Are language and social communication intact in children with congenital visual impairment at school age?

Valerie Tadić, Linda Pring, and Naomi Dale

Abstract: Development of children with congenital visual impairment (VI) has been associated with vulnerable socio-communicative outcomes often bearing striking similarities to those of sighted children with autism. To date, very little is known about language and social communication in children with VI of normal intelligence. Methods: We examined the presentation of language and social communication of 15 children with VI and normal-range verbal intelligence, age 6–12 years, using a standardised language assessment and parental reports of everyday social and communicative behaviours. Their profiles were compared to those of typically developing sighted children of similar age and verbal ability. Results: Compared to their sighted peers, and relative to their own good and potentially superior structural language skills, children with VI showed significantly poorer use of language for social purposes. Pragmatic language weaknesses were a part of a broader socio-communicative profile of difficulties, present in a substantial proportion of these children and consistent with the pattern found in sighted children with autism. Conclusions: There are ongoing socio-communicative and pragmatic language difficulties in children with congenital VI at school age, despite their good intellectual abilities and advanced linguistic skills. Further research is required to unpack the underlying causes and factors maintaining this vulnerability in such children. Keywords: Visual impairment, language, social communication, autism.

Congenital visual impairment (VI) has been associated with vulnerabilities in socio-communicative and socio-cognitive development, including behavioural similarities with sighted children with autism (Pring, 2005). Difficulties have been reported in early social interaction and communicative competence (Preisler, 1991; Urwin, 1983), theory of mind (Green, Pring, & Swettenham, 2004; Peterson, Peterson, & Webb, 2000; Roch-Levecq, 2006) and emotional expressiveness and recognition (Dyck, Farrugia, Shochet, & Holmes-Brown, 2004; Minter, Hobson, & Pring, 1991; Roch-Levecq, 2006).

The underlying reasons for such difficulties in children with VI are unknown. Similar to sighted children with autism, their adverse outcomes in social communication and social cognition have been attributed to disruptions in visually guided experiences (e.g., dyadic and triadic joint attention) and visual behaviours (e.g., eye-gaze monitoring, directing and following) in early childhood, which are seen as precursor milestones for subsequent social development (Hobson, 1993). However, very little is known about the non-visual aspects of early social interaction and social attention that are not eye-gaze dependent, although some recent empirical advances in understanding the mechanisms of attention in a social context in young children with congenital VI have occurred (Tadić, Pring, & Dale, 2009).

Importantly, evidence suggests individual variation and greater success in social communication in some children with VI (Preisler, 1991; Urwin, 1978). Language may be a developmental domain which provides children with VI with alternative non-visual strategies for social development (Landau & Gleitman, 1985; Pérez-Pereira & Conti-Ramsden, 1999). For instance, research shows that children with VI who pass a standard theory of mind task have significantly higher verbal IQs (VIQ) and verbal mental ages than do those who fail it (Green et al., 2004; Minter, Hobson, & Bishop, 1998). Similarly, Brown, Hobson, Lee, and Stevenson (1997) found that children with VI who had higher verbal ability (VIQ > 70) showed fewer autistic-like socio-communicative difficulties on an autism rating checklist than did those with lower verbal ability (VIQ < 70). However, language-based measures are commonly used to assess general intelligence in children with VI, making it more difficult to isolate the contribution of language irrespective of general cognitive ability. Studies have been erratic in their inclusion of children with varying intellectual levels. Thus, it is necessary to consider the presentation of language in children with VI where cognitive ability has been controlled.
for and where a systematic comparison with a well-controlled sighted sample is appropriate.

Language comprises a rich network of functions and skills that provide building blocks for communicative and social interaction. While structural language skills (i.e., articulation of speech, use of grammar and vocabulary) may enable a fluent conversation, pragmatic language skills (i.e., using language socially and appropriately in a given context) are required for successful socio-communicative functioning. Specific language disorders provide evidence for a possible dissociation between different language functions. For instance, children with pragmatic language impairment (PLI) typically experience difficulties with use of semantics and pragmatic language, compared to their generally intact verbal fluency and adequate syntax and phonology (Norbury, Nash, Baird, & Bishop, 2004).

