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Different visual development: norms for visual acuity in children

3

with Down's syndrome

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7 **Abstract**

8

9 **Background:** Visual acuity is known to be poorer in children with Down's syndrome than in
10 age-matched controls. However, to date, clinicians do not have access to norms for children
11 with Down's syndrome that allow differential discrimination of healthy from anomalous visual
12 development in this population.

14 **Methods:** The Down's Syndrome Vision Research Unit at Cardiff University has been
15 monitoring visual development in a large cohort of children since 1992. Cross-sectional data
16 on binocular visual acuity were retrospectively analysed for 159 children up to 12 years of age
17 in order to establish binocular acuity norms. Longitudinal binocular acuity data were available
18 for nine children who were seen regularly over the 12 years age-range. Monocular acuity was
19 successfully recorded less often in the cohort, but analysis of scores for 69 children allowed
20 assessment of inter-ocular acuity differences and binocular summation.

22 **Results:** In comparison with published norms for the various test used, binocular acuity was
23 consistently poorer in children with Down's syndrome from the age of three years and
24 stabilised at around 0.25 logMAR from the age of four years. Inter-ocular acuity difference
25 and binocular summation were both 0.06 logMAR, which is similar to the reported values in
26 children without Down's syndrome.

28 **Conclusions:** The study provides eye-care practitioners with the expected values for
29 binocular acuity in children with Down's syndrome and demonstrates the visual disadvantage
30 that children with Down's syndrome have when compared with their typically developing
31 peers. The results emphasise the responsibility that practitioners have to notify parents and
32 educators of the relatively poor vision of children with Down's syndrome, and the need for
33 classroom modifications.

34 **Introduction**

35 It is well reported that children with Down's syndrome have poorer visual acuity than expected for
36 age(1-3) even when refractive errors are corrected. Objective measurements of acuity by visual
37 evoked potentials have shown that the deficit is not explained by lack of concentration, motivation
38 or persistence in acuity testing(4) and other studies implicate the quality of the optics in reducing
39 visual acuity in children with Down's syndrome(5). This may present a problem for clinicians
40 examining children with Down's syndrome, in discriminating between an acceptable or 'normal'
41 level of visual acuity and a poor acuity indicative of amblyopia or pathology.

42 Children with Down's syndrome exhibit a number of characteristics different from typically-
43 developing children; among them is retarded growth - children with Down's syndrome are generally
44 small for age. The Down's Syndrome Medical Interest Group, a UK and Ireland organisation of health
45 care practitioners (mainly paediatricians), publishes growth charts specifically for children with
46 Down's syndrome, which paediatricians can use to monitor a child's growth compared to the
47 appropriate norms. Refractive error profiles are available for children with Down's syndrome up to
48 the age of 15 years, but no norms for visual acuity have yet been published.

49 Our research group, the Down's Syndrome Vision Research Unit, has been involved in a longitudinal
50 study of visual and ocular development of children with Down's syndrome since 1992 and we
51 therefore have the data available to develop norms for the use of eye care practitioners. At the
52 outset, recruitment of very young children was through paediatricians in South and West Wales, but
53 since then we have targeted older children in the local area, at various times for specific research
54 studies. More recently, as parents have become aware of our work, families have contacted us
55 directly requesting to join our studies; we have no exclusion criteria for families wishing to enrol,
56 except that the child must have a diagnosis of Trisomy 21. Although most children in the study
57 cohort live in South and West Wales, some children travel considerable distances to take part.

58 Children participate in conventional eye examinations as well as in laboratory-based experiments.
59 Over the years, eye examinations have been conducted in the children's homes, on school premises
60 and/or in the clinic at Cardiff University School of Optometry & Vision Sciences. Clinical data include
61 visual acuity, refractive error, accommodation, and binocular status and are available for a total of
62 226 participants at various ages. We therefore have ample data to establish normative values.

63 The longitudinal study has had continual and on-going ethical approval from the appropriate bodies
64 covering NHS ethics in Wales (the actual institutions have changed over the 25 years of the study).
65 Parental consent was given for all data collected and the study conducted in accordance with the
66 Declaration of Helsinki.

