Adaptive behaviour and quality of life in school-age children with congenital visual disorders and different levels of visual impairment

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Abstract

Background: Adaptive behaviours are vital skills that allow individuals to function independently and are potentially amenable to behavioural interventions. Previous research indicated that adaptive behaviours are reduced in children and adolescents with severe to profound VI, but it was unclear if this was also the case for children with mild to moderate VI.

Aim: The aim of the study was to assess differences in adaptive behaviour in children with congenital visual disorders and different levels of visual impairment and their influence on quality of life and everyday strengths and difficulties.

Methods and Procedures: Questionnaires about adaptive behaviour, strengths and difficulties, and quality of life were completed by parents of school-age children with severe-to-profound VI (S/PVI, n=9, 0.9 logMAR – light perception only), mild-to-moderate VI (MVI, n=9, 0.1-0.7 logMAR), or typical sight (control, n=18, -0.3-0.1 logMAR). Differences in questionnaire domains by the severity of VI and relationships between adaptive behaviour and quality of life were analysed in general linear models.

Outcomes and Results: The questionnaire ratings indicated reduced adaptive behaviour, more difficulties, and reduced quality of life in children with S/PVI compared to typically-sighted peers. Effects were smaller for children with MVI, but indicated a significant reduction in quality of life compared to typically-sighted children. The effect of visual impairment on quality of life in school was partially mediated by adaptive behaviour.

Conclusion and Implication: Severe congenital visual impairment affects adaptive behaviour in children with verbal abilities in the typical range. This effect is less pronounced in children with mild-to-moderate VI, but still impacts on quality of life, particularly in school.

Keywords: adaptive behaviour, adolescence, childhood, quality of life, visual impairment

What this paper adds:

- Adaptive behaviour and quality of life are affected by congenital visual impairment (VI) during mid-childhood
- The difficulties are most pronounced in children with the more severe to profound VI, with all degrees affected
- Adaptive behaviour is partially mediating the effect of VI on difficulties in school

Adaptive behaviour and quality of life in school-age children with congenital visual disorders and different levels of visual impairment

Between 3 and 4 in 10,000 children in the UK are born with a severe congenital visual impairment (VI) identifiable in the first year of life (Rahi, Cable, BCVISG, 2003). Whilst this incidence is low, the associated life-long burden for affected individuals, their families, and society can be high (Frick, Gower, Kempen, & Wolff, 2007). A potential reason is that congenital VI impacts on the acquisition of skills needed to respond appropriately to environmental demands across a range of contexts, so-called adaptive behaviour. Aspects of adaptive behaviour include practical, social, and conceptual function. For instance, this includes being able to keep one's belongings tidy, being polite to other people, and being able to tell the time (Harrison & Oakland, 2008). Children with visual impairment have been shown to have a range of differences which all potentially reflect and impact on practical, social and conceptual function. For instance, they show reduced independence due difficulties with motor skills and mobility (Metsiou, Papadopoulos, & Agaliotis, 2011; Papadopoulos, Metsiou, & Agaliotis, 2011a). They often have slower development of social cognition and a relatively large proportion displays behaviours akin to autism spectrum disorder, including stereotyped movement, echolalic speech, and lack of engagement with caregivers and peers (Brown, Hobson, Lee, & Stevenson, 1997; Jure, Pogonza, & Rapin, 2016, Jure et al., 2016). Cognitive differences have also been found, notably delayed acquisition of semantic and pragmatic language skills (Tadić, Pring, & Dale, 2010), deficits in spatial memory and spatial navigation (Schmidt, Tinti, Fantino, Mammarella, & Cornoldi, 2013), and difficulties with cognitive control and behavioural regulation (Bathelt, de Haan, Salt, & Dale, 2016).

However, children with congenital VI are a heterogeneous group, including considerable variation in visual function ranging from no vision to relatively mild reduction in visual acuity. Previous studies on adaptive behaviour in VI have been based on samples of children with more severe vision acuity reduction and impairment. Consequently, there is no information regarding children with mild to moderate forms of VI that could guide clinicians, practitioners, and families. The current study, therefore, set out to investigate adaptive behaviour in children with varying levels of congenital visual impairment. The study focused on children with 'potentially simple' congenital disorders of the peripheral visual system (Sonksen & Dale, 2002), with no known disorder of central brain structures according to the visual disorder diagnosis and therefore potentially isolated vision disorders. In children with additional brain defects, as is common in cerebral VI (Rahi et al., 2003), the likelihood of comorbid learning difficulties is greatly increased. This would pose a significant confound as any differences in adaptive behaviour may be potentially linked to the learning disability rather than to the impact of visual impairment as adaptive behaviour progresses developmentally across childhood and is normative (Ditterline, Banner, Oakland, & Becton, 2008). To further minimise the possibility of additional learning difficulties which can commonly occur in children with congenital VI (Alimovic, 2013), a sample of higher functioning children with normal range verbal intelligence (according to standardized age norms for the typically sighted population) were selected for the study. Adaptive behaviour, strengths and difficulties in everyday life, and quality of life were assessed using parent questionnaires commonly used in clinical practice. This study was part of a larger study where children had direct visual acuity and neuropsychological and neuroimaging assessments (Bathelt et al., 2016; Bathelt, Dale, & de Haan, 2017). The current analysis investigated if different levels of visual impairment were associated with reduced adaptive behaviour and if adaptive behaviour was related to everyday difficulties and to quality of life in children with visual impairment.

