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Executive abilities in children with congenital visual impairment in mid-childhood

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ABSTRACT

The role of vision and vision deprivation in the development of executive function (EF) abilities in childhood is little understood; aspects of EF such as initiative, attention orienting, inhibition, planning and performance monitoring are often measured through visual tasks. Studying the development and integrity of EF abilities in children with congenital visual impairment (VI) may provide important insights into the development of EF and also its possible relationship with vision and non-visual senses. The current study investigates non-visual EF abilities in 18 school-age children of average verbal intelligence with VI of differing levels of severity arising from congenital disorders affecting the eye, retina, or anterior optic nerve. Standard auditory neuropsychological assessments of sustained and divided attention, phonemic, semantic and switching verbal fluency, verbal working memory, and ratings of everyday executive abilities by parents were undertaken. Executive skills were compared to age-matched typically-sighted (TS) typically-developing children and across levels of vision (mild to moderate VI [MVI] or severe to profound VI [SPVI]). The results do not indicate significant differences or deficits on direct assessments of verbal and auditory EF between the groups. However, parent ratings suggest difficulties with everyday executive abilities, with the greatest difficulties in those with SPVI. The findings are discussed as possibly reflecting increased demands of behavioral executive skills for children with VI in everyday situations despite auditory and verbal EF abilities in the typical range for their age. These findings have potential implications for clinical and educational practices.

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Executive function (EF) is used as an umbrella term for a set of inter-related cognitive abilities—including goal planning, control of attention, working memory, inhibition, and cognitive flexibility (Anderson, 2002; Diamond, 2013)—that are highly important for educational attainment and academic success in childhood and adolescence (de Haan, 2014; McDermott, Westerlund, Zeanah, & Fox, 2012; Stevens, Lauinger, & Neville, 2009). Current theoretical models about the early

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development of EF are largely based on observations of visual behaviors, even though the importance of early vision for early and later EF development is unknown (Colombo, 2001; Johnson & de Haan, 2011; Richards, Reynolds, & Courage, 2010). A link between vision and EF is possibly suggested by the close connection between visual processing streams with prefrontal regions and the fronto-parietal attention network (Kravitz, Saleem, Baker, & Mishkin, 2011; Ptak, 2012) and also the close relationship between visuospatial working memory, spatial abilities, and EF (Hecke, Mundy, & Acra, 2007; Miyake, Friedman, Rettinger, Shah, & Hegarty, 2001; Mundy, Block, Delgado, & Pomares, 2007). However, it is currently not clear if early or later visual behaviors are necessary for the development of executive abilities and the integration of EF networks in the brain or if experiences in other non-visual modalities (auditory, haptic) are sufficient for the development of EF in the absence of vision. Consequently, studying the development of children with congenital visual impairment (VI) may shed light on the relationship between EF, vision, visual experience and potential vulnerabilities and compensatory factors in the development of EF abilities in this clinical population. In addition, this is of high clinical and educational importance, as children with VI may have to rely more on their ability to plan, organize, and hold information in working memory when visual cues are inaccessible.

Congenital VI is associated with differences of large-scale structural and functional brain network organization (Amedi, Raz, Pianka, Malach, & Zohary, 2003; Liu et al., 2007; Noppeney, 2007; Raz, Striem, Pundak, Orlov, & Zohary, 2007; Shu, Li, Li, Yu, & Jiang, 2009), which may affect the distributed networks involved in EF (Cavezian et al., 2013). Evidence from an observational study indicates differences in potential precursors of executive behaviors, specifically attention shifting, in preschoolers with VI compared to matched typically-sighted (TS) peers (Tadic, Pring, & Dale, 2009). The authors report reductions in the frequency at which the preschoolers with severe to profound VI (SPVI) responded to adult attempts to elicit or maintain their attention, and in particular to shift their attention from one object to another through auditory, haptic or visual cues, with those with profound VI (PVI, i.e. light perception at best) observed as having the greatest difficulty. Interestingly, individual weaker responses to attention shifting were found to be significantly related to more problems in everyday behaviors requiring EF on the Behavior Rating of Executive Function (BRIEF), in particular Shifting, when the same children were observed at school age (Tadic, 2009). Neurodevelopmental differences that are potentially related to EF have also been reported in mid-childhood to adolescence in other samples of children with VI. A comprehensive survey in a sample of 264 children aged 4 to 17 years attending specialist clinics found a substantially higher prevalence of attention deficit hyperactivity disorder (ADHD) diagnoses in children with VI (22.9% compared to 14.3% in the TS population in the same geographical area; Decarlo et al., 2014), and EF has been shown to be a significant component in ADHD in sighted samples (Willcutt, Doyle, Nigg, Faraone, & Pennington, 2005). Further, a recent study indicates higher parental ratings of behavioral EF deficits, compared with normative population expectations, in a small sample of high-functioning adolescents with congenital VI and age-appropriate verbal IQ (Greenaway, Pring, Schepers, Isaacs, & Dale, 2016).

