performance was measured monocularly using crowded Lea symbols with 45 young people with CP (mean age 11.8 ± 4 years), 44 with DS (mean age 10.5 ± 3 years), and 211 controls (mean age 11.4 ± 5 years). Refractive status was confirmed with cycloplegic retinoscopy.

RESULTS. DS and CP groups had significantly lower acuities than controls at all contrasts (P < 0.001). Mean (±SD) high-contrast VA was as follows: DS = -0.39 ± 0.2 logMAR; CP = +0.18 ± 0.2 logMAR; controls = -0.04 ± 0.1 logMAR. Mean 2.5% LCA was as follows: DS = +0.73 ± 0.2 logMAR; CP = +0.50 ± 0.2 logMAR; controls = +0.37 ± 0.1 logMAR. For controls, the mean difference between VA and 2.5% LCA was 0.40 logMAR (95% limits of agreement, ±0.22 logMAR). While there was a positive relation between VA and 2.5% LCA scores (linear regressions, P < 0.0001), considerable variation existed, with VA explaining only 36% of the variance in LCA performance for control data.

CONCLUSIONS. VA and LCA performance was significantly poorer in DS and CP groups than in controls, and high-contrast VA did not reliably predict low-contrast performance. Therefore both high- and low-contrast acuity assessment are valuable to fully describe an individual’s visual function, and this may be particularly relevant in DS and CP in cases in which patients are unable to articulate visual difficulties. Age-specific reference data from a large sample of typically developing young people across a broad age range are presented for clinicians using high- and low-contrast Lea symbols. (Invest Ophthalmol Vis Sci. 2013;54:251–257) DOI:10.1167/iovs.12-10506

Down syndrome (DS) and cerebral palsy (CP) are common causes of intellectual and physical impairment in children with special needs. In both DS and CP, deficits in visual acuity (VA) are known to be common1–6; however, there is a paucity of information in the literature regarding these groups’ visual performance at low contrast. A few studies have reported contrast sensitivity losses among children and adults with developmental delay of various etiologies,7,8 but these studies grouped individuals with developmental delay or special needs together, and it is not possible to extract information about contrast sensitivity performance for those with DS or CP. In routine clinical practice, measures of low-contrast acuity are likely to be performed only if a patient reports symptoms of difficulties associated with poor contrast sensitivity or pathological signs that may degrade low-contrast performance. While DS and CP are commonly encountered by clinicians, such patients may be unable to report symptoms of reduced visual performance at low contrast. Furthermore, it has been suggested that low-contrast acuity or contrast sensitivity is an important functional measure to obtain and should be routinely attempted.9,10 Visual acuity is a resolution measurement of high-contrast black-and-white targets and does not reflect the lower contrasts present in everyday life, including tasks such as working in subdued lighting, reading, mobility, and facial recognition. Measures of contrast sensitivity have been found to predict performance in these tasks11–14 in studies of visual function in older adult populations with vision loss.

However, it may be challenging to obtain additional measures of contrast sensitivity or low-contrast acuity in children with developmental disability. What is the evidence that this is necessary, and will such measures add useful detail on visual performance, over and above standard measures of high-contrast VA?

The Lea symbols acuity charts utilize four simple symbols (house, circle, heart/apple, and square) and were designed to assess the VA of preschool children.15-17 Studies have shown that the Lea symbols acuity test tends to overestimate VA when compared to Early Treatment Diabetic Retinopathy Study (ETDRS),17 Landolt-C,18 and Bailey-Lovie charts,19 but provides a useful alternative to letter charts. Chen et al.20 examined the repeatability of Lea symbols in amblyopic and non-amblyopic children and found no significant difference in repeatability among normal, fellow, or amblyopic eyes. They report a test-retest measurement difference of ±0.10 logMAR in 93% of eyes, with 95% limits of agreement of ±0.18 logMAR.
Lea low-contrast acuity charts are used by some clinicians for functional visual assessments, sampling medium- and low-contrast acuity performance in addition to high-contrast VA. While data exist supporting the use of (high-contrast) Lea symbols acuity tests in young children, there are no published data relating to the low-contrast charts. As Lea symbols charts are frequently used by clinicians to obtain measures of visual function from older children and adults with learning or communication difficulties, normative data for these charts would be valuable.