67 **Methods**

68 The Down's Syndrome Vision Research Unit database was used to retrospectively examine the
69 record cards of all children seen between December 1992 and April 2017. 'Normal values' should
70 ideally be collected from a non-clinical population, since a subject group presenting at a clinic cannot
71 be expected to be representative of the general population. The published normal values for
72 typically developed children (6-9) generally report on a non-clinical population, but use exclusion
73 criteria, particularly for refractive errors, to ensure 'normal vision'. The only norms available for the
74 Keeler Crowded test, on the other hand (10), were obtained from children referred to an orthoptic
75 clinic, and determined to be non-strabismic. For the early years of our longitudinal study, children
76 with Down's syndrome in South and West Wales were identified by the Cytogenetics Department of
77 the University Hospital Wales, and then recruited through the children's paediatricians. Only two

78 families refused to join the study at this stage. An additional recruitment campaign was initiated for
79 the bifocal spectacle trial (11), through Educational Psychologists, without reference to visual
80 concerns. Children who did not go on to participate in the bifocal trial, either because they did not
81 have an accommodative deficit, or because they could not be satisfactorily matched to another
82 child, remained in the cohort. At this stage then, the study group of 'early' recruits was not a clinical
83 population. Since the early 2000's, as our work has become increasingly well-known, families have
84 contacted us directly requesting to join our studies; we have no exclusion criteria for families
85 wishing to enrol, except that the child must have a diagnosis of Trisomy 21. Many families joining the
86 study in this way have no prior concerns about their child's vision; nevertheless, it is possible that
87 children joining through this route are more likely to have visual deficits. We therefore identified
88 these children in the current analysis as 'late' recruits. Each year a large number of families from all
89 over the UK simply request clinical appointments for their child with Down's syndrome. These
90 children are not enrolled into the longitudinal study, although they may take part in other aspects of
91 the team's studies.

92 From the database of 226 participants, children with visually-impairing conditions such as aphakia
93 (N=1) or nystagmus (N= 39) were excluded from analysis, as were children who joined the study
94 after the age of 12 years (N=9); 177 children remained, including those with strabismus.

95 The database contains data on children before they were prescribed spectacles for significant
96 refractive errors. Those visits at which the child had uncorrected refractive error were excluded
97 according to the following criteria. The criterion for significant hypermetropia was identical to that
98 used for the studies of normal values of visual acuity in children, for the tests we used. For Teller
99 cards this was $>+5.00D$ (8), for Cardiff Acuity Test (6) and Keeler LogMAR (10) this was $>+3.00D$ and
100 for Kay Pictures (9) $>+2.00D$. Three children were excluded because there were no later visits when
101 refraction was corrected; other children had later data after provision of spectacles. Low myopia
102 was not considered detrimental to PL acuity, and there were no exclusions for uncorrected myopia.
103 When distance optotype tests were used, no child had uncorrected myopia. Further exclusions (N=5)
104 were visits on which binocular acuity data were not obtained.

105 The database of 159 children was then inspected without acuity data, to prevent any bias, and
106 children were allocated to age groups. Grouping was at 6-monthly intervals for up to 2 years, since
107 acuity is expected to change rapidly in infancy. Thereafter, grouping was two-yearly up to 11.9 years.
108 The following 9 age groups were created:

109 1-5.9 months, 6-11.9 months, 12-17.9 months, 18-23.9 months, 2-3.9 years, 4-5.9 years, 6-7.9 years,
110 8-9.9 years, and 10-11.9 years.

111 Each of the 159 children was allocated to only one age group, to provide cross-sectional data on
112 visual acuity development, and allocations were made without reference to visual acuity scores, and
113 so that an approximately even distribution of participants numbers resulted across the age groups.
114 When a child had been seen on more than one occasion within a designated age group, the visit
115 which was closest to the centre of the age range was chosen (e.g. the centre of the 2-3.9 year age
116 group was 34.5 months).

117 Seven optometrists have been involved in the study over the years, and all contributed some of the
118 data used in this analysis. All were highly experienced at examining children with Down's syndrome.

119 Visual acuity was measured on each occasion by age and ability-appropriate tests, and thus varied
120 among the participants even within one age group. The tests used were, however, limited to the
121 following, which all have LogMAR-based acuity scales and for which norms are available:

122 Teller Acuity Cards(12), Cardiff Acuity Test(13), Kay Pictures LogMAR (singles or crowded)(14), Keeler
 123 LogMAR Crowded test (formerly Glasgow Acuity Cards – only the crowded version was used)(15).

124 Binocular acuity was available for all children in the final database, and the norms derived for
 125 binocular acuity. Monocular acuity was measured if the child could tolerate both occlusion and
 126 repeated measurements, which was relatively infrequent. If the child had spectacles for a distance
 127 refractive error, acuity was ‘corrected acuity’ with current spectacles. If a change of refractive error
 128 was found during an examination, ‘best corrected’ acuity was rarely recorded with the trial frame,
 129 since the unfamiliar experience, discomfort of the trial frame and further repeat of the
 130 measurement was unlikely to be tolerated. If no spectacles had been prescribed, then acuity was
 131 obviously ‘uncorrected acuity’.