Whilst these preliminary small-scale studies suggest that behaviors related to EF may be negatively affected in children with VI, it is not clear how VI might impact on EF and whether specific aspects of EF are more vulnerable than others. Certain executive abilities might be more dependent on visual information during development, whereas other abilities may develop typically or be more amenable to compensatory mechanisms even when visual information is largely inaccessible or very degraded. The auditory and haptic modalities, and the mechanism of language, have been proposed to modulate developmental processes in the absence of vision (Perez-Pereira & Conti-Ramsden, 1999; Warren, 1994).

In this study, the aim is therefore to investigate the development of EF in the context of congenital VI during mid-childhood. It has been argued that this period is important for EF development, as rapid advances in executive ability have been observed in this age period (Xu et al., 2013). Further, executive abilities are believed to be more differentiated in mid-childhood compared to preschool years (Anderson, 2002; Diamond, 2013), allowing for a more fine-grained assessment of the possible impact of VI on EF development. To investigate this potential relationship, this study focuses on children with congenital visual disorders. The subpopulation of interest is those with disorders affecting the anterior or peripheral part of the visual system with no known involvement of central brain structures according to the visual disorder diagnosis (i.e., "potentially simple" congenital disorders of the peripheral visual system [CDPVSs]; Sonksen & Dale, 2002). In children with additional brain defects, as is common in cerebral VI (Rahi, Cable & BCVISG, 2003), the likelihood of comorbid learning difficulties is greatly increased. This would pose a significant confound, as any differences in cognitive performance may be potentially linked to the learning disability rather than to the impact of vision reduction per se (Sonksen & Dale, 2002). To further minimize the possibility of additional learning difficulties which can commonly occur in children with congenital VI (Alimovic, 2013), a sample of higher-functioning children with VI and normal-range verbal intelligence was selected. Standard auditory and verbal assessments of EF were employed, including assessments of working memory, auditory attention, and verbal fluency, in order to cover a range of executive tasks that do not require vision for performance. In addition, parents completed a standard questionnaire on everyday behaviors associated with EF. To further test the relationship between vision level and EF, children with differing degrees of VI —mild to moderate VI (MVI) through to SPVI—are included in the VI sample, which permits comparisons between children with a broad spectrum of congenital visual disorders along with sighted controls, as well as comparisons between different degrees of vision and vision reduction (PSVI versus MVI versus TS). The study design is therefore selected to permit novel insights into the potential impact of congenital vision reduction on EF in mid-childhood, including a comparison of those who remained profoundly or severely visually impaired with those who have continued to develop significant functional visual acuity by mid-childhood. Previous research of younger children has suggested that those with the most profound degrees of VI (especially light perception at best) exhibit the greatest developmental impact, with significant delays in cognition, language and social development (Dale, Tadić, & Sonksen, 2014; Dale & Sonksen, 2002), leading to the prediction that children with no or very low vision will also show a negative impact in EF abilities.

Assuming that visual input is necessary for the typical development of EF, it was hypothesized that children with VI would have lower standard scores on auditory tests of EF compared with TS matched controls, and that the scores would be even lower in children with the greatest severity of vision reduction (PSVI) compared with those with moderate vision reduction (MVI) and TS children. Standard neuropsychological assessment measures with good construct validity that make no demand on vision were selected for this study. In the absence of well-validated tactile or haptic assessments, these measures are either auditory or verbal. The only available auditory tasks that are suitable for children with VI and are all arguably tapping into EF are those of working memory, auditory attention and verbal fluency (Delis et al., 2007; Jurado & Rosselli, 2007; Manly, Nimmo-Smith, Turner, Watson, & Robertson, 2001). A parent-rated standard questionnaire measure is also included to assess performance and any difficulties with everyday behaviors associated with executive abilities.

Method

Participants

This project was approved by the NHS Paediatric Research Ethics Committee (Ref: 12/LO/0939) and written consent was obtained from all parents/guardians according to the Declaration of Helsinki.

A prospective cross-sectional study was undertaken with 18 children with VI aged between 8 and 13 years. Congenital disorders of the peripheral visual system with severe VI are rare, with an estimated prevalence of less than 2 to 3 per 10,000 children in the United Kingdom (UK), which raises challenges for recruitment and sampling (Rahi et al., 2003). Children were therefore recruited through national specialist clinics at the Great Ormond Street Hospital for Children NHS Foundation Trust and the Moorfields Eye Hospital NHS Foundation Trust. The investigations reported here are part of a larger study to investigate neural, cognitive and behavioral correlates in this sample. The inclusion criteria are as follows: (1) "potentially simple" CDPVSs (Sonksen & Dale, 2002), i.e., any visual disorder affecting the globe of the eye, the retina, or the anterior optic nerve up to the optic chiasm with no other known central nervous system involvement or brain insult in the pediatric diagnosis, originally diagnosed by pediatric ophthalmology; (2) English as a first language or sufficiently fluent level of English to participate in assessments; and (3) within the normal range for verbal reasoning (verbal IQ > 79). Identification of children was initially made through clinical databases and also self-recruitment where parents were asked if their child was attending school at the age-appropriate level. One participant was subsequently found to have a verbal IQ slightly below the inclusion criterion but did not act as an outlier on other assessments and was therefore retained in the analysis.

