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To cite this article: Mathijs P. J. Vervloed, Ellen C. G. van den Broek & Ans J. P. M. van Eijden (2019): Critical Review of Setback in Development in Young Children with Congenital Blindness or Visual Impairment, International Journal of Disability, Development and Education, DOI: 10.1080/1034912X.2019.1588231

To link to this article: https://doi.org/10.1080/1034912X.2019.1588231

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Published online: 13 Mar 2019.

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Critical Review of Setback in Development in Young Children with Congenital Blindness or Visual Impairment

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\section*{ABSTRACT}
A presumed setback in development between 16 and 27 months of age was studied by reviewing the literature on setback in development and longitudinal studies of children with blindness or visual impairment. The results showed that the period around the second year of life is a vulnerable period for these children, in which about 25–33\% of the children who are blind show a setback. Setback is far less common in children with less severe visual impairments (±3\%). Important risk factors are profound visual impairments, neurological abnormalities and social adversity. Individual differences in maturation and development and methodological issues complicate conclusive statements about the existence and the nature of this setback. Alternative explanations for a developmental setback are described, such as measurement error and an over-diagnosis of Autism Spectrum Disorder. The main suggestion for future research is that in order to be able to capture the phenomenon of developmental setback and to pinpoint moderating and mediating factors, children with congenital blindness and visual impairment need to be monitored prospectively in a longitudinal research design.

\section*{Introduction}
In 2008 Dale and Salt published a review paper on developmental setback (DS) in children with blindness and visual impairment (Dale & Salt, 2008). They described DS as a stasis or regression in development starting between the ages of 16 and 27 months, following an initial period of normal developmental progress. They based their review on an existing database of 1300 children with visual impairment (VI). This retrospective research focused on children with blindness and severe VI with ‘potentially simple’ congenital disorders of the peripheral visual system (CDPVS) that is disorders of the globe, retina, and anterior pathways of the optic nerve. The word ‘simple’ refers here to disorders that are not part of neuro-ophthalmologic conditions. Children with potentially brain damaging events (i.e. complex CDPVS) were excluded. Of the children with CDPVS 33\% showed a DS if the visual impairment was profound (PVI, i.e. no form vision; comparable to the WHO definition of blindness) in contrast to 3\% in children with Severe Visual Impairment (SVI, i.e. form vision or better visual acuity) (Dale & Salt, 2008).
One of the problems with this presumed setback in the development of children with blindness is that it is difficult to determine whether this is a regression (i.e. loss of skills), stasis (i.e. plateauing of scores), and delay in development or is an adaptive but atypical developmental pathway caused by blindness. Lack of reliable prospective longitudinal studies is one reason for this problem. The great variability in outcome reported in the literature is another reason. Children with blindness often follow atypical developmental paths and show an increased risk of developmental delay throughout the infancy and preschool years (Hatton, Bailey, Burchinal, & Ferrell, 1997; Hatton, Schwietz, Boyer, & Rychwalski, 2007; Reynell, 1978). The high prevalence of additional disabilities in children with visual disabilities – 68% according to Hatton et al. (1997, 2007) and 97% in children with cerebral visual impairments (Boonstra et al., 2012) – complicates the picture, since it obscures whether, when a setback or delay is detected, this is the result of the blindness or visual impairment, other disabilities or both.

The low incidence of blindness in children, the heterogeneity of the group, and the individual variation pose three problems. Firstly, even for experienced clinicians, it is difficult to assess skills and problems in the development of children who are blind correctly. Secondly, it obscures alarm signals and risk factors associated with developmental problems. Lastly, it is hard to differentiate transient behavioural problems often found in young children with blindness from more enduring precursors of psychopathology. Despite the extensive studies on children with blindness (see Brambring, 2005, 2006, 2007a, 2007b; Dik, 2005; Hatton et al., 1997; Jan, Freeman, & Scott, 1977; Pérez Pereira & Conti-Ramsden, 1999; Warren, 1994) normative data on developmental patterns of cognition, language, sensory-motor and psychosocial development in children who are blind or have visual impairment are very limited, and hardly existent after 2000. Note that the studies of Brambring but also Dale and Salt all refer to data gathered mainly before 2000.