132 In line with common clinical practice, spectacle prescriptions were not issued for infants. The
 133 youngest child with corrected acuity was 12.9 months old. In general, because accommodation is
 134 known to be poor in the majority of children with Down’s syndrome(16, 17), our protocol is to
 135 prescribe for hypermetropia whenever uncorrected accommodation at near is poor (outside the
 136 normal lag)(18, 19); this often means prescribing for lower amounts of hypermetropia than the
 137 protocols adopted by many clinics and prescribing the full amount of hypermetropia. Myopic
 138 children are prescribed spectacles when the child begins to take an interest in distance viewing; this
 139 is determined in discussion with parents and is usually between the ages of 2 and 3 years.
 140 Astigmatism is always incorporated in prescriptions for hypermetropia or myopia. Astigmatism alone
 141 (i.e. when the equivalent sphere is emmetropia) is corrected when it is over 2.00DC. Visual acuity is
 142 never used as a criterion for spectacle prescription. When children were examined later than our
 143 bifocal trial(11), bifocals were prescribed for all children with a persistent accommodative lag (i.e.
 144 present on two consecutive visits) with full distance error correction, over the age of 2 years.

145 **Results**

146 Table 1 shows the numbers of children in each age group, the numbers of children using each
 147 available test and the numbers of children wearing spectacle correction for acuity testing. Key:
 148 T=Teller cards, C=Cardiff Acuity Test, KC= Kay Pictures crowded, KS=Kay Pictures singles, KL= Keeler
 149 Crowded.

Age group	1-5.9 months	6-11.9 months	12-17.9 months	18-23.9 months	2-3.9 years	4-5.9 years	6-7.9 years	8-9.9 years	10-11.9 years
No. of children	17	18	16	16	16	20	21	16	19
No. using tests	17 T	16 T 2 C	8 T 8 C	5 T 11 C	5 T 11 C	15 C 4KC 1 KS	10 C 9 KC 2 KL	6 C 8 KC 1 KS 1 KL	6 C 9 KC 4 KL
No. wearing Rx	0	0	1	2	5	10	12	12	12

150

151 Binocular acuity data were not normally distributed for one of the age groups (2-3.9 years, Shapiro-
 152 Wilk, $p < 0.04$), but since the remaining age groups had normally distributed data, it was decided to
 153 use means and confidence intervals in Figure 1, which shows the binocular acuity values for each
 154 group, in line with published normal values for typically developing children.

155 Table 2 shows the binocular acuity norms that eye care practitioners can expect in children with
 156 Down’s syndrome without significant uncorrected refractive error.

Age group	1-5.9 months	6-11.9 months	12-17.9 months	18-23.9 months	2-3.9 years	4-5.9 years	6-7.9 years	8-9.9 years	10-11.9 years
Mean binocular acuity (LogMAR)	1.15	0.81	0.67	0.5	0.37	0.28	0.28	0.18	0.25
Range (95% confidence interval)	0.75 to 1.54	0.51 to 0.10	0.18 to 1.16	0.14 to 0.85	-0.01 to 0.74	-0.04 to 0.59	0.08 to 0.49	-0.12 to 0.47	-0.01 to 0.50
Proportion failing vision screening criteria						11/20 55%	12/21 57%	5/16 31%	8/19 42%

158

159 Although the normal range expressed in Figure 1 and Table 2 suggests that acuities as good as -0.12
160 can be scored for children with Down's syndrome, in practice this may not be achieved. Among the
161 cross-sectional data presented here, only five children achieved LogMAR 0.0 acuity binocularly; one
162 was aged 4-5.9 years, three were aged 8-9.9 years and one was aged 10-11.9 years; no child
163 achieved better than 0.0 LogMAR. In the UK, the national guidelines for vision screening of 4-5 year
164 olds specify the pass criteria of LogMAR 0.2 (monocular acuity; since binocular acuity is usually
165 slightly better than monocular, applying the criteria to this current data would be expected to
166 maximise pass rates). Applying this criterion to children with Down's syndrome in the over 4 years
167 age groups, as Table 2 shows, means that a total of 46% of children would fail standard vision
168 screening.

169 Acuity may vary considerably with the test used. In particular, the pooling of data recorded by
170 preferential looking with data from optotype tests in establishing norms may be questioned. To test
171 this, all binocular acuities were categorised as PL (Teller or Cardiff) or optotype (Kay Pictures or
172 Keeler letters) and an analysis of co-variance was carried out, with age as the covariate. There was
173 no significant difference between acuities recorded by PL and by optotype when age was taken into
174 account ($F=1.17$, $p=0.28$). Since at the older ages, some children were still dependent on preferential
175 looking, combining the test scores to give a single norm is justified.