Vision is implicated in general language development, as visually driven joint attention experiences in early childhood provide a framework within which language learning occurs (Morales, Mundy, & Rojas, 1998; Tomasello & Farrar, 1986). However, despite some early delays and irregularities in the early vocabulary acquisition and production, syntactic knowledge, and semantic concepts acquisition (Andersen, Dunlea, & Kekelis, 1984; McConachie & Moore, 1994), the structural language skills of children with VI can develop without major concern (Landau & Gleitman, 1985; Mulford, 1988; Urwin, 1983), and any early difficulties appear to resolve and are generally overcome by school age (Landau & Gleitman, 1985; Mills, 1993; Mulford, 1988). The picture is less clear regarding their pragmatic language use, which may have features similar to that of children with PLI (Mills, 1993) and autism (Hobson & Bishop, 2003). These involve the extensive, and sometimes inappropriate, use of questions, an absence of communicative gestures, and the extensive use of imitative speech, repetitions and verbal routines in children with VI (Mulford, 1983; Norgate, Collis, & Lewis, 1998; Preisler, 1991). It has been argued that such characteristics of language use in children with VI may have an important function in promoting their cognition and social interaction by providing an adaptive strategy by which to gather information, analyse speech, reduce memory load, and avoid isolation (see Pérez-Pereira & Conti-Ramsden, 1999). However, concern has been raised regarding these language features in children with VI (e.g., stereotypic speech and echolalia), which are often seen in children with autism and thus may contribute to the autism-like presentation of some children with VI (Brown et al., 1997).

Based on the aforementioned studies, most of which involved preschool children with congenital VI, structural language appears to be an area of relative strength for such children, whereas their pragmatic skills may be challenged. Irregular presentation of language skills in children with congenital VI has been illustrated recently in an uncontrolled study using a parental communication checklist with a small school-age sample (James & Stojanovik, 2007). However, the discrepancy between structural and pragmatic language in children with VI needs to be further substantiated by research.

Overall, little is known about the presentation of language in children with congenital VI in middle childhood. Our understanding of the nature of autism-like socio-communicative difficulties seen in such children is still in its infancy, and the prevalence of learning difficulties continues to impose complex methodological constraints. Empirical attempts to enhance this understanding remain a major challenge, given the rarity of children with congenital VI and the need to adapt assessment procedures to their available sensory channels. The aim of this study was to examine variation in language presentation and social communication in school-age children with congenital VI, while controlling for general intelligence. To achieve this we i) focused on children with a significant congenital vision loss who had intelligence in the normal range, ii) utilised a comparison group of typically developing sighted children of similar age and verbal ability, iii) used a standardised test designed specifically to assess language function, and iv) utilised a parental report of everyday language and communicative behaviours. We compared the language and socio-communicative profiles of children with VI and sighted children with a view to examining the extent to which the two groups differ, and to gain better appreciation of specific strengths and weaknesses that may characterise the VI group.

Method

Participants

The participants were 15 children with congenital VI and 26 sighted children.

The children with congenital VI were recruited through the Developmental Vision Clinic at the Great Ormond Street Hospital, London, UK to which children with vision loss of heterogeneous aetiology and severity are referred for developmental and functional vision assessments in their preschool years. A consecutive clinic sample of children with a significant congenital vision loss (i.e., profound – no form vision, light perception or worse; or severe – severely degraded form vision), which was of peripheral, not cerebral origin (i.e., without an additional paediatric diagnosis involving the central nervous system) (see Sonksen & Dale, 2002), was identified for the purposes of longitudinal research. Informed parental consent for participation in this study was sought for those children who were age 6–12 at the time. Of 20 children for whom parental consent was obtained, 15 children with a verbal IQ (VIQ) > 80 and no additional impairments were eligible for inclusion in this research. Their VI severity was assessed by...
the paediatrician of the clinic at regular intervals throughout their preschool years (see Appendix A for individual diagnoses and vision levels). All but one were Braille readers at the time of this research.

A comparison group of sighted children was recruited through local primary schools in the South East of London and Kent, UK. Of 32 children for whom parental consent was obtained, and who were subsequently assessed, the 26 most closely resembling the VI group in terms of age and VIQ were included. We excluded children from sibling pairs.

The final groups were therefore comparable on (t_{39} = -.105; \ p = .917), age (t_{39} = -.502; \ p = .618), verbal mental age (VMA) (t_{39} = -.672; \ p = .506), and gender distribution (\chi^2 (1) = .702; \ p = .754) (Table 1). The majority of all children were from White British majority ethnic background (no between-group differences: \chi^2 (1) = 1.036; \ p = .309). However, the insufficient data available on socio-economic status precluded us from drawing conclusions with regards to this variable.

Materials

The Verbal scale from the Wechsler Intelligence Scales for Children-III (WISC-III; Wechsler, 1992) was used to assess developmental level. Each child’s VIQ and VMA were derived from five verbal subtests that require no presentation of visual stimuli: Information, Similarities, Vocabulary, Comprehension and Digit Span.