Participants & Methods

Participants

The assessments presented here were part of a wider study on the neural and cognitive sequelae of congenital VI during mid-childhood. This study was performed in accordance with the Declaration of Helsinki. The study was approved by Bloomsbury Research Ethics Committee - approval: 12/LO/0939. All parents, guardians or next of kin provided written informed consent and children provided verbal assent to participate in this study. A prospective cross-sectional study was undertaken with eighteen children with VI aged between 8 and 13 years. The rarity of 'potentially simple' congenital disorders of the peripheral visual system raised challenges for recruitment and sampling (Rahi et al., 2003; Sonksen & Dale, 2002). Children were therefore recruited through national specialist clinics at Great Ormond Street Hospital for Children NHS Foundation Trust and Moorfields Eye Hospital NHS Foundation Trust. Inclusion criteria were i) children with a primary ophthalmological diagnosis falling into the classification of 'potentially simple' congenital disorders of the peripheral system (CDPVS), that is disorders affecting the globe of the eye, retina, or anterior optic nerve up to the optic chiasm, with no known brain disorder indicated by the paediatric or ophthalmological diagnosis (Sonksen & Dale, 2002), ii) vision reduction from mild/moderate to severe/profound VI iii) between 8 and 13 years, iv) normal range verbal functioning (verbal IQ at the last assessment >75 or attending mainstream school at age-appropriate level), iv) fluent speaker in English. Parent participants who rated questionnaires on their child also had to be relatively fluent speakers in English. Children with indications of additional neurological or endocrine abnormalities, including motor or hearing impairment, in their clinical records were excluded. Recruitment was undertaken through initial identification through clinical databases of children who had previously attended a tertiary paediatric specialist clinic at the hospital research site in their infancy and preschool years and open recruitment call through voluntary agencies associated with VI. Sample characteristics are summarized in Table 1.

Control sample: Eighteen children with normal or corrected-to-normal vision were recruited through local advertisement to match according to age. Children in the control group had to attend mainstream school at the age-appropriate level and have no known neurological or psychiatric conditions and be fluent English speakers. Their parent rater of the questionnaires also had to have fluent English to fill in questionnaires. See Table 2 for descriptive statistics.

The experimenter (J.B.) was trained by a neurodisability paediatrician specialised in VI (Dr Alison Salt) to undertake the visual acuity assessments using the Sonksen logMAR test of Visual Acuity (Salt, Wade, Proffitt, Heavens, & Sonksen, 2007). For children, who were not able to see the largest items on the Sonksen logMAR test, the Near Detection Scale (Sonksen, Petrie, & Drew, 1991) was used to assess their basic level of detection vision. Severe/Profound VI (S/PVI) in this study is defined as limited form vision with logMAR above 0.8 (Snellen worse than 6/36) to no or light perception only (Near Detection Scale, points 0 and 1). Mild/moderate VI (MVI) is defined as reduced visual acuity with logMAR between 0.4 and 0.8 (Snellen 6/18-6/36). Two children in the current sample were diagnosed with mild/moderate VI, but had better visual acuity scores than the MVI cut-off. Their diagnoses indicate that these children had difficulties with visual function that may interfere with everyday life, but that may not be adequately characterized through acuity measurement alone, e.g. nystagmus.

There was no significant difference in the number of males vs females between the combined VI and control group (χ^2=0.22, p=0.635), nor in participant age (independent sample t-test: t(17)=0.53, p=0.598).

Verbal comprehension was assessed using verbal subtests of the Wechsler Intelligence Scale for Children 4th UK edition (WISC-IV). The administered subtests included all items of the Verbal Comprehension composite score (Vocabulary, Similarities, Comprehension). Two items were altered that required direct visual experience: the first practice item on the Similarities subtest which includes knowledge of colour was not administered. The Comprehension question that asks about a situation in which ‘you see thick smoke’ was changed to ‘you smell thick smoke’. These alterations were used for the whole sample, including the typically-sighted control group. All other items were administered verbatim according to the WISC-IV administration manual. There was no significant difference in verbal IQ scores between the groups (S/PVI: mean=98.58 SE=7.012; MVI: mean=103.25, SE=3.004; control: mean=113.17, SE=3.866; one-way analysis of variance: F(2,35)=2.46, p=0.099).