The current study assessed low-contrast acuity in DS and CP populations using Lea symbols acuity charts in order to explore whether low-contrast measures provided additional information about visual performance. The study deliberately recruited a sample of participants with a wide range of acuities and ages. High- and low-contrast acuities measured from a large control group of typically developing young people are also presented to provide comparative data for participants with DS and CP across a wide age range.

**METHODS**

**Participants**

**Exclusion Criteria.** All participants were receiving ophthalmological/optometric eye care, and those with ocular pathology such as retinopathy of prematurity, keratoconus, retinal pathology, or cataracts were not invited to participate. During testing, the presence of any pathology was assessed by an experienced pediatric optometrist, and data from any participants with evident corneal or lenticular pathology were excluded from the analyses. The presence of strabismus did not exclude participants from the study, as acuity was measured with the dominant eye. This information was recorded during the testing protocol and incorporated into analyses.

**Cerebral Palsy.** Participants were 45 young people (25 male) with CP aged between 4 and 18 years (mean age 11.8 ± 4 years). Five participants from different regions sent an invitation to parents of young people with CP under their care to participate in the study. Information was also obtained from the CP register in Northern Ireland regarding Gross Motor Function Classification System (GMFCS) and CP subtype. According to the GMFCS, 1 participant was level I; 14 were level II; 13 were level III; 6 were level IV; and 11 were level V. Thirty-five participants had the spastic subtype of CP; 7 had the dyskinetic form of CP; and 3 had ataxic CP. The GMFCS categorizes those with CP by self-initiated movement and uses a grading scale from level I (walks without limitations) up to level V (requires the use of a wheelchair). Nystagmus was noted in 6 participants, and 11 had strabismus (6 exotropic, 2 hypertropic, and 3 esotropic).

**Down Syndrome.** Participants were 44 young people with DS (24 male) aged between 6 and 17 years (mean age 10.5 ± 3 years). They were invited to participate with written information after identification by local ophthalmologists and DS parent support groups. Nystagmus was noted in 4 participants, and 4 had strabismus (all esotropic).

**Controls.** A total of 211 typically developing young people (94 male) aged between 5 and 17 years acted as age-matched controls for this study (mean age 11.4 ± 3 years). Control participants were recruited from state primary and postprimary schools that were nonselective in academic ability and drew children from a wide range of socioeconomic backgrounds. None of the control group had clinically evident nystagmus. Six esotropias, one microtropia, and two exotropias were noted.

The parents of all participants were sent an information pack explaining the nature of the study. Informed consent was obtained from the parents/guardians of the participants, and verbal assent was given by the participant (where possible) on the day of testing. Data collection was conducted across a range of sites including the participant’s school and community eye clinics.

Ethical approval for the study was obtained from the University of Ulster Research Ethics Committee and NHS Research Committees and Research Governance, and the research followed the tenets of the Declaration of Helsinki.

**Procedure**

High- and low-contrast visual acuities were measured monocularly for the dominant eye using four crowded Lea symbols charts at (high-contrast) VA and 2.5% contrast placed within a light box of high illumination (85 cd/m²). The charts were consistently presented in descending order of contrast level. Ocular dominance testing using a finger-pointing technique established the dominant eye, and this eye was used for subsequent monocular acuity measures. Where ocular dominance was difficult to interpret, high-contrast VA was measured for right and left eyes, and the eye with the better acuity was used for subsequent monocular measures. Testing was carried out under full room illumination with the participant seated 5 m from the chart. Participants either matched or verbally indicated their choice of symbol during testing. Measurement began by familiarizing participants with the symbols and, starting from the top line, asking participants to identify the first and last symbol on each line. Once the participant had difficulty identifying the first and/or last symbol on the line, he or she was directed to attempt all symbols two lines above that. If the participant identified three out of the five symbols correctly, he or she was directed to the next line below. Acuity was recorded using by-symbol scoring (~0.02 logMAR for each symbol correctly identified). Test time was approximately 4 to 6 minutes for each participant.