The Clinical Evaluation of Language Fundamentals-3 (CELF-3; Semel, Wiig, & Secord, 2000) was used to assess language function. Only two core and two supplementary subtests were suitable for use with children with VI, as they do not use visual stimuli. These were two receptive language subtests (i.e., Word Classes – semantic word grouping; and Listening to Paragraphs – verbal recall, comprehension and interpretation) and two expressive language subtests (i.e., Recalling Sentences – verbal short-term memory; and Word Associations – word fluency). Typically, Expressive, Receptive and Total Language composites can be derived based on the six core subtests. Instead, we derived a Total Language score for each child, based on the sum of the four scaled scores obtained. This was so as to obtain an index of language ability separate from VIQ. However, the incomplete CELF-3 assessment may have reduced reliability.

The Children’s Communication Checklist-2 (CCC-2; Bishop, 2003) was used to evaluate every-day language and communicative skills. This 70-item parental questionnaire taps a wider range of language functions, such as structural language skills (i.e., Speech and Syntax scales) and meaningful use of language (i.e., Semantics and Coherence scales), as well as pragmatic language use (i.e., Inappropriate Initiation, Stereotyped Language, Use of Context, and Non-Verbal Communication scales). The questionnaire also includes scales assessing social behaviours that are typically impaired in cases of autism spectrum disorders (ASD) (e.g., Social Relationships and Interests scales). The CCC-2 is not diagnostic, although it can be useful in screening for a potential communication disorder (e.g., ASD and specific language impairment – SLI). The General Communication Composite (GCC) is used to identify children likely to have clinically significant communication problems, whereas the Social Interaction Deviance Composite (SIDC) can help identify children in whom pragmatic language and social interaction skills are disproportionately impaired relative to their structural language. The CCC-2 item 14 from the Non-Verbal Communication scale (i.e., ‘does not look at the person s/he is talking to’) was removed from analyses for both groups here, as it was omitted by more than half of the parents of children with VI.

The Social Communication Questionnaire (SCQ) (Lifetime Autoscore) (Rutter, Bailey, & Lord, 2003) was used to screen for socio-communicative behaviours associated with autism (scores of ≥15 are considered potentially clinically significant), and which map onto the three core diagnostic domains: Reciprocal Social Interaction, Communication and the Restricted, Repetitive and Stereotyped Behaviours.

Procedure

The verbal tests were presented in the same order for all children, the WISC-III subtests followed by the CELF-3, and in the order specified in the test manuals. All questionnaires were completed by parents, except for two children with VI for whom these were completed by a teacher who knew them well.

The study, including the recruitment with informed parental consent, conduct towards the participants and study procedure, was carried out following the research protocol approved by the NHS research ethical committee for the UCL Institute of Child Health and Great Ormond Street Hospital, and the ethical committee for Goldsmiths, University of London.

Results

Data screening did not suggest any serious violation of assumptions (e.g., normality). Box-plots revealed only one extreme outlier in the sighted group on the CELF-3 Word Associations and the inclusion or removal of this case did not affect the results.

**Table 1** Demographic characteristics – group means and SDs

<table>
<thead>
<tr>
<th>Comparison criteria</th>
<th>VI</th>
<th>Sighted</th>
<th>p level</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean (SD)</td>
<td>(N = 15)</td>
<td>(N = 26)</td>
<td></td>
</tr>
<tr>
<td>VIQ</td>
<td>105.9 (10.7)</td>
<td>106.3 (11.1)</td>
<td>n.s.</td>
</tr>
<tr>
<td>Range</td>
<td>84–128</td>
<td>80–130</td>
<td></td>
</tr>
<tr>
<td>VMA</td>
<td>108.2 (21.8)</td>
<td>113.3 (24.8)</td>
<td>n.s.</td>
</tr>
<tr>
<td>Range</td>
<td>103.1 (23.0)</td>
<td>106.5 (20.3)</td>
<td>n.s.</td>
</tr>
<tr>
<td>Mean in months (SD)</td>
<td>6:06–12:11</td>
<td>6:02–11:11</td>
<td></td>
</tr>
<tr>
<td>Age</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Note: n.s.: not significant.

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The skills required for the two tests may not be
independent.

VI was Recalling Sentences (the only individual subtest superior for children with
Classes (38.9) = 1.702; t(39) = 1.742; p = .097) and Word Associa-
tions (t(38.9) = 1.702; p = .097) and Word Associations (t(39) = 1.256; p = .217).

Overall performance on the two standardised tests (CELF-3 Total Language and WISC-III VIQ) was sig-
ificantly correlated in both groups (VI: r = .596; p = .019; Sighted: r = .703; p ≤ .001), signifying that
the skills required for the two tests may not be independent.