Table 1 about here

Table 2 about here

Questionnaire measures of adaptive behaviour, strengths & difficulties, and wellbeing

Parents filled in the questionnaire while children participated in other parts of the study. The levels of missing data are reported below; not all responses were available for all participants because parents missed out items on the questionnaire or decided to not fill in a questionnaire. The data inclusion criterion was set as not more than 20% missing or uncoded questionnaires or not more than 15% of an individual item missing per sample.

Adaptive behaviour: The Adaptive Behaviour Assessment Systems (ABAS-II) 2nd edition was administered (Harrison & Oakland, 2008). Parents rate the adaptive behavior on questions relating to 9 skill areas:

- Communication: speech, language, and listening skills required to communicate with other people
- Functional academics: basic reading, writing, and arithmetic and other academic skills required for daily independent functioning
- Self-direction: Skills needed for independence, responsibility, and self-control, e.g. completing tasks, keeping to a schedule, following directions and similar
- Leisure: Skills needed for engaging in and planning leisure activities, e.g. playing with others, following rules in a game and similar
- Social: Skills needed to interact socially and get along with others, e.g. having friends, showing and recognising emotions, using manners and similar
- Community Use: Skills needed for functioning in the community, including using community resources, shopping skills, using public transport and similar
- Home living: Skills needed for basic care of a home or living setting, e.g. cleaning, performing chores and similar

- Health and Safety: Skills needed for protection of health and to respond to illness and injury, e.g. following safety rules, using medicines, showing caution and similar.
- Self-care: Skills needed for personal care, e.g. eating, dressing, bathing, grooming, hygiene and similar

Strengths & difficulties: The UK version of the Strengths and Difficulties Questionnaire (SDQ) was administered (Goodman, 2001). This is a questionnaire designed to assess the presence of behavioural and emotional problems. The SDQ scoring website (http://www.sdqinfo.org/py/sdqinfo/c0.py, last visited 25/7/14) was used for scoring. Useable data was available for 17 children in the VI group and 18 children in the control group.

Quality of life: The Paediatric Quality of Life (PedsQL) questionnaire was administered (Varni, Seid, & Kurtin, 2001). This questionnaire is designed to assess a child's wellbeing in areas of physical, emotional, social, and school functioning. 17 parents of children in the VI group and 18 parents of children in the control group filled in the questionnaire.

Statistics

Differences between the three groups of children (S/PVI, MVI, typically-sighted) were investigated using analysis of variance (ANOVA) models. Follow-up contrasts were based on independent-sample t-tests. Bonferroni correction was used to adjust for multiple comparisons and corrected values are reported as p_corr throughout the manuscript. Effect size was expressed as Hedges' g. This metric can be interpreted similarly to Cohen's d, but is more suitable for small sample sizes (small effect: $\geqslant 0.2$, medium effect: $\geqslant 0.5$, large effect: $\geqslant 0.8$).

The association between adaptive behaviour and quality of life scores was investigated using a general linear model with adaptive behaviour as a predictor and vision group as a factor. Only children with VI were considered in this analysis, because the variation of quality of life ratings was limited and near ceiling level in the typically-sighted control group. The continuous variables met the normality assumption according to Shapiro-Wilk tests (all $p>0.05$) in the S/PVI and MVI subset.

Statistical analyses were carried out using the scientific python (SciPy) v0.18.1 and statsmodels v0.6.1 packages for Python. Visualizations were created using the matplotlib v1.5.1 package for Python.

Mediation analysis was carried out under R version 3.2.3 using the lavaan package v0.5 (Rosseel, 2012).

Results

Adaptive behaviour

Level of visual impairment (S/PVI and MVI versus controls) was compared with everyday adaptive behaviour, as measured by the Adaptive Behavior Assessment System (ABAS). A comparison of the composite scores indicated a significant effect of vision group on the General Adaptive Behaviour Composite (GAC) score (see Table 3 for descriptive statistics, 1-factor ANOVA: $F(2,31)=7.13$, $p=0.003$) with significantly lower scores in the S/PVI group compared to typically-sighted controls (see Table 3). There were also a significant effect of vision group for the Practical, Social, and Conceptual composite scores (2-factor ANOVA for scale and vision group - vision group effect: $F(2,93)=10.27$, $p<0.001$, see Table 3). Follow-up analysis indicated significantly lower scores in the Practical domain for children with S/PVI compared to controls (see Table 3).

Comparison across the individual questionnaire scales indicated a significant effect of vision group for the Functional Academic scale ($F(2,31)=6.79$, $p=0.004$, $p_{corr}=0.032$) with lower scores in the S/PVI group compared to typically-sighted controls (S/PVI: mean=5.91, SE=1.178; control: mean=11.00,SE=0.822; $t(25)=3.64$, $p=0.001$, $p_{corr}=0.033$, $g=1.508$) (see Figure 1). No other comparisons for the individual subscales showed significant differences.