The exclusion criteria are as follows: (1) hearing impairment and/or severe motor impairment; (2) retinopathy of prematurity; and/or (3) pediatric diagnoses of comorbid neurological disorders or indication of other brain involvement or endocrine abnormalities, e.g. hypopituitarism (Garcia-Filion & Borchert, 2013).

A total of 18 children with normal or corrected-to-normal vision were recruited through local advertisement to match the VI children according to age. Children in the control group had to attend mainstream school at an age-appropriate level, have no known neurological or psychiatric conditions, and have English as their first language.

Table 1. Characteristics of Participants in the VI Group.

ID	Gender	Age (years)	VerbComp	logMAR	Near Detection	Vision Group	Visual Disorder
MVI1	F	9.19	114	0.100	-	MVI	Congenital nystagmus
MVI2	F	13.32	95	0.400	-	MVI	Ocular fibrosis
MVI3	F	11.91	104	0.500	-	MVI	Bilateral optic nerve hypoplasia
MVI4	M	12.34	-	0.540	-	MVI	Rod-cone dystrophy
MVI5	F	8.27	104	0.600	-	MVI	Oculocutaneous albinism
MVI6	M	12.06	104	0.600	-	MVI	Congenital nystagmus
MVI7	M	10.64	116	0.600	-	MVI	Congenital nystagmus
MVI8	M	9.82	93	0.700	-	MVI	Ocular albinism, congenital nystagmus
MVI9	F	12.26	96	-	Left: 0.23; Right: light perception	MVI, PVI	Unilateral optic nerve hypoplasia
SVI1	F	10.98	87	0.900	-	SVI	Hereditary progressive cone dystrophy
SVI2	M	11.69	148	0.900	-	SVI	Oculocutaneous albinism
SVI3	F	10.98	78	1.100	-	SVI	FEVR, *LRP5* mutation
SVI4	M	9.57	119	1.200	-	SVI	Leber's congenital amaurosis
SVI5	M	9.01	-	1.225	-	SVI	Ocular albinism, nystagmus
SVI6	M	9.91	96	1.225	-	SVI	Norrie's disease
SVI7	F	11.04	75	-	1.5 cm sweet from 20 cm	SVI	Leber's congenital amaurosis
SVI8	F	9.86	95	-	12.5 cm wooly ball 50 cm	SVI	Bilateral micro-ophthalmia, *SOX6* mutation
PVI1	M	10.36	134	-	Light perception only	PVI	Leber's congenital amaurosis
	9 female 9 male	*M* = 10.73 *SE* = 0.31	*M* = 103.63 *SE* = 4.41				

Note. FEVR = familial exudative vitreoretinopathy; logMAR = Sonksen logMAR test of Visual Acuity; MVI = mild to moderate VI (degraded visual acuity); PVI = profound VI (light perception at best); SVI = severe VI (basic form vision); VerbComp = WISC-IV Verbal Comprehension age-normed score; VI = visual impairment; WISC-IV = Wechsler Intelligence Scale for Children – Fourth Edition. Demographic information and the results of the verbal ability and visual acuity assessments are listed.

The sample characteristics are summarized in Table 1 and Table 2. The experimenter (JB) was trained by a neurodisability pediatrician who specializes in VI (AS) to undertake the visual acuity assessments using the Sonksen logMAR test of Visual Acuity (Sonksen, Wade, Proffitt, Heavens, & Salt, 2008). For children who were unable to see the largest items on the logMAR, the Near Detection Scale was used to assess the basic level of detection vision (Sonksen, Petrie, & Drew, 1991).

SPVI is defined as limited form vision with a logMAR above 0.8 (Snellen worse than 6/36) to no perception or light perception only (on the Near Detection Scale). MVI is defined as reduced visual acuity with a logMAR of between 0.6 and 0.8 (Snellen 6/24–6/36).

Procedure and Testing Environment

Participants were tested by an experimenter trained in the assessment of children with VI (JB) under the supervision of a clinical psychologist who specializes in VI (ND). Assessments were carried out in a quiet testing room in the university hospital center. Children were given frequent breaks between assessments to maintain optimal performance and promote well-being.

Table 2. Characteristics of the Typically-Sighted (TS) Control Group.

ID	Gender	Age (years)	VerbComp	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
	8 female	*M* = 10.49	*M* = 113.17	
	10 male	*SE* = 0.32	*SE* = 3.87	

Note. logMAR = Sonksen logMAR test of Visual Acuity; VerbComp = WISC-IV Verbal Comprehension age-normed score; WISC-IV = Wechsler Intelligence Scale for Children – Fourth Edition.