Each participant’s refractive error was assessed by an experienced optometrist 30 minutes after instillation of 1.0% cyclopentolate hydrochloride. For the CP group, refractive errors had been measured on a previous visit, and prescriptions were changed where necessary so all were known to be wearing an up-to-date prescription. For the control and DS groups, data were collected at a single visit and participants wore their habitual correction. For the control and DS participants, cycloplegic refraction was conducted after vision testing, and it was beyond the limits of the study protocol to retest on occasions when a significant (or a significantly different) refractive error was found. In the DS group, seven participants’ VA data were excluded from analysis due to significantly undercorrected or uncorrected spectacle prescriptions (defined as an undercorrected mean spherical equivalent greater than +1.50 diopter (D) or less than −0.50 D, or an undercorrected cylindrical prescription greater than 1.25 D of cylinder), and data from four of the control group were excluded using the same criteria. Notification of this requirement for an updated refractive correction was sent to the clinician responsible for these individuals’ eye care.

**Statistical Analysis**

Statistical software packages Origin 7.5 (OriginLab Corp., Northampton, MA) and StataCorp LP (College Station, TX) were used for graphical presentation and data analysis. Linear regression analyses, Student’s t-test, and one-way and multivariate analyses of variance were used to compare data within and between groups, with statistical significance defined at 5% levels. Mann-Whitney analyses were employed for nonparametric statistical tests. Data for 2.5% contrast was further explored, as the instructions in use of the low-contrast Lea symbols report this to be the most clinically useful low-contrast test and indicate that one should expect 2.5% contrast scores to be half those recorded at high contrast. Assuming that this indicates a 2-fold reduction in performance, this would equate to a 0.3 logMAR reduction in acuity from high to 2.5% contrast.

The sample sizes used in this study were powerful with regard to detecting differences in acuity, comparing the current study’s control
TABLE 1. Control Group Data: Median Visual Acuities and Low Contrast Acuities

<table>
<thead>
<tr>
<th>Acuity Chart</th>
<th>Control Group, Median (IQR) (LogMAR)</th>
</tr>
</thead>
<tbody>
<tr>
<td>5-8 to 7.9</td>
<td>0.08 (-0.16 to -0.00)</td>
</tr>
<tr>
<td>8 to 9.9</td>
<td>0.08 (-0.14 to -0.00)</td>
</tr>
<tr>
<td>10 to 11.9</td>
<td>0.08 (-0.12 to -0.00)</td>
</tr>
<tr>
<td>12 to 13.8</td>
<td>0.12 (0.04 to 0.26)</td>
</tr>
<tr>
<td>14 to 16.8</td>
<td>0.16 (0.02 to 0.44)</td>
</tr>
<tr>
<td>16.8 to 18</td>
<td>0.26 (0.04 to 0.44)</td>
</tr>
</tbody>
</table>

Median and interquartile range (IQR) for high- and low-contrast acuity data for the control group (LogMAR). Control data have been grouped into five age categories, and the final column summarizes data for the whole group.

Success Rates

High-contrast VA was successfully measured in 92% of the DS and 84% of the CP group. Acuities could not be measured for three participants with DS (8%) and seven participants with CP (16%). The main reason some in the CP group were not able to undergo testing was limitations in mobility (impacting the ability to match symbols) and communication (nonverbal). Six of these nonsuccessful participants were level V on the GMFCS scale, and one had a level IV classification. Five of the nonsuccessful participants had strabismus. For the DS group, despite careful instruction and practice on symbol matching, three participants could not perform this task. Acuities at all contrast levels were successfully measured on 20 (59%) participants with DS and 25 (66%) participants with CP. The lowest-contrast chart yielded significantly poorer success rates for the DS and CP groups (one-way analysis of variance, F(3,4) = 11.3, P = 0.02).

Comparison of High-Contrast Acuity Data and Age of Participants

There were no significant differences in the age of participants between groups (t-test between DS and control groups, t = 1.21, P = 0.25, and between CP and control groups, t = -0.92, P = 0.36). Data were found to be normally distributed for the DS (P = 0.13) and CP groups (P = 0.55), but not normally distributed for the control group (P = 0.02); therefore nonparametric statistics were used for comparisons between control and DS/CP data.

There was no significant association between acuity and age in either the DS or the CP group (linear regression analyses, R² between 0.02 and 0.08, P > 0.1). For the CP group, inspection of high-contrast VA according to level of motor impairment (by GMFCS) indicated a reduction in acuity with increasing GMFCS, but a one-way analysis of variance comparing acuities between GMFCS classifications did not reach significance (F(4,33) = 1.7, P = 0.19). Within the control group, there was a significant improvement in high-contrast VA with age (linear regression analysis, R² = 0.18, F(1,206) = 45.6, P < 0.0001).