Table 3 about here

Figure 1 about here

Strengths & Difficulties

Level of visual impairment (S/PVI and MVI versus controls) was compared with strengths and difficulties across domains of everyday function, as measured by the Strengths and Difficulties questionnaire (SDQ). Statistical comparison indicated a significant difference in the Total Difficulties score ($F(2,32)=5.41$, $p=0.009$) with significantly higher difficulties scores in the S/PVI and MVI group compared to typically-sighted controls (S/PVI: mean=8.30, SE=1.758; MVI: mean=9.25, SE=2.562; control: mean=3.29, SE=0.685; S/PVI vs control: $t(25)=-3.12$, $p=0.005$, $p_{corr}=0.014$, $g=-1.29$; MVI vs control: $t(23)=-2.99$, $p=0.006$, $p_{corr}=0.019$, $g=-1.34$). There was no significant difference in Total Difficulties scores between the S/PVI and MVI group ($t(16)=0.32$, $p=0.757$, $p_{corr}=1.0$, $g=0.16$). Comparison across the individual questionnaire scales indicated a significant effect of group for the Peer Relationship Problems scale ($F(2,32)=9.28$, $p<0.001$, $p_{corr}=0.003$) with lower scores in the S/PVI group compared to typically-sighted controls (S/PVI: mean=2.5, SE=0.511; control: mean=0.35, SE=0.171, $t(25)=-4.89$, $p<0.001$, p_corr=0.001, $g=-2.02$, see Figure 2).

Figure 2 about here

Quality of Life

Level of visual impairment (S/PVI and MVI versus controls) was compared with children's quality of life and wellbeing, as measured by the Paediatric Quality of Life (PedsQL). Statistical analysis indicated significant group effects for all PedsQL scales (see Table 4). Follow-up analysis with simple contrasts indicated lower ratings on the Physical, Social, and School scales for children with S/PVI or MVI compared to typically-sighted controls. Differences on the Emotional scale did not survive correction for multiple comparisons. There were no significant differences between the S/PVI and MVI group for any of the scales.

Table 4 about here

Relationship between ABAS-II and PedsQL scores

Next, the relationship between adaptive behaviour and quality of life ratings was investigated. For Physical aspects of quality of life (PedsQL-Phys) as the outcome, a regression model with ABAS-II general adaptive behaviour scores (GAC) indicated a significant fit with GAC scores explaining around 25% of variance in PedsQL-Phys ($F(1,31)=10.89$, $p<0.001$, $R^2=0.26$; GAC: $\beta=0.61$, $p=0.002$; $\beta_0=24.98$). Adding vision group (S/PVI, MVI) as an additional predictor improved the model fit and added around 20% of explained variance in ratings ($F(3,29)=11.65$, $p<0.001$, $R^2=0.55$, Adj.-$R^2=0.5$). Significant interactions between ABAS-II GAC scores and vision group were found for the S/PVI and MVI group (S/PVI: $\beta=-29.68$, $p<0.001$; MVI: $\beta=-24.02$, $p=0.005$). ABAS-II GAC scores were also associated with higher ratings for PedsQL Social and School scales (Social: $F(1,31)=16.10$, $p<0.001$, $R^2=0.34$; School: $F(1,31)=44.91$, $p<0.001$, $R^2=0.59$). Both models were improved by adding vision group as a predictor, which added around 30% of explained variance (Social: $F(3,29)=31.78$, $p<0.001$, $R^2= 0.77$, Adj.-$R^2=0.74$; School: $F(3,29)=21.39$, $p<0.001$, $R^2=0.69$, Adj.-$R_2=0.66$). The association between GAC and PedsQL scores were significant in both the S/PVI and MVI group (Social: $\beta_{(S/PVI)}=-40.42$, $p_{(S/PVI)}<0.001$, $\beta_{MVI}=-25.99$, $p_{MVI}<0.001$; School: $\beta_{(S/PVI)}=-17.12$, $p_{(S/PVI)}=0.009$, $\beta_{MVI}=-14.88$, $p_{MVI}=0.031$). These findings indicate that better adaptive behaviour is associated with better quality of life ratings in domains of Physical, Social, and School functioning with the strongest effects observed for children with S/PVI.

Next, a potential mediating effect of adaptive behaviour on quality of life was investigated. The results indicated that the relationship between vision group (S/PVI, MVI, control) and the quality of life associated with school (PedsQL-School) was mediated by general adaptive behaviour (ABAS-II GAC). As Figure 3 illustrates, the standardised regression coefficient between visual group and ABAS-II GAC was statistically significant, as was the standardised regression coefficient between ABAS-II GAC and PedsQL School. We tested the significance of this indirect effect by using a bootstrapping procedure (Rosseel, 2012). The bootstrapped standardised indirect effect based on 10,000 boot-strapped samples was 0.36, and the 95% confidence interval ranged from 0.127 to 0.593. Thus, the indirect effect was statistically significant ($p<0.003$, p_corr=0.012). There were no significant mediation effects for other PedsQL scales (Emotional, Social, Physical).