Table 2 CELF-3 – group means and SDs

<table>
<thead>
<tr>
<th>Measure</th>
<th>VI</th>
<th>Sighted</th>
<th>p level</th>
</tr>
</thead>
<tbody>
<tr>
<td>CELF-3 (scaled scores)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Receptive language subtests</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Word Classes</td>
<td>11.4 (2.4)</td>
<td>10.04 (2.4)</td>
<td>n.s.</td>
</tr>
<tr>
<td>Listening to</td>
<td>9.9 (1.5)</td>
<td>8.8 (2.6)</td>
<td>n.s.</td>
</tr>
<tr>
<td>Paragraphs</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Expressive language subtests</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Recalling Sentences</td>
<td>12.6 (2.7)</td>
<td>9.8 (3.1)</td>
<td>**</td>
</tr>
<tr>
<td>Word Associations</td>
<td>11.9 (3.5)</td>
<td>10.7 (2.6)</td>
<td>n.s.</td>
</tr>
<tr>
<td>Total Language (sum of 4 scaled scores)</td>
<td>45.9 (8.2)</td>
<td>39.4 (6.6)</td>
<td>**</td>
</tr>
</tbody>
</table>

Note: n.s.: not significant; ** significant at p ≤ .01.

Parental reports

Table 3 shows the group means and SDs on the
CCC-2 (scaled scores) and the SCQ (raw scores). First, a Profile Analysis was used to compare the
groups’ profiles on the CCC-2. Subsequently, the test
of Flatness showed significantly different within-
group performance (i.e., non-flat profiles) across the
CCC-2 scales (Pillai’s Trace: F(9,29) = 5.323; p ≤ .001). Additionally, the test of Levels indi-
cated significant between-group differences when
children’s scores were averaged across the CCC-2 scales (F(1,37) = 26.6; p ≤ .001). A significant test of
Parallelism showed distinguishable (i.e., unparallel)
profiles between the two groups across the CCC-2 scales (Pillai’s Trace: F(9,29) = 7.266; p ≤ .001).

Figure 1 illustrates the divergent CCC-2 profiles of
the two groups. The sighted children’s profiles were
in line with the CCC-2 developmental norms, falling
around the mean scaled score of 10. Despite the
irregularity of profiles in the VI group, mean scores
across the CCC-2 scales remained largely within
normal range limits (i.e., scaled score ≥ 6), except for
the Non-Verbal Communication and Social Relations
scales, where mean scores fell below the normal
range (i.e., scaled score < 6).

Following from the results of the Profile Analysis,
the between-group difference on individual CCC-2
scales were examined, using a corrected alpha level
of p = .01 (adjusted for 10 comparisons). Children
with VI were comparable to sighted children on the
structural language scales assessing articulation of
Speech (t(37) = −.401; p = .691) and Syntactic
knowledge (t(16.99) = −1.250; p = .228). However,
they obtained significantly poorer parental ratings
on the scales assessing language Semantics
(t(37) = −2.717; p = .01), as well as on the four scales
assessing pragmatic language use (Inappropriate
Initiation: t(37) = −3.838; p ≤ .001; Stereotyped Lan-
guage: t(37) = −3.18; p ≤ .003; use of Context: t(37) =
−5.105; p ≤ .001; and Non-Verbal Communication:

Table 3 CCC-2 and SCQ – group means and SDs

<table>
<thead>
<tr>
<th>Measure</th>
<th>VI</th>
<th>Sighted</th>
<th>p level</th>
</tr>
</thead>
<tbody>
<tr>
<td>CCC-2 (scaled scores)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Speech</td>
<td>9.9 (3.5)</td>
<td>10.3 (2.6)</td>
<td>n.s.</td>
</tr>
<tr>
<td>Syntax</td>
<td>9.6 (3.6)</td>
<td>10.9 (1.9)</td>
<td>n.s.</td>
</tr>
<tr>
<td>Semantics</td>
<td>8.6 (3.1)</td>
<td>11.1 (2.7)</td>
<td>**</td>
</tr>
<tr>
<td>Coherence</td>
<td>8.7 (3.3)</td>
<td>11 (2.5)</td>
<td>n.s.</td>
</tr>
<tr>
<td>Inappropriate Initiation</td>
<td>7.5 (3.1)</td>
<td>11 (2.6)</td>
<td>**</td>
</tr>
<tr>
<td>Stereotyped Language</td>
<td>6.6 (3.6)</td>
<td>10.3 (3.3)</td>
<td>**</td>
</tr>
<tr>
<td>Use of Context</td>
<td>6.5 (2.4)</td>
<td>11.2 (2.9)</td>
<td>**</td>
</tr>
<tr>
<td>Non-Verbal Communication</td>
<td>4.3 (2.6)</td>
<td>10.9 (2.7)</td>
<td>**</td>
</tr>
<tr>
<td>Social Relations</td>
<td>5.4 (2.8)</td>
<td>10.4 (2.9)</td>
<td>**</td>
</tr>
<tr>
<td>Interests</td>
<td>6.5 (2.5)</td>
<td>9.5 (2.9)</td>
<td>**</td>
</tr>
<tr>
<td>General Communication Composite/GCC</td>
<td>61.8 (18.8)</td>
<td>86.8 (14)</td>
<td>**</td>
</tr>
<tr>
<td>SCQ</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total raw score</td>
<td>14.3 (3.9)</td>
<td>4.4 (3.9)</td>
<td>**</td>
</tr>
</tbody>
</table>