Verbal Comprehension

In order to exclude the possibility that any difference in EF may be due to underlying differences in intellectual ability, a standard test of verbal comprehension was administered. Verbal comprehension was assessed using verbal subtests of the Wechsler Intelligence Scale for Children – Fourth Edition (WISC-IV; Wechsler, 2004). Verbal subtests of previous and current editions of the WISC have also been used with children with VI (Dekker, 1993; Greenaway et al., 2016; Tillman, 1973; Tillman & Bashaw, 1968; Witkin, Birnbaum, Lomonaco, Lehr, & Herman, 1968).

The administered subtests include all the items of the Verbal Comprehension composite score (Vocabulary, Similarities, Comprehension). Two items were altered that required direct visual experience: the WISC-IV first practice item on the Similarities subtest which includes color was not administered, and 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 TS control group. All other items were administered verbatim according to the WISC-IV administration manual (Wechsler, 2004).

Analysis of verbal comprehension by vision group (SPVI, MVI, TS) does not indicate significant differences between the groups: SPVI: $M = 100.78$, $SE = 8.94$, range = 75–148; MVI: $M = 103.25$, $SE = 3$, range = 93–116; TS: $M = 113.17$, $SE = 3.87$, range = 83–144; $F(2, 32) = 1.665$, $p = .205$.

Ef Tasks

Working Memory

Tasks comprising the Working Memory composite of the WISC-IV (Wechsler, 2004) were administered to determine working memory performance. The Working Memory composite was calculated from the Digit Span and Letter-Number Sequence scale scores.

Sustained and Divided Auditory Attention

Auditory attention was assessed using tests from the Test of Everyday Attention for Children (TEA-Ch; Manly et al., 2001). In the Score! subtest, children have to count infrequently presented sounds in several trials over a 6-minute period. Because of the long pauses between tones and simple task demands, children have to actively sustain their attention to perform the task (Anderson, 2002). The Score Dual Task condition requires children to count the number of scoring sounds while listening out for an animal name in a simultaneously presented news broadcast (Manly et al., 2001).

Verbal Fluency

The Verbal Fluency task of the Delis-Kaplan Executive Function System (D-KEFS; Delis et al., 2007) consists of three conditions. In the Letter Fluency task, the participant has to name as many words as possible that start with a given letter within 60 seconds. In the Category Fluency task, the participant has to name words within 60 seconds that belong to a semantic category. In the Category Switching task, participants have to switch between words that belong to different semantic categories. All tests were administered according to the test manual. The DKEFS Verbal Fluency subtest typically requires the assessor to talk through the rules as well as present them visually in print. As the participants were unable to access the print, the assessors ensured that the participants had understood the rules by talking through these carefully and clearly, providing repetition if required.

A total of 17 children in the VI group (7 male, 8.27–13.32 years, WISC Verbal Comprehension: 75–148) and 17 children in the TS group (10 male, 8.56–12.92 years, WISC Verbal Comprehension: 83–144) completed the Verbal Fluency tasks, as 1 participant from each group did not complete them.

Everyday Executive Skills

The BRIEF is an 86-item questionnaire suitable for children aged 5 to 18 years (Gioia, Isquith, & Kenworthy, 2000). The questionnaire rates executive skills in the domains of Inhibition, Shifting, Emotional Control, Initiation, Working Memory, Planning/Organizing, Organization of Materials and Monitoring. Only one of the items used to create these scores makes a reference to visual behavior (Item 31: "Has poor handwriting"), but does still apply to the majority of the children in this study with MVI. Another 2 items may be indirectly related to vision—Item 67 ("Cannot find things in room or school desk") and Item 68 ("Leaves messes wherever he/she goes")—but also reflect executive contributions. These tasks may be harder for children with VI, but they do not necessarily depend on vision. For this reason, parents were given the full questionnaire without any modifications.

Inconsistency scores are below the 98th percentile and are therefore in the acceptable range according to the questionnaire manual. There are two cases of highly elevated Negativity scores in the VI group (above the 98th percentile). High negativity scores may indicate an excessively negative attitude of the rater, but may also suggest extreme executive dysfunction (Gioia et al., 2000). Separate analysis showed no effect of the inclusion or exclusion of these cases for the group results; therefore, the presented results include the cases with high negativity ratings.

Statistical Analysis

The statistical analysis is based on analysis of variance (ANOVA) models. Mauchly's test was used to assess violations of the sphericity assumption (Mauchly, 1940). In the case of violated sphericity assumptions, the Greenhouse–Geisser correction was applied (Greenhouse & Geisser, 1959). All statistical tests were performed in *R* v2.15.3 (R Development Core Team, 2008). Follow-up contrasts were based on Student's *t*-tests. Welch correction was applied to account for differences in variance between the groups (Welch, 1947). Visualization were based on ggplot2 algorithms (Wickham, 2009). A significance level of $p < .05$ was used for all statistical analyses. Values between .05 and .10 are discussed as trend-level effects.

Results

Working Memory

Statistical analysis does not indicate significant differences in the Working Memory composite score between the vision groups (SPVI, MVI, TS), $F(2, 31) = 0.079$, $p = .971$ (see Table 3 for descriptive statistics). There is also no significant effect of vision group on the Digit Span, $F(2, 32) = 0.824$, $p = .448$, and Letter-Number Sequence, $F(2, 32) = 1.033$, $p = .368$, scores.