Figure 3 about here

Discussion

The current study investigated adaptive behaviours and quality of life in children with congenital visual impairment arising from congenital disorders of the peripheral visual system using parent-rated standard questionnaires that are commonly used in clinical practice. This is the first study to also investigate how mild-to-moderate visual impairment affects adaptive behaviour in children and whether it differs from children with more severe visual impairment. The children were in the average range intellectually and were compared with typically sighted peers of similar intellectual ability.

The results of the current study indicated reduced adaptive behaviour in children with visual impairment across domains of practical, social, and conceptual function. These results mirror previous studies that also reported reduced adaptive behaviour in school-age children and in adolescents with congenital VI (Greenaway, Pring, Schepers, Isaacs, & Dale, 2017; Metsiou et al., 2011; Papadopoulos, Metsiou, & Agaliotis, 2011a). In line with these reports, the most pronounced difficulties were found for children with S/PVI in practical aspects of adaptive behaviour, i.e. skills required to take care of oneself and to effectively interact with other people (Harrison & Oakland, 2008). Overall, adaptive behaviour in children with MVI was rated at an intermediate level between typically-sighted controls and children with S/PVI, i.e. the distribution of scores overlapped with the control and S/PVI group. Ratings along the individual questionnaire scales suggested that children with MVI have similar problems to children with S/PVI in some domains, e.g. Self-Direction, Community Use, and Health & Safety, while being more like typically-sighted children in other aspects, e.g. Functional Academics, Self-Care.

Different factors may be contributing to lower ratings for practical adaptive skills in children with VI. First, children with VI may have fewer opportunities for self-determined behaviour (Robinson & Lieberman, 2004), (Engel-Yeger & Hamed-Daher, 2013) that would help them to develop these skills, because parents may be overprotective or underestimate their ability to cope with tasks by themselves (Papadopoulos, Metsiou, & Agaliotis, 2011a). Second, daily living tasks may pose additional challenges for children with VI. For instance, it is easy to imagine that task like noticing that the waste basket is full and emptying it (Item 17) are a more challenging task for a child with VI compared to a typically-sighted child. Further, additional factors like mobility or physical skills that are known to be affected in children with VI (Hallemans, Ortibus, Truijen, & Meire, 2011; Papadopoulos, Metsiou, & Agaliotis, 2011a), (Aslan, Calik, & Kitiş, 2012) are likely to have influenced the ratings, e.g. "Attends fun activities at another's home" (Item 4). Biases of the questionnaire towards behaviours and activities that are heavily reliant on vision may also play a role, highlighting the need to develop specific questionnaires that rate adaptive behaviours in children with VI.

Ratings of strengths and difficulties (SDQ) further indicated that peer relationships are an area of concern for children with VI. Difficulties in social function have been consistently observed across pre-school, middle childhood, and adolescents in congenital VI (Bathelt et al., 2017; Greenaway et al., 2017; Jure et al., 2016; Tadić et al., 2010). Reported difficulties were most pronounced for children with S/PVI, but ratings in the MVI group overlapped with both the

S/PVI and control group. This may suggest that some peer relational difficulties are present in the MVI group, but to a lesser extent compared to the S/PVI group. Difficulties in the social domain may arise due to multiple influences. First, social cognition may develop differently in children with VI (Bathelt et al., 2017; Dale & Salt, 2008), potentially because salient social information like facial expressions are less accessible to children with congenital VI. Other constraints may arise from reduced opportunities for social interaction. For instance, children with visual impairment have been found to participate less in activities with their peers (Engel-Yeger & Hamed-Daher, 2013), which may be driven by peer rejection or exclusion from mainstream activities, e.g. sports (Cervantes & Porretta, 2013). Reduced mobility and independence may introduce additional boundaries that make it difficult for children with VI to take part in age-typical social activities with their peers (Metsiou et al., 2011; Papadopoulos, Metsiou, & Agaliotis, 2011a)

Importantly, variation in adaptive skills was found to relate to children's quality of life (QoL). In general, children with VI were rated lower for quality of life across all domains (Physical, Emotion, Social, and School) in line with previous studies (Chadha & Subramanian, 2011). Difficulties with Physical, Social, and School functioning were observed for both children with S/PVI and children with MVI. Notably, better adaptive behaviour was associated with higher QoL. Indeed, the effect of visual impairment on school-related aspects of QoL was significantly mediated by adaptive behaviour. Adaptive behaviour is therefore of considerable significance in improving the quality of life of children with VI. However, adaptive behaviour is multi-faceted and extra-curricular in relation to education for children with VI and may need continuing targeted focus from a range of professional supports including special educational needs coordinators (SENCo), specialist teachers for the visually impaired child, habilitation and mobility specialists, occupational therapists and clinical and educational psychologists. As adaptive behaviour transcends school and home, parents, specialist family support and youth workers also need to be brought into programmes of assistance. Of relevance, children with MVI also appeared to require this support even if they had relatively mild to moderate VI, in particular regarding community use, health and safety, home living. Adaptive behaviour is potentially amenable to behavioural interventions or adjustments in the environment, which could have a positive impact on the quality of life in school and at home for children with all degrees of congenital visual impairment.