Note: n.s.: not significant; ** significant at p ≤ .01.
The between-group difference on language Coherence failed to reach significance at the adjusted alpha level ($t_{37} = -2.404; \ p = .021$). The VI group obtained significantly lower ratings on scales associated with autistic presentation (Social Relationships: $t_{37} = -5.17; \ p \leq .001$; and Interests: $t_{37} = -3.15; \ p = .003$) (see Appendix 2 for an illustrative breakdown of everyday behaviours on the CCC-2).

The sighted group achieved significantly higher General Communication Composite/GCC scores than the VI group, signifying their higher communicative skills ($t_{37} = -5.105; \ p \leq .001$). A between-group comparison on the Social Interaction Deviance Composite/SIDC was not considered useful as this provides qualitative information about the pattern of impairment at an individual child level and cannot be interpreted without the GCC (Bishop, 2003). Thus, the relationship between the two composites has been considered qualitatively for each child.

In relation to this, three reference lines plotted in Figure 2 indicate the clinical cut-off points used to subgroup children with specific communication difficulties on the CCC-2. According to Bishop (2003), the top-right section of the scatter-plot (i.e., GCC ≥ 55 and SIDC > −15) marks the distribution of profiles associated with children without suspected socio-communicative difficulties (typically developing range). The bottom-left section of the scatter-plot (i.e., GCC < 55 and SIDC < 0) marks a region of profiles which are frequently seen in autism spectrum disorders (ASD) (i.e., both composites are below normal range, and there is a significant mismatch between structural language and social/pragmatic communication skills).
skills). Finally, the bottom-right section of the scatter-plot highlights the profiles of children whose social/pragmatic skills are disproportionately low (SIDC ≤ −15), relative to their normal-range GCC score (GCC ≥ 55). Such profiles are frequently seen in Asperger syndrome (AS).

Figure 2 shows that the CCC-2 communicative profiles of all children in the sighted group were within the normal range. Profiles of five children with VI were also within this range. However, four children with VI showed CCC-2 profiles resembling those of children with ASD, and five showed profiles usually associated with children with AS. Overall, 64% of children with VI showed CCC-2 socio-communicative characteristics consistent with the broader autism spectrum.

In line with the CCC-2 pattern, children with VI obtained significantly higher total raw scores on the SCQ – indicative of lower socio-communicative outcome – than sighted children ($t_{\text{aw}} = 7.727; p ≤ .001$). Five children with VI (34%) obtained SCQ scores above clinical cut-off for potential autism concern. Four of these children were also within range for clinical concern of a potential communication disorder on the CCC-2. Additionally, a number of other children with VI achieved overall SCQ scores that were just below the clinical cut-off of 15. The profiles of three of these children fell within the CCC-2 section associated with AS.

This degree of overlap between SCQ and CCC-2 was expected due to the underlying social and communicative constructs of the two measures. In line with this were the significant correlations between the children's SCQ scores and their scaled scores on the Social and Non-Verbal Communication scales on the CCC-2 (VI: $r = −.631$, $p = .015$ and $r = −.653$, $p = .011$; Sighted: $r = −.469$, $p = .018$ and $r = −.580$, $p = .002$). This suggests some degree of consistency between the parental reports.

Although the potentially confounding effects of variation in severity of VI could not be examined statistically due to small numbers, scrutiny of the data revealed that clinically elevated scores on the two socio-communicative measures were not confined to the group of children whose VI was of greater severity, but were also seen in children with some limited levels of functional vision. Additionally, the prevalence of autistic-like behaviours in the VI group could not be explained by differences in verbal competence. The overall performance scores on the standardised language measures (VIQ WISC-III and CELF-3 Total Language Sum) did not correlate with any of the parental questionnaire scores (SCQ raw score and the CCC-2 scaled scores) for either the VI or sighted group ($p$ values > .05). Examination of individual characteristics of those children whose socio-communicative profiles scores fell in the normal range did not reveal any insight with respect to this individual variation (e.g., see Appendix A).