Sustained and Divided Auditory Attention

Statistical analysis does not indicate significant differences in the Working Memory composite scores between vision groups, $F(2, 32) = 0.515$, $p = .602$ (see Figure 1 and Table 3 for descriptive statistics). There is also no significant effect of vision group in the divided attention condition, $F(2, 32) = 1.599$, $p = .218$. A high proportion of participants in both groups reached scores in the superior to highly superior range compared to the normative sample of the test (Figure 2). However, there is also considerable within-group variability in the VI group, including scores in the low range ($n = 2$).

Verbal Fluency

Statistical analysis does not indicate significant differences in the Letter Fluency scores between vision groups, $F(2, 32) = 0.711$, $p = .499$ (see Figure 1 and Table 3 for descriptive statistics). There is no significant effect of vision group on category fluency scores, $F(2, 30) = 0.737$, $p = .487$. There is also no significant effect of vision group on the number of responses in the switching condition, $F(2, 30) = 0.128$, $p = .88$, or on switching accuracy, $F(2, 30) = 0.314$, $p = .733$.

Table 3. Descriptive Statistics of Mean Scores and Standard Errors of the Mean (*SE*) across EF measures.

		SPVI		MVI		TS	
		M	*SE*	*M*	*SE*	*M*	*SE*
WM	Total	99.10	12.59	101.25	4.08	100.50	2.41
	Digit Span	10.44	1.21	8.75	0.70	9.22	0.66
	L-N Seq	9.56	1.59	10.12	0.97	11.17	0.32
Aud Att	Sustained	10.11	1.02	10.38	0.84	10.94	0.70
	Divided	10.78	1.39	11.00	1.10	12.89	0.64
Verb Fl	Letter	12.75	1.95	13.00	1.30	12.63	0.99
	Category	9.25	1.86	11.12	0.93	12.06	0.87
	Swtch Resp	11.12	1.52	12.50	1.32	11.76	0.62
	Swtch Acc	10.50	1.50	12.00	1.16	12.06	0.52
BRIEF	GEC	62.86	5.43	58.12	7.33	45.11	3.25
	BRI	59.14	6.51	58.88	6.96	42.83	4.10
	MI	62.86	4.68	57.75	6.85	43.50	1.93

Note. Aud Att = auditory attention; BRI = Behavioral Regulation Index; BRIEF = Behavior Rating Inventory of Executive Function; GEC = global executive composite; L-N Seq = Letter-Number Sequence; MI = Metacognitive Index; MVI = mild to moderate VI; SPVI = severe to profound VI; Swtch Acc = switching accuracy; Swtch Resp = switching responses; Verb Fl = verbal fluency; VI = visual impairment; WM = working memory.

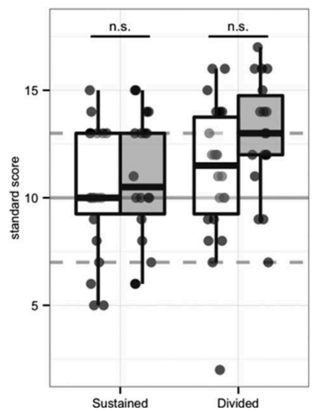

Figure 1. Results of the sustained and divided auditory attention task.

Note. The distribution of standardized scores is shown for the VI group (black) and the TS group (gray). The solid gray line indicates the mean of the normative sample. The dashed lines show 1 *SD* variance of the mean of the normative sample (Manly et al., 2001). There are no significant differences between the groups in either condition.

Everyday Executive Skills

Half of the children with VI reached the threshold for clinical concern regarding executive deficits on the BRIEF (n = 9, 4 MVI, 5 SPVI, see Figure 3) over global executive composite (GEC, cut-off at 65, > 93rd percentile). Statistical comparison indicates a main effect of vision group, $F(2, 27) = 4.444$, $p = .022$. Follow-up contrasts indicate significantly higher scores in the SPVI group compared to the TS group, $t(10.58) = 2.806$, $p = .018$. Other contrasts do not reach significance level.

A total of 8 children with VI scored above the cut-off on the Behavioral Regulation Index (BRI, 45%, 4 MVI, 4 SVI, cut-off at 65, > 93rd percentile). Scores on the BRI also show a significant effect of vision group, $F(2,27) = 6.248$, $p = .006$). Post-hoc contrasts

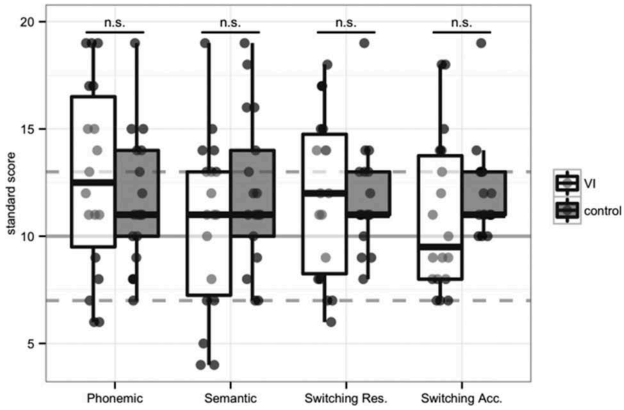

Figure 2. Results of the semantic, phonemic, and switching verbal fluency assessment.