Control Group High- and Low-Contrast Acuity Data

Table 1 reports the control reference data as median (and interquartile range) acuities for all contrasts for the control group. These have been separated into five different age categories, as high-contrast and 10% contrast acuity varied by age (P < 0.0001). For 1.25% low-contrast acuity, linear regression analysis revealed a slight worsening of acuity with age (R² = 0.02, P = 0.04). There was no significant difference in 2.5% low-contrast acuities across age groups (linear regression analysis, P = 0.13).

High- and Low-Contrast Acuity Data for DS and CP Groups

Table 2 reports the mean acuities for all contrasts for the CP and DS groups. Measures of acuity were not significantly affected by the presence of strabismus in any of the groups.
Table 2. CP and DS Group Data: Mean Visual Acuities and Low-Contrast Acuities

<table>
<thead>
<tr>
<th>Acuity Chart</th>
<th>CP Group, Mean ± SD (LogMAR)</th>
<th>Participants Successful in Completing Testing at All Contrasts, n = 25</th>
<th>DS Group, Mean ± SD (LogMAR)</th>
<th>Participants Successful in Completing Testing at All Contrasts, n = 20</th>
</tr>
</thead>
<tbody>
<tr>
<td>High contrast (VA)</td>
<td>+0.18 ± 0.18, n = 38</td>
<td>+0.18 ± 0.14, n = 25</td>
<td>+0.39 ± 0.19, n = 34</td>
<td>+0.32 ± 0.15, n = 20</td>
</tr>
<tr>
<td>10% contrast</td>
<td>+0.30 ± 0.19, n = 36</td>
<td>+0.32 ± 0.13, n = 25</td>
<td>+0.55 ± 0.19, n = 34</td>
<td>+0.45 ± 0.13, n = 20</td>
</tr>
<tr>
<td>2.5% contrast</td>
<td>+0.50 ± 0.15, n = 34</td>
<td>+0.53 ± 0.14, n = 25</td>
<td>+0.75 ± 0.16, n = 31</td>
<td>+0.68 ± 0.13, n = 20</td>
</tr>
<tr>
<td>1.25% contrast</td>
<td>+0.67 ± 0.12, n = 25</td>
<td>+0.67 ± 0.12, n = 25</td>
<td>+0.82 ± 0.11, n = 20</td>
<td>+0.82 ± 0.11, n = 20</td>
</tr>
</tbody>
</table>

Mean high- and low-contrast acuity data for the CP and DS groups, for all participants, and for participants who successfully completed testing for all contrast levels (mean ± standard deviation presented; LogMAR units).

Discussion

The measurement of high- and low-contrast acuities was successful in the majority of participants with CP (66%) and DS (59%), demonstrating that Lea symbols are appropriate for participants with a broad range of motor and intellectual impairment. This is superior to the results of Neilsen et al., who reported that it was possible to evaluate contrast sensitivity using Cambridge low-contrast grating tests in 40% of their population of children with developmental disability aged between 4 and 15 years. In the present study, acuities could not be measured for seven participants in the CP group due to limitations in mobility and communication. Six of these nonsuccessful participants were level V on the GMFCS scale, and one had a level IV classification. This is consistent with findings of Ghasia et al., who report a significant reduction in success with optotype acuity testing for participants classified as level V on GMFCS. Success rates also decreased as the testing procedure continued, as attention and cooperation could not be maintained for the lower-contrast acuity levels with some participants in the DS and CP groups, and the lowest-contrast chart yielded significantly poorer cooperation rates. Four (2%) control participants reported that they could not discern any symbols on the lowest-contrast chart, despite relatively good acuity at other contrasts.

This study presents age-specific low-contrast reference data for typically developing children, which provide a useful guide for clinicians using the Lea symbols charts to assess vision of older children with learning or communication difficulties. In addition, these data suggest that, on average, a four-line (0.4 logMAR) reduction in acuity should be expected when one is comparing high-contrast acuity with a 2.5% low-contrast acuity measurement using Lea symbols.