### Discussion

The present study highlights three key findings. Despite being comparable on age and verbal intelligence, children with VI performed significantly better than sighted children on a standardised test of language function. By contrast, children with VI showed significantly poorer socio-communicative skills than their sighted peers, based on parental reports, with particular weakness in the use of language for pragmatic and social purposes. Finally, judging by the parental reports, a substantial proportion of children with VI showed a level of socio-communicative difficulties consistent with the broader autism spectrum in sighted children.

The finding that children with VI outperformed sighted children on structural language is surprising, particularly in light of early difficulties cited in literature (Andersen et al., 1984; McConachie & Moore, 1994). However, the language domain is more salient to children with VI than sighted children (Pérez-Pereira & Conti-Ramsden, 1999), and thus may serve subtly different functions for the two groups. The CELF-3 may have, to an extent, allowed us to separate this language function from general intelligence. Being a test of language ability, it is not surprising to find that CELF-3 performance correlated with verbal IQs as assessed by the WISC-III (see also Semel et al., 2000). However, unlike the verbal WISC-III subtests, which essentially measure crystallised intelligence and ‘fluid’ reasoning, the CELF-3 is less reasoning-based, largely tapping linguistic elements such as content and structure, which can be evaluated independently and context-free. This potential of the CELF-3 to isolate children’s language-specific strengths and difficulties has also been demonstrated in research with other clinical groups (e.g., SLI and autism) (Lloyd, Paintin, & Botting, 2006).

Given that the CELF-3 assessment here was based only on four (i.e., non-visual) out of the six possible subtests, it is possible that this combination of auditory subtests was especially favourable to children with VI. The group’s superior performance on Recalling Sentences in particular may reflect a short-term memory advantage, which has traditionally been demonstrated using Digit Span (Hull & Mason, 1995; Smits & Mommers, 1976). Thus, good verbal short-term memory may boost an overall language outcome and explain the superior CELF-3 performance of the VI group.

Furthermore, it is possible that the achieved language competence in children with VI is dependent upon the assessment context, as the parental ratings of specific language skills (e.g., CCC-2 Semantics) revealed a potentially contrasting picture. These children may have benefited from the structured context of a traditional one-to-one assessment, which may provide scaffolding for
successful performance and may better capture the strength that is not necessarily apparent in an everyday conversation with children with VI. Conversely, parental reports of everyday language skills (which are used in an inherently social context) may be more likely to reveal a VI-related disadvantage in meaningful use of language than would a standardised assessment.

In contrast to robust structural language skills, parental reports also captured a particularly striking weakness in the pragmatic language of children with VI, mirroring some earlier research (Hobson & Bishop, 2003; Mulford, 1983; Preiser, 1991). Here, we demonstrate that pragmatic language difficulties are present at school age even in those children with VI who are linguistically advanced, highlighting the non-verbal aspects of pragmatics as a particular challenge.

It is possible that parental ratings on the CCC-2 were negatively biased towards the children with VI. This questionnaire has not been developed with such children in mind, potentially over-emphasising their inherent developmental weaknesses (Norbury et al., 2004). However, poorer socio-communicative outcomes of these children may reflect the developmental – as well as methodological and theoretical – challenge of separating pragmatic language skills from behaviours supported by vision. The non-verbal aspects of pragmatics, such as use of facial expressions and gestures, may be the most potent communicative tools for maintaining a conversational partner’s focus of attention. Similarly, the ability to initiate conversations appropriately, to understand irony and sarcasm, and to adjust conversational topics based on others’ levels of interest may be more easily achieved through monitoring of the conversational partner’s facial expressions and gestures. Thus, this visual basis of pragmatic language may explain why pragmatic language fails to benefit from scaffolding in the same way as does structural language (i.e., it may rely more heavily on joint attention, which itself is vulnerable in these children). This is in line with the patterns observed in autism, as even high-functioning children with autism, who show better language outcomes, sustain poor socio-pragmatic language (Dennis, Lazenby, & Lockyer, 2001; Klin, 2000).