Note. The distribution of standardized scores is shown for the VI group (black) and the TS group (gray). The number of responses and switching accuracy are shown separately for the switching condition (Delis, Kramer, & Kaplan, 2001). The solid gray line indicates the mean of the normative sample and the dashed lines show 1 *SD* variance from the norm mean.

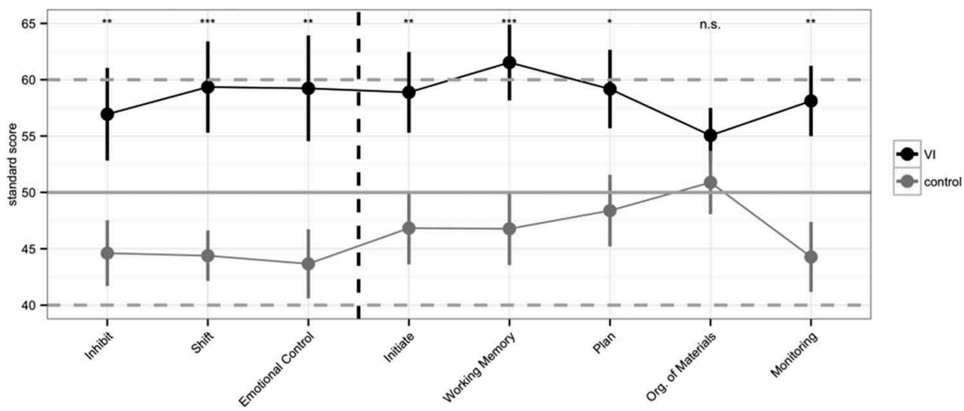

Figure 3. Results of the everyday executive ability parent questionnaire.

Note. The mean score and standard error on each scale are shown for children in the VI group (black) and the TS group (gray). The solid gray line indicates the mean scores of the normative sample and the dashed lines show 1 *SD* of variance from the mean. The scales on the left of the vertical black line made up the Behavioral Regulation Index, while the scales to the right are summarized in the Metacognitive Index (Gioia et al., 2000). There are significant differences on all scales, except for Organization of Materials.

reveal a significantly higher score in the SPVI group compared to the TS group, $t(7.827) = 2.339$, $p = .048$, and a trend-level difference between the MVI group and the TS group, with higher scores in the MVI group, $t(8.851) = -2.171$, $p = .058$. There is no significant difference between the two VI groups.

A total of 7 children with VI reached scores above the cut-off on the Metacognitive Index (MI, 38%, 3 MVI, 4 SPVI, cut-off at 65, > 93rd percentile). Statistical analysis also indicates a significant difference between vision groups on the MI, $F(2, 27) = 8.020$, $p = .001$. Follow-up contrasts indicate significantly higher scores in the SPVI group compared to the TS group, $t(3.82) = 8.127$, $p = .005$, as well as a trend-level difference between the MVI group and the TS group, with higher scores in the MVI group, $t(8.134) = -2.01$, $p = .079$. The difference between the VI groups (MVI vs. S/PVI) is not statistically significant, $t(13.683) = -0.405$, $p = .692$.

Discussion

The relationship between congenital VI and EF abilities in mid-childhood has not been systematically studied before and the available theories and evidence suggest that a lack of vision and deprivation of visual information from the environment might adversely impact on the developmental and/or behavioral aspects of EF. Nevertheless, alternative sensory functions of audition and touch might provide compensatory avenues for developing EF abilities. To investigate this further, this study focuses on the performance of EF abilities in a sample of 18 children with congenital VI in mid-childhood, compared with age-matched typically-developing TS controls. The precautionary methodological approach to reducing potential confounding influences of comorbid learning disability (which is high in children with congenital visual disorders) or an inability to perform the EF task because of lacking the vision necessary to see the materials consisted of the following inclusion criteria: 1) children with VI in the "simple" CDPVS subpopulation; 2) higher-functioning range of verbal intelligence; and 3) no tasks requiring vision. Contrary to arguments leading to the hypothesis that EF abilities would be adversely constrained in children with VI and particularly in those with the most severe VI (light perception or low levels of "form" vision), no significant differences were found in standard scores of EF tasks of working memory, sustained and divided attention, and phonemic, semantic, and switching verbal fluency between the VI group and the age-matched TS groups. Moreover, the mean standard scores of the VI group are on average in line with age-appropriate population norms.