176 Another confounding factor in the study is the different recruitment sources, and in particular the
177 likelihood of self-selection bias when parents elected to join the study. An analysis of co-variance
178 was again used to determine any effect of recruitment source, define as 'early' or 'late' on the
179 binocular acuity score, with age as the covariate. There was no significant difference between
180 acuities recorded from the early or late recruits, when age was taken into account ($F=0.01$, $p=0.90$)

181 Nine children were identified who had data for at least 8 of the 9 age groups, which therefore
182 allowed longitudinal analysis of binocular acuity and this is shown for the individuals in Figure 2.
183 Each child shows a progression of acuity in keeping with the cross-sectional data. Note that no
184 acuity score is better than LogMAR 0.0.

185 Monocular acuity was available for 69 children without strabismus, with ages ranging from 3.9
186 months to 11.6 years. Mean interocular acuity difference was calculated, using the absolute

187 difference between right and left eyes. The mean interocular acuity difference was LogMAR 0.06
188 (± 0.12) and was not influenced by age ($r=0.22$, $p=0.86$) or by test type (preferential looking v.
189 optotype, $t=0.26$, $p=0.80$). The differences between better-eye and binocular acuity (i.e. binocular
190 summation) was available for 64 children, and mean difference was LogMAR 0.06 (± 0.07) and was
191 not influenced by age ($r=0.16$, $p=0.2$) or by test type ($t=0.26$, $p=0.80$).

192 **Discussion**

193 This retrospective analysis confirms the previous reports of poor acuity in children with Down's
194 syndrome, even when refractive errors are corrected, at all but the youngest ages. Courage and
195 Adams(1) used the Teller Acuity Cards to record acuity for participants with Down's syndrome from
196 2 months of age, and noted a deviation from published norms after 6 months of age. Woodhouse et
197 al(3) used Teller Acuity Cards for younger children and Cardiff Acuity Test for older children both
198 with and without Down's syndrome aged 3 to 57 months and showed poorer acuity in Down's
199 syndrome after 24 months. The current analysis (see Figure 1) also suggests a deviation from the
200 expected norms between 3-4 years such that almost half of all children with Down's syndrome over
201 the age of 4 years would fail standard vision screening.

202 A recent longitudinal evaluation by Tomita(2) suggested that development of visual acuity is delayed
203 rather than abnormal in Down's syndrome, since 50% achieved 0.0 LogMAR by 3 years and 100% by
204 6 years. However, the authors presented no comparative data from typically developing children,
205 and used non-standardised test procedures. Whether the pictures or Landolt C tests used for 3-6
206 year olds were presented singly or crowded and whether LogMAR scales were incorporated was not
207 stated.

208 The current analysis of both cross-sectional and longitudinal data suggests that acuity lies within the
209 typical norms for infants, when preferential-looking tests are used exclusively, but the rate of acuity
210 improvement separates from that of typically developing children from about 3-4 years, and there
211 appears little further acuity development beyond 5 years. Of course a limitation of the current
212 analysis is the use of different tests for acuity; theoretically, preferential looking and optotype tests
213 measure different aspects of vision. In clinical settings, ability appropriate tests must be used; it is
214 clear that a poor score would result from the use of too complex a test for a child and similarly, the
215 use of too simple a test could mean loss of interest in a child subject. In general, children with
216 Down's syndrome would be expected to use a simpler test for age than typically developing children,
217 and in this analysis, we report that a proportion of children were still reliant on preferential looking
218 (the Cardiff Acuity Test) beyond the age of 4 years, when typically developing children would be
219 expected to have progressed to Kay Pictures or letter tests. There is evidence that preferential
220 looking tests over-estimate acuity and are less sensitive to refractive errors and amblyopia (20, 21) in
221 typically developing children, although our analysis showed no difference in scores between children
222 using the preferential looking and optotype tests. The purpose of this analysis is the development of
223 norms for clinical purposes, and we find no difference between the type of test used. This suggests
224 that choosing the test to suit a child's ability is entirely appropriate and the norms can be considered
225 equivalent. Further, the use of simpler tests in children with Down's syndrome could be expected to
226 minimise any differences in acuity. Instead, the mean values for binocular acuity in children with
227 Down's syndrome are 0.1 to 0.4 LogMAR poorer than typically developing children from the age of 4
228 years.