The uneven CCC-2 profiles of children with VI, marked by a disproportionate weakness in social and pragmatic skills relative to spared structural language, are in fact reminiscent of the presentation of autism in sighted children. A substantial proportion of children with VI here reached or exceeded clinical cut-off scores for autism concern on both parental checklists. These findings support previous research raising concern about the alarming prevalence of autism-like features in children with congenital VI of different ages (Brown et al., 1997; Cass, Sonksen, & McConachie, 1994; Hobson, Lee, & Brown, 1999). Brown et al., for instance, reported autism-like clinical features across a cognitively heterogeneous population of preschoolers and primary school-aged children with congenital VI, although these were significantly more prominent in children with learning difficulties. The present research provides additional insight, revealing similar vulnerabilities in an intellectually homogeneous group of children with VI and advanced language skills, some of whom had above average verbal IQs. Additionally, judging by the CCC-2 outcomes, the study also highlights close similarities to children with PLI who, despite spared structural language, show difficulties with semantic and pragmatic language use (Norbury et al., 2004). Sighted children with high-functioning autism and good language outcome, as well as children with PLI, may provide useful comparison groups in further research, to help unpack the reasons for the irregular language presentation and socio-communicative difficulties seen in verbally proficient children with congenital VI.

While Brown et al. (1997) investigated autistic-like characteristics exclusively in children with total sight loss, the present research provides unique evidence that such characteristics remain prevalent amongst children with some, albeit severely degraded, limited form vision. However, it is still unclear why children with varying degrees of VI are at risk of adverse socio-communicative outcomes. One explanation is that significant vision loss in early childhood may impose seriously limited opportunities to engage in that very special form of relatedness, affective sharing and perspective taking provided in the context of triadic joint attention with an interactional partner, which may create a developmental vulnerability with possible long-term consequences (including autistic-like behaviours) (Hobson, 1993). This vulnerability may explain why difficulties persists even in middle childhood and why even those children with VI whose socio-communicative profiles fall within normal range limits did not reach the levels of socio-communicative competence typical of the majority of sighted children. Another explanation is that VI may interact with an inherent neurological susceptibility and adverse environmental circumstances at a critical stage of development (Cass et al., 1994; Dale & Sonksen, 2002). Individual variation within the VI sample is suggestive evidence for a multi-factorial aetiology. Despite good language skills and verbal intelligence, some children with VI presented more strongly with autistic-like behaviours than did others.

Although utilising additional structured measures and direct clinical assessments to complement the parental questionnaires would have been useful, the existing clinical and observational measures targeting autism-related socio-communicative difficulties are not developed for children with VI and are likely to be less sensitive to their developmental strengths.
This emphasises the need for VI-specific measures to be developed. With this in mind, parents are a valuable source of knowledge about their children. Although not diagnostic, parental reports can provide a window into their children’s characteristics that may not otherwise be easily evaluated, as it has been achieved here. Further research is required to establish the developmental origin and maintaining factors of socio-communicative vulnerabilities in children with congenital VI. Greater insight into these aspects may provide a platform for potential preventative and habilitative interventions that can assist their socio-communicative and pragmatic development.

Key points

- Children with congenital VI have been associated with vulnerable socio-communicative outcomes, including resemblances to sighted children with autism.
- The current study demonstrates that, even with intact verbal intelligence and superior structural language skills, children with congenital VI are at risk of socio-communicative difficulties, with an ongoing concern at primary school age.
- Pragmatic language skills of such children seem to be disproportionately impaired compared to their strengths in structural language.
- Further research, with a focus on individual variation and the development of suitable measures, is necessary to establish the developmental cause and maintaining factors of these vulnerabilities in children with congenital VI.

References


Acknowledgements

The authors thank the children and their families who participated in this research. The research was supported by a 1 + 3 ESRC studentship (PTA-031-2004-00211) awarded to the first author while completing a PhD at Goldsmiths, University of London.

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Appendix A: Clinical and demographic characteristics of individual children with VI