As one would expect, low-contrast performance was positively correlated with high-contrast VA measurements, but the coefficient of determination demonstrates that only 36% of the variance of 2.5% low-contrast acuity is explained by high-contrast VA. Other studies have shown a wide range of contrast sensitivity measures in pediatric and adult populations with normal VA, with West et al. reporting a 66% variance between VA and Pelli-Robson contrast sensitivity measures. Furthermore, while on average, the contrast data suggest a 0.4 logMAR (four line) reduction from high- to low-contrast performance, the coefficient of determination demonstrates that only 36% of the variance of 2.5% low-contrast acuity is explained by high-contrast VA.
contrast acuity, there are broad 95% limits of agreement for this value (±0.22 logMAR; from two to six lines of reduction). Consequently, some individuals in all participant groups showed marked deviations from this average relationship, suggesting that for clinicians to obtain a full picture of a child's visual function, both high- and low-contrast measures should be attempted. This is particularly relevant where learning or communication difficulties prevent a child from reporting symptoms of poor function at low contrast. While this appears surprising for control data, analysis of those with greater than expected differences revealed no significant relation between the magnitude of refractive error and high-contrast acuity. The only significant relationship from postestimation of multivariate analysis of variance was that participants with a greater difference in low-contrast acuity compared with high-contrast acuity tended to be older.

Chen et al. reported a test-retest difference between ±0.10 logMAR in 93% of intrasubject acuity measures using the
related to reading speed 10,11 and daily living tasks and comparison across participants and with reference data. Using a single technique, which allows more rigorous without logMAR design). In the current study, VA was assessed and picture uncrowded and crowded optotypes (with and et al.,24 who reported mean VA of their DS group ((0.35 logMAR) worse than controls. However, both these studies pooled VA data from a number of different acuity tests, including spatial sweep VEP,23 preferential looking,24 and letter and picture uncrowded and crowded optotypes (with and without logMAR design). In the current study, VA was assessed using a single technique, which allows more rigorous comparison across participants and with reference data. Previous authors have reported that contrast sensitivity is related to reading speed10,11 and daily living tasks and mobility14 in older adult populations. While there are limited data regarding the additional impact that contrast sensitivity deficits may have for children with impaired VA, Neilsen et al.7 highlight the importance of raising awareness of contrast sensitivity as an aspect of vision that may be impaired in children with developmental disability. Consequently, where high-and low-contrast acuity performance is impaired, parents, teachers, and caregivers of children with DS and CP should be provided with advice regarding good lighting and appropriate sizing and contrast of educational and recreational material to ensure that visual difficulties do not restrict access to visual material, impede communication and independence, and/or compound underlying intellectual deficits.

**Strengths**

Participants were recruited to ensure that the relation between high- and low-contrast acuity could be explored across a spectrum of visual, communication, and intellectual abilities and ages. Acuity data were included only in cases in which it was established that participants had been tested wearing an up-to-date spectacle correction ascertained by cycloplegic refraction. There may be a variety of underlying causes for the reduced acuity and contrast sensitivity in the CP and DS groups (including amblyopia, cerebral visual impairment, optic nerve/foveal hypoplasia, and optic neuropathy).2,5,25,27 However, the aim of the present study was to explore the additional benefit of measuring low-contrast acuity in a typical clinical population of DS and CP participants. The sample size was sufficient for the study to have a power of over 99% for both groups.

**Limitations**

Acuity was measured in descending order of contrast. This may have caused some fatigue effects with regard to the lowest-contrast chart, and may explain the greater variation both in compliance and in measured acuities found at this level. However, participants were given plenty of time and breaks during testing to ensure that the protocol was not onerous.

In conclusion, this study demonstrates that for a full clinical picture of any child’s functional ability, measures of both high- and low-contrast acuity are valuable. The reason is that while good performance at high contrast generally predicts good performance at low contrast, individual variation exists, and some individuals with excellent high-contrast acuity concurrently demonstrate poor low-contrast performance. However, low-contrast acuity data are likely to be more important when VA is already reduced, as additional contrast deficits will further impact the visual ability of the given individual. For typically developing children with good VA and no symptoms of visual difficulty, information on low-contrast acuity may not aid management. Reference data are provided that the clinician can use to interpret results obtained from children with a developmental disability between the ages of 5 and 16 years to ascertain whether they suggest the presence of a visual impairment.

It is likely that clinicians assessing both high- and low-contrast acuity will employ a single high- and a single low-
contrast chart rather than assess across all the contrasts available in the Lea series. Consistent with the Lea symbols instructions, the authors recommend the 2.5% contrast chart for the latter. In addition, this study reveals that one should expect a $0.40 \pm 0.22$ logMAR (mean $\pm$ 95% limits of agreement) reduction in acuity from high- to low (2.5%)-contrast acuity.

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References