229 In our study, 'best corrected' acuity was not measured, since trial frames were not used for acuity.
230 However, none of the published norms use best corrected acuity and one (norms for Kay Pictures
231 LogMAR (9)) did not measure refractive errors at all in the subject group; uncorrected refractive

232 errors could be a confounder in their data. The study is included here simply because it provides
233 norms for an age group not otherwise represented with the Kay Pictures test. Even with the
234 potential for uncorrected refractive errors, the acuity scored for typical children is, on average, 0.2
235 better than for children with Down's syndrome.

236 Accommodative deficits are common among children with DS(16, 17, 22), and our group now
237 corrects such defects with bifocals (11). However, children whose visits were in the early years of
238 our studies would not have bifocals. The presence of a bifocal segment or an accommodative lag
239 would not be expected to influence acuity measures with Kay Pictures or Keeler LogMAR tests,
240 which are conducted at 3 metres. Preferential looking tests (Teller Cards and Cardiff Acuity Test) are
241 conducted at closer working distances and could be influenced by an uncorrected accommodative
242 deficit. However, the norms for Down's syndrome match the norms for typical children when
243 children are very young and entirely dependent on preferential looking. The ages at which a
244 significant proportion of children are using tests at 3 metres, are the ages at which acuities in
245 Down's syndrome are poorer than acuities for typical children.

246 Little et al (5) assessed acuity in children with Down's syndrome and typical children by conventional
247 grating targets and by interferometric generated grating targets, which eliminated the optical quality
248 of the eyes. Both tasks required the children to identify whether the stripes were horizontal or
249 vertical and were as closely matched as possible for cognitive demand. Acuity for children with
250 Down's syndrome improved for interferometric targets by a factor of FOUR compared to typical
251 children, although acuities were still significantly poorer in Down's syndrome. Since the tasks were
252 the same, this suggests that the poor acuity for conventional targets was not due to behavioural
253 issues. Similarly, the poor acuity demonstrated in the current study is unlikely to be behavioural in
254 nature.

255 Binocular summation and interocular acuity difference values are available for typically-developing
256 children, for some of the tests used here. Binocular summation for Teller Acuity Cards (estimated
257 from the published data(8)) is up to 0.07 LogMAR, and for Cardiff Acuity Test(6) up to 0.3 LogMAR.
258 For children with Down's syndrome, we found a mean value of 0.06 across all test types. Interocular
259 acuity differences are reported to be up to 0.15 LogMAR for Teller Acuity Cards(8), up to 0.1 LogMAR
260 for Cardiff Acuity Test(6), up to 0.15 LogMAR for Kay Pictures Crowded(9) and, on average, 0.03
261 LogMAR for Keeler LogMAR Crowded test(10). We found a mean interocular acuity difference of
262 0.06 LogMAR across all test types. Thus for children with Down's syndrome these aspects would
263 appear to be within the expected range, suggesting that acuity measurements are reliable in
264 children with Down's syndrome but the absolute level of visual acuity can be expected to be poorer
265 than for typical children.

266 The availability of 'normal' values as produced here, will allow eye care practitioners to reassure
267 parents when their child's vision is within the expected range for Down's syndrome. However, visual
268 acuity is poorer than expected when compared to typically developing children. Studies suggest that
269 contrast sensitivity too is reduced in children with Down's syndrome(4, 23). It is, therefore,
270 imperative that practitioners explain to parents and to teachers that a child's vision is below normal
271 when compared to classroom peers. Children with Down's syndrome are considered to be visual
272 learners(24), that is, they are more reliant on vision for learning than are typically-developing
273 children. It could be argued then, that reduced vision is more detrimental to learning in a child with
274 Down's syndrome, than it is in a typically-developing child, who can compensate for vision loss by
275 making use of their auditory and cognitive skills. Teachers need to be aware that the child in their
276 classroom who has Down's syndrome is not seeing their school work as readily as the other children.
277 If the teacher is not aware that a child's vision is poor, then inability to carry out tasks in the

278 classroom may be considered to be due to the learning disability and nothing done to address the
279 issue. Simple enlargement of learning material may be all that is needed. Eye care practitioners have
280 a responsibility to keep parents and teachers fully informed of a child's visual development, *in*
281 *comparison to typical children*, as well as with reference to the expected values for Down's
282 syndrome, in order that everyone associated with a child can understand their capabilities and what
283 needs to be done to support their learning.

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356 Figure 2. Mean binocular acuity values for individual children with Down's syndrome followed
 357 longitudinally, in comparison with the cross-sectional data (mean and 95% confidence limits) from
 358 Figure 1. Note that each child in the longitudinal data set contributes only ONE data point to the
 359 cross-sectional values.