<table>
<thead>
<tr>
<th>N</th>
<th>Age</th>
<th>Gender</th>
<th>Visual diagnosis</th>
<th>VI level</th>
<th>VIQ</th>
<th>SCQ raw (clinical cut-off ≥ 15)</th>
<th>CCC-2 profile</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>6:08</td>
<td>M</td>
<td>Familial exudative vitreo-retinopathy/Norrie’s Syndrome</td>
<td>Severe</td>
<td>109</td>
<td>9</td>
<td>Normal range</td>
</tr>
<tr>
<td>2</td>
<td>7:00</td>
<td>F</td>
<td>Leber’s amaurosis</td>
<td>Profound</td>
<td>110</td>
<td>14</td>
<td>AS-like</td>
</tr>
<tr>
<td>3</td>
<td>9:02</td>
<td>M</td>
<td>Bilateral microphthalmia and optic nerve aplasia</td>
<td>Profound</td>
<td>100</td>
<td>23</td>
<td>ASD-like</td>
</tr>
<tr>
<td>4</td>
<td>7:00</td>
<td>M</td>
<td>Bilateral optic nerve hypoplasia</td>
<td>Profound</td>
<td>101</td>
<td>12</td>
<td>ASD-like</td>
</tr>
<tr>
<td>5</td>
<td>12:11</td>
<td>F</td>
<td>Bilateral optic nerve hypoplasia</td>
<td>Profound</td>
<td>99</td>
<td>9</td>
<td>Data missing</td>
</tr>
<tr>
<td>6</td>
<td>8:03</td>
<td>F</td>
<td>Bilateral microphthalmia</td>
<td>Profound</td>
<td>99</td>
<td>12</td>
<td>Normal range</td>
</tr>
<tr>
<td>7</td>
<td>11:04</td>
<td>M</td>
<td>Bilateral aniridia and glaucoma</td>
<td>Severe</td>
<td>113</td>
<td>14</td>
<td>AS-like</td>
</tr>
<tr>
<td>8</td>
<td>6:06</td>
<td>F</td>
<td>Leber’s amaurosis</td>
<td>Severe</td>
<td>128</td>
<td>19</td>
<td>AS-like</td>
</tr>
<tr>
<td>9</td>
<td>9:11</td>
<td>M</td>
<td>Bilateral microphthalmia with multiple corneal opacities</td>
<td>Profound</td>
<td>108</td>
<td>13</td>
<td>ASD-like</td>
</tr>
</tbody>
</table>

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Appendix A: (continued)

<table>
<thead>
<tr>
<th></th>
<th>Age</th>
<th>Gender</th>
<th>Visual diagnosis</th>
<th>VI level</th>
<th>VIQ</th>
<th>SCQ raw (clinical cut-off ≥ 15)</th>
<th>CCC-2 profile</th>
</tr>
</thead>
<tbody>
<tr>
<td>10</td>
<td>8:05</td>
<td>F</td>
<td>Multiple opacities and sclerocornea</td>
<td>Severe</td>
<td>119</td>
<td>13</td>
<td>AS-like</td>
</tr>
<tr>
<td>11</td>
<td>8:07</td>
<td>M</td>
<td>Leber’s amaurosis</td>
<td>Severe</td>
<td>100</td>
<td>16</td>
<td>Normal range</td>
</tr>
<tr>
<td>12</td>
<td>7:03</td>
<td>F</td>
<td>Persistent primary hyperplastic vitreous and sclerocone</td>
<td>Severe</td>
<td>113</td>
<td>16</td>
<td>ASD-like</td>
</tr>
<tr>
<td>13</td>
<td>8:01</td>
<td>F</td>
<td>Bilateral microphthalmia and sclerocornea</td>
<td>Severe</td>
<td>95</td>
<td>20</td>
<td>AS-like</td>
</tr>
<tr>
<td>14</td>
<td>10:11</td>
<td>F</td>
<td>Leber’s amaurosis</td>
<td>Severe</td>
<td>84</td>
<td>12</td>
<td>Normal range</td>
</tr>
<tr>
<td>15</td>
<td>6:06</td>
<td>F</td>
<td>Leber’s amaurosis</td>
<td>Severe</td>
<td>111</td>
<td>12</td>
<td>Normal range</td>
</tr>
</tbody>
</table>

Appendix B: An illustrative vignette of everyday behaviours shown by a child with VI, as assessed by the CCC-2 scales

Here, we describe behaviours of a 7-year-old girl with congenital and profound VI (ID number 2, Appendix A). Her mother rates her highly, in terms of her ability to speak very clearly and fluently (Speech), and her use of long and complex sentences (Syntax) and abstract words to refer to general concepts (Semantics). She is able to talk very clearly about past events and plans in the future (Coherence). However, she often talks repetitively about things that other people are not interested in, telling them things that they already know, and asking questions even though she has been given an answer (Inappropriate Initiation). She has a tendency to include over-precise information in conversation (e.g., telling date and time if asked when she had been on a holiday) (Stereotyped Language) and misses the point of jokes (Context). She tends to appear blank in situations where other children would show clear facial expression, e.g., when happy or angry (Non-Verbal Communication). However, her mother reports that, despite her frequent tendency to shift conversations to favourite topics, even when others do not seem to be interested in them (Interests), people generally have enjoyable, interesting conversations with her (Stereotyped Language). According to her mother, this child shows a good repertoire of socio-interactive behaviours. For instance, she shows concern about others when they are upset, and often talks about her friends, showing interest in what they do (Social Relationships), which are the types of behaviours some of her VI peers in this study were reported to have difficulties with.