Limitations

The current study has some limitations which affect generalisability of findings. First, the sample size was very limited, which is mainly due to the population rarity of the congenital disorders of the peripheral visual system and consequent recruitment challenges of children in this population who were also of normal intelligence during the time limited project. The individual vision level groups are small and the MVI group covered a wide range of functional vision with a few children who had measurable functional vision in the near normal range or at least in one eye in the MVI group at the time of the study. An associated potential issue concerns the heterogeneity of the sample. To reach a sample size that allowed for a meaningful group comparison, a range of congenital disorders were included. The individual ophthalmological disorders are extremely rare with often little understood and complex genetic causes so that varied visual and functional ability is even found within diagnostic categories. The representativeness of the current sample also demands cautious interpretation. Children were recruited to show an intellectual function within the typical range for their age, which is not the case for a large proportion of children with visual impairment who have additional intellectual disabilities (Alimovic, 2013). Greater difficulties on the SDQ sample have been reported in children with visual impairment and intellectual disabilities (Dijkhuizen, Hilgenkamp, Krijnen, van der Schans, & Waninge, 2016), (Harris & Lord, 2016). Further, the children were mostly recruited through specialised developmental vision and eye hospital clinical services so that the sample is potentially clinically biased towards children with specific problems even though these children were mostly referred for standard clinical care during the early years and were later discharged. A further limitation is that only parental questionnaire ratings were administered without further validation by behavioural measures or teacher-rated questionnaires or self-ratings. In the future, further investigation of adaptive behaviour in VI will be improved by using robust observational assessments that are specifically designed for and validated on children with VI. Vision related quality of life may also be appropriately compared with more generalized paediatric quality of life measures in the future (Tadic et al., 2016). Further, understanding the role of other factors that may influence adaptive behaviour and quality of life, like cognitive skills (Bathelt et al., 2016; Greenaway et al., 2017) and mental health (Harris & Lord, 2016) are important avenues for future research.

In conclusion, the findings of the current study indicate that severe VI is linked to reduced adaptive behaviour during mid-childhood, even when verbal abilities are in the age-expected range. Children with mild to moderate VI showed less pronounced decrements in adaptive behaviour, but parents still reported lower quality of life compared to typically-sighted peers. Reduced adaptive behaviour was found to mediate the relationship between visual impairment and quality of life in school indicating that adaptive behaviour may be a promising avenue for behavioural interventions.

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Figure captions

Figure 1: Profiles of parent ratings for adaptive behaviours in the vision groups. The lines show the average score of each group along the questionnaire scale. The error bars show two standard errors around the mean. Abbreviations: Communication (Com), Functional Academics (FA), Self-Direction (SD), Community Use (CU), Home Living (HL), Health & Safety (HS), Self-Care (SC), Leisure (LS), Social (Soc).

Figure 2: Profiles of parent ratings for strengths and difficulties. The markers indicate the mean in each group. The error bars show two standard errors around the mean. Abbreviations: Emotional Problems (Emotion), Conduct Problems (Conduct), Hyperactivity/Impulsivity (Hyperact/Impuls), Peer Relationship Problems (Peer Rel), Prosocial Behaviour (Prosocial).

Figure 3: Illustration of the statistical model indicating that the effect of the degree of visual impairment on the quality of life in school is mediated by general adaptive behaviour. Abbreviations: confidence interval (CI), quality of life (QoL).

Table captions

Table 1: Characteristics of children with visual impairment. Abbreviations: female (f), intelligence quotient (IQ), logarithm of the minimum angle of resolution (logMAR), male (m), mild-to-moderate visual impairment (MVI), profound visual impairment (PVI), severe visual impairment (SVI), standard error (SE), years (y)

Table 2: Characteristics of children with typical sight. Abbreviations: female (f), intelligence quotient (IQ), logarithm of the minimum angle of resolution (logMAR), male (m), standard error (SE), years (y)

Table 3: Composite scores for parent ratings of adaptive behaviour in groups with severe-to-profound VI (S/PVI), mild-to-moderate VI (MVI), or typical sight. Abbreviations: General Adaptive Behavior Composite (GAC), Practical (Prac.), Social (Soc.), Conceptual (Con.)