A number of theoretical positions could explain this "typical" performance in EF neuropsychological tasks in children with VI. Firstly, infancy and later experience in auditory and haptic sensory modalities—including possibly the mediating role of language and non-verbal physical and object experiences—assists the development of metacognitive thought processes and mental abstraction involved in executive skills. In terms of the possible origin of EF abilities in childhood, present theoretical models of possible precursors in infant behavior are largely based on the observation of visually-mediated behaviors, like saccades (Colombo, 2001; Richards et al., 2010). For instance, the ability to shift visual fixation from an intrinsically attractive visual stimulus to a less intrinsically attractive but task-relevant visual stimulus is seen as a precursor of top-down executive control in longitudinal studies (Nakagawa & Sukigara, 2013; Papageorgiou et al., 2014). To the authors' knowledge, there are currently no theories of infancy EF development based on other auditory or haptic modalities, potentially due to methodological difficulties in assessing these functions in infants—though auditory oddball paradigms may be revealing in the future (Gomes, Molholm, Christodoulou, Ritter, & Cowan, 2000). Investigations of auditory or haptic (tactile) aspects of EF precursors in infants with congenital VI will need to be pursued, although the methodological challenges cannot be understated. The present finding in relation to auditory EF function raises the possibility that alternative non-visual modalities may provide a compensatory route to the development of EF.

Secondly, there may be modality-specific executive skills that are tied to the availability of sensory information and, in this sense, verbal and auditory EF skills would be expected to develop smoothly to a relatively well-preserved level in children with VI. Others have also argued for the important role of verbal ability in EF function in childhood (Henry, Messer, & Nash, 2015). A related model to this is that if children with VI are restricted to modality-specific EF skills then they might be expected to have much greater difficulty in areas of EF associated with vision, such as design fluency and spatial working memory. This dimension is not explored in this paper but is worthy of further investigation to see if effects are amodal or modality-specific.

A third possible model is that EF abilities are a unitary construct in mid-childhood. If EF is unitary in mid-childhood and this sample of children with VI scored in the age-appropriate range compared to the TS sample then one might deduce that EF is amodal in mid-childhood and can be executed through visual or auditory/verbal means. Xu et al. (2013) demonstrated that from 5 to 7 and 8 to 11 years of age, children's performance on different EF tasks is explained best by a single-factor model rather than the three-factor model of working memory, inhibition, and shifting commonly described for adults (Miyake et al., 2000). The similar performance on the different auditory neuropsychological tests in both the VI and TS groups could reflect the early unitary nature of EF. This might explain the finding that the children with VI did relatively well on all aspects of the EF assessment tasks used in this study. Executive abilities might diversify into discrete EF abilities in adolescence, with differences between VI and TS participants emerging at this later developmental stage. The preliminary results presented by Greenaway et al. (2016) suggest that this may be the case. However, despite apparent similar abilities on the group level, some children with VI displayed highly uneven neuropsychological profiles, with extreme weaknesses on certain tasks (see too Greenaway et al., 2016 in higher functioning adolescents with VI). For reasons not yet understood, there is extreme variation between and within some of the children, with scores ranging from extremely low to a superior level. A more detailed investigation of the potentially multiple factors contributing to these individual differences (Sonksen & Dale, 2002) will only be possible through the assessment of larger samples in future studies.

In contrast to test performance, the results of the behavioral ratings (BRIEF, parent ratings) show significant differences between the VI and TS groups and indicate that around half of the children with VI reached the clinical threshold for EF difficulties (> 93rd percentile). According to expectations for TS children, these scores would indicate significant difficulties in the domains of behavioral regulation and metacognition. These findings replicate the results of an independent sample of 6- to 12-year-old children with SPVI and typical intelligence (Tadić et al., 2009) and results-based teacher reports in a wider sample of children with VI (Heyl & Hintermair, 2015). Further, the current study provides evidence that behavioral EF is also affected in some children with MVI.

It has been argued that standardized neuropsychological tests of EF place reduced demands on executive skills by providing an adult-directed environment with clear instructions, training items, and a problem-solving scaffold. These aids are rarely available in everyday dynamic situations requiring EF abilities such as taking initiative, generating new ideas, making plans, achieving goals and self-organization of materials (Isquith, Roth, & Gioia, 2013). The discrepancies found in this study between the assessment scores and the parent ratings might therefore indicate that "core" cognitive skills in standard EF tasks are similar in both the VI and the control group, but that everyday demands on dynamic performance requiring executive skills are much higher for children with VI, e.g., lack of access to visual information from the environment may increase the cognitive load of a task (Bertone, Bettinelli, & Faubert, 2007) and reduce the environmental supports for the basic mobility and orientation required for executing any physical or goal-focused activity (Warren, 1994). This argument is further reinforced by the finding that more severe levels of VI (SPVI vs. MVI group comparisons) are significantly associated with more everyday behavioral executive difficulties. Further, children with SPVI who are likely to receive more assistance may have less opportunities to practice relevant behaviors, leading to less proficiency at performance level despite intact "core" skills.