Table 4: Composite score for parent ratings of strengths and difficulties n groups with severe-to-profound VI (S/PVI), mild-to-moderate VI (MVI), or typical sight. Abbreviations: Physical (Phy), Emotional (Emo), Social (Soc), School (Sch).

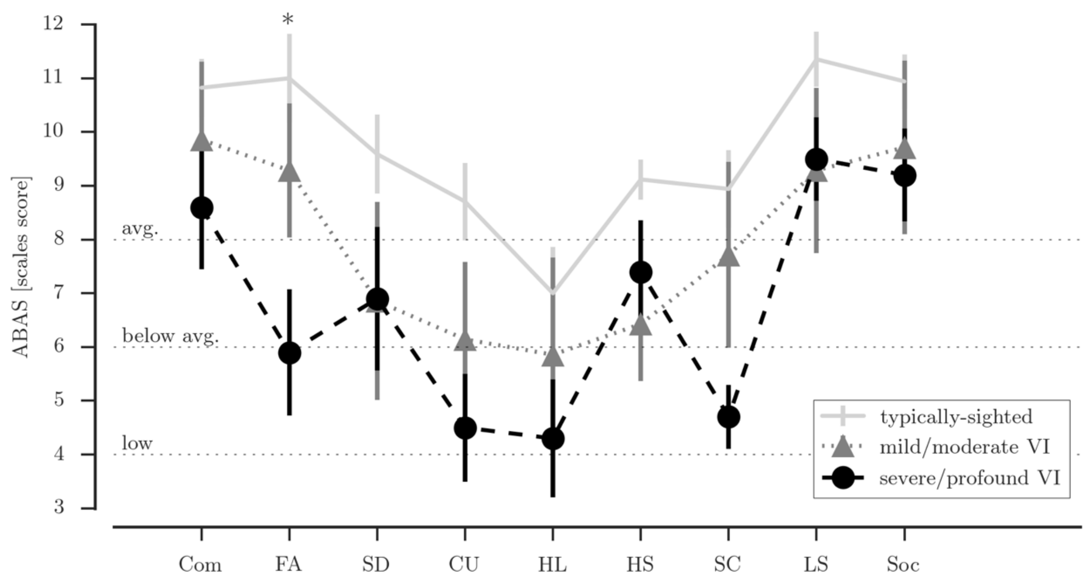

Figure 1

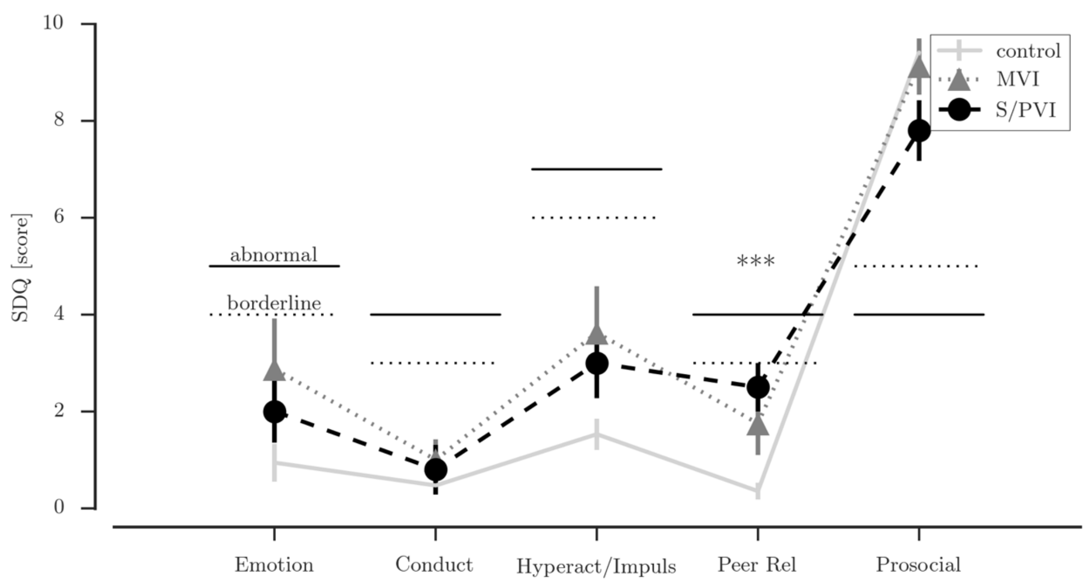

Figure 2

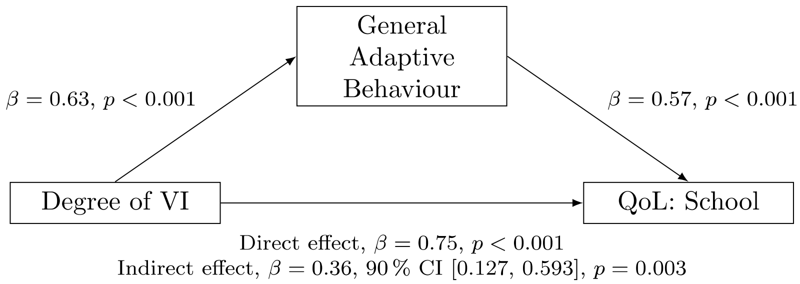
General Adaptive Behaviour
$\beta = 0.63$, $p < 0.001$
$\beta = 0.57$, $p < 0.001$
Degree of VI
QoL: School
Direct effect, $\beta = 0.75$, $p < 0.001$
Indirect effect, $\beta = 0.36$, 90 % CI [0.127, 0.593], $p = 0.003$

Figure 3

ID	Sex	Age	verbal IQ	logMAR	Near Detection	Vision Group	Visual Disorder
MVI 1	f	9.19	114	0.1	-	MVI	congenital nystagmus
MVI 2	f	13.32	95	0.4	-	MVI	ocular fibrosis
MVI 3	f	11.91	104	0.5	-	MVI	bilateral optic nerve hypoplasia
MVI 4	m	12.34	-	0.54	-	MVI	rod-cone dystrophy
MVI 5	f	8.27	104	0.6	-	MVI	oculocutaneous albinism
MVI 6	m	12.06	104	0.6	-	MVI	congenital nystagmus
MVI 7	m	10.64	116	0.6	-	MVI	congenital nystagmus
MVI 8	m	9.82	93	0.7	-	MVI	oculocutaneous albinism
MVI 9	f	12.26	96	left: 0.23	right: light perception	MVI	unilateral optic nerve hypoplasia
SVI 1	f	10.98	87	0.9	-	SVI	hereditary progressive cone dystrophy
SVI 2	m	11.69	148	0.9	-	SVI	oculocutaneous albinism
SVI 3	f	10.98	78	1.1	-	SVI	familial exudative vitreoretinopathy
SVI 4	m	9.57	119	1.2	-	SVI	Leber's congenital amaurosis
SVI 5	m	9.01	-	1.225	-	SVI	oculocutaneous albinism
SVI 6	m	9.91	96	1.225	-	SVI	Norrie's disease
SVI 7	f	11.04	75	-	1.5cm lure from 20cm	SVI	Leber's congenital amaurosis
SVI 8	f	9.86	95	-	12.5cm lure from 50cm	SVI	Bilateral micro-ophthalmia

ID	Sex	Age	verbal IQ	logMAR
C1	f	8.56	98	-0.3
C2	f	8.73	110	0.1
C3	m	8.90	116	-0.3
C4	m	9.08	102	0.1
C5	f	9.12	98	-0.1
C6	m	9.34	108	-0.2
C7	m	10.07	96	0.1
C8	m	10.16	134	0.0
C9	m	10.37	106	0.0
C10	m	10.74	102	-0.2
C11	f	10.78	134	0.1
C12	f	10.82	116	-0.2
C13	f	10.89	83	0.0
C14	f	11.09	130	-0.3
C15	f	11.78	144	0.1
C16	m	12.70	106	-0.2
C17	m	12.77	130	-0.2
C18	m	12.92	124	-0.3
	8f	mean 10.49	mean 113.17	

Table 3

	S/PVI		MVI		control		S/PVI vs control		MVI vs control		S/PVI v
	mean	SE	mean	SE	mean	SE	$p_{corr.}$	g	$p_{corr.}$	g	$p_{corr.}$
GAC	75.7	4.904	84.00	8.527	97.82	2.621	0.001	1.74	0.207	0.91	>0.999
Prac.	65.2	7.965	78.14	14.021	93.24	2.474	0.124	0.90	0.166	0.95	>0.999
Soc.	88.9	8.298	85.00	13.243	106.12	2.307	0.083	0.97	0.113	1.03	>0.999
Con.	83.1	9.823	83.57	11.729	102.18	2.894	0.002	1.62	0.511	0.69	>0.999

Table 4

	S/PVI		MVI		control		S/PVI vs control		MVI vs control		S/PVI vs MVI	
	mean	SE	mean	SE	mean	SE	$p_{corr.}$	g	$p_{corr.}$	g	$p_{corr.}$	g
Phy	60.94	3.332	69.59	3.204	94.30	1.277	<0.001	2.52	0.002	2.08	>0.999	0.45
Emo	62.40	3.709	62.86	3.171	83.82	2.435	0.075	1.24	0.107	1.35	>0.999	0.02
Soc	52.00	2.635	66.43	1.758	96.18	1.427	<0.001	3.86	<0.001	3.33	0.715	1.07
Sch	58.50	2.996	61.43	4.118	90.59	1.553	<0.001	2.50	0.005	1.94	>0.999	0.14