Alternatively, the current findings could be explained by both higher vulnerabilities in the VI group in some EF skills—such as taking initiative and achieving goals—in addition to higher performance demands, particularly in the children with the most severe VI. Moreover, further evidence of a highly similar discrepancy between test performance on similar neuropsychological EF tasks and the parent-rated BRIEF in a small sample of 12- to 16-year-olds with VI suggests that this may be a longstanding and continuous pattern across later childhood (Greenaway et al., 2016) and that further research is required to identify the specific constraints underlying this apparent behavioral vulnerability.

Limitations

The current investigation is constrained in several ways which potentially limit the generalizability of the findings. First, the sample size is limited to 18 cases due to the recruitment challenges of the very rare "simple" congenital disorders of the peripheral visual system (Rahi et al., Cable, 2003); other studies on VI are often of a similar size for this reason (Absoud, Parr, Salt, & Dale, 2011; Tadic et al., 2009). Because of this small sample size, only large effects between group means could be detected, and thus the investigation of subtler group differences may have been underpowered (Button et al., 2013). Further, in order to recruit a sufficient number of individuals, a range of congenital visual disorders are included that share common functional symptoms. Despite this heterogeneity, the overall similarity of the test scores across the VI sample suggests that common functional issues (VI and degree of severity of vision reduction) are of greater relevance than individual anatomical disorders of globe, retina or optic nerve (Sonksen & Dale, 2002).

Second, there is an absence of EF tests that are designed for and have been validated on children with VI. However, the similar performance between the VI sample and the TS control group on most of the standard tasks implies that validity and reliability are unlikely to be seriously constrained. This also means that some areas of EF such as set shifting, problem-solving and design fluency could not be assessed due to the lack of suitable tests; the current study can therefore not be viewed as a broadly comprehensive investigation of EF abilities in children with VI.

Particularly striking is the discrepancy in the results between the neuropsychological tests and the behavioral questionnaire, which may reflect methodological issues. The questionnaire measure may tap into different dimensions or constructs related to EF compared to standard assessment or lab-based measures (Chan, Shum, Toulopoulou, & Chen, 2008; Ten Eycke & Dewey, 2015; Toplak, Bucciarelli, & Jain, 2008; Toplak & West, 2013). This is supported by similar discrepancies that have been reported in other clinical populations e.g., frontal lobe patients (Chan et al., 2008; Shallice and Burgess, 1991). However, parent ratings may also be less accurate than direct standardized testing and may reflect unrealistic parental expectations of the performance of children with VI.

Conclusion

The present study is the first—to the authors' knowledge—to report on EF abilities based on systematic neuropsychological assessments in a group of higher-functioning children with congenital VI and to then relate this to current precise levels of vision reduction. The study provides persuasive evidence that children with VI—including with severe to profound vision reduction—can succeed in auditory and verbal neuropsychological tests of working memory, attention and verbal fluency to the same level as age-matched TS controls.

The results of the current investigation have potentially important implications for clinical and educational practice. The results of the parent behavioral questionnaire may indicate that even though a child may be doing relatively well at school on academic tasks, some of their behavioral EF abilities may not be developing as smoothly—and any constraint in this area could impact on secondary school years, where higher autonomy and independence are required. Further research would be useful in a larger sample of 11- to 15-year-olds to investigate whether children can apply their cognitive and behavioral EF abilities in a secondary school environment. In mid-childhood, parents may be the first to be concerned about their child's difficulties at home, but educators and clinicians also need to be alerted when a child is struggling in sustaining or dividing attention in a busy classroom, taking initiative, shifting between mental sets or tasks, and/or generating new ideas to devise and follow goal-directed plans. Of further clinical concern, EF difficulties in older children with VI predict greater behavioral problems and socio-emotional difficulties (Heyl & Hintermair, 2015). In these circumstances, a specialized clinical neuropsychological assessment could be valuable in identifying needs and providing guidance or intervention for supporting EF abilities. Further research would be beneficial for developing and evaluating interventions to assist the more vulnerable school-aged children with VI and weaker EF abilities. Greater severity of VI is a particular risk factor, but even in MVI some children struggle in this area.

This study is limited to higher-functioning children with VI, and there are many children with VI who also have additional neurological impairment (Rahi et al., 2003); it is predicted that they will struggle to a greater extent with EF-related abilities (Heyl & Hintermair, 2015). Autism-related difficulties are present in a significant proportion of children with VI as well (Mukkades, Kilincaslan, Kucukaltun-Yildirim, Sevketoglu, & Tuncer, 2007; Parr, Dale, Shaffer, & Salt, 2010), and according to research on children with isolated autism, a higher level of EF-related difficulties is predicted in this subgroup (Kleinhans, Akshoomoff, & Delis, 2005; Lopez, Lincoln, Ozonoff, & Lai, 2005; Ozonoff, Pennington, & Rogers, 1991). Intellectual disabilities are also highly prevalent in children with congenital VI and thus likely to impact on executive abilities (Alimovic, 2013), but it not yet clear if this arises as a consequence of visual deprivation or as a comorbid disorder. Further research and clinical investigations and interventions are recommended for these vulnerable subgroups.

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