The literature results were in line with our own clinical experience. In preparation of the current study, an analysis of available client records showed us that most children who were blind develop well, sometimes atypical but this was adaptive. However, about 30% of the children who were blind and had no apparent additional disabilities showed a DS in development and behaviour, starting at approximately 18 months of age after a period of seemingly normal development and without clear immediate environmental cause. Children, for example, whose development of language skills decelerated or stagnated, while some children even lost language skills. Other children developed tactile defensiveness and/or stopped exploring their surroundings. Some of them became more and more passive and withdrawn. In some children an increase in stereotyped behaviour was seen, such as eye poking, biting and twirling. Lastly, we saw an increase in rigid behaviour, temper tantrums and uncooperative behaviour. Testing of the children was not performed at fixed ages and under controlled situations. Therefore these data were anecdotal and not reliable enough to make inferences.

Notwithstanding our clinical observations, there are still some questions left unanswered after the study of Dale and Salt. Firstly, all empirical data stemmed from just one setting, leaving it unknown whether a DS is a problem in children with visual impairment and blindness in general or just for a specific population from one geographical area. Secondly, most data were collected retrospectively. As a result, the exact timing of the DS is hard to determine and a selection bias could have obscured the prevalence of DS. Lastly, just a few developmental domains were studied, mainly language, cognition
and sometimes social development. A possible DS for other domains, such as motor development and adaptive skills, is therefore unknown.

A closer look at the development of children with visual impairment and blindness and a possible DS is important for clinical practice. Clinicians want to know what the factors are that constitute a DS, and more importantly, whether it is possible to prevent or overcome a setback. On the other hand, as seen from a biopsychological perspective it could be that certain behaviours are temporary adaptive behaviours helping the child to cope with environmental challenges. The goal of the present study is, therefore, to examine whether in longitudinal studies on the development of children with congenital blindness or visual impairment there is empirical evidence for the existence of a DS as postulated by Dale and Salt in 2008.

Methods

The literature search followed the PRISMA statement for reporting items for systematic reviews (Moher, Liberati, Tetzlaff, & Altman, 2009). The statement consists of a checklist, flow diagram, and a summary paper. Its aim is to help authors improve the reporting of systematic reviews and meta-analyses (Moher et al., 2009). First, we looked for articles that described longitudinal studies in children with visual impairment and blindness, whether or not DS or a regression in development was mentioned. The following databases were used: PubMed/Ebsco, Medline, Psychinfo and Science Direct. The following keywords in all possible spelling variations were used: longitudinal, developmental setback or regression or delay, and visual impairments or vision disorders or, visual disability, or low vision. Additional references were found by manually checking the reference sections of the articles found in the automatic search process. Articles were included when: (1) the participants were between 0 and 4 years of age; (2) children were congenitally blind/had visual impairment, (3) a longitudinal design was used; (4) assessment instruments were named or described; (5) a setback, regression, stasis or delay was described and/or development was studied longitudinally; (6) they were published in journals and books after 1955; and (7) original empirical data were given. Articles were included as long as they had a longitudinal design, irrespective of whether the age range 16–27 months was covered and irrespective of whether a setback, regression, stasis or delay was mentioned. Selection of articles was performed by the second and third author independently of each other after which they reached consensus about the content of the data. The data were checked for correctness by the first author.

Two literature searches were carried out. A first preliminary search looked for longitudinal studies that specifically investigated a DS in young children who are blind. No other than three studies were found, all by researchers connected to the same institute as Dale and Salt. To complement this search, we looked for more general longitudinal studies without specific mention of regression or delay in children who are blind or visually impaired, because, if the prevalence of a DS is significant, then one would expect this phenomenon to be described in general longitudinal studies on these children as well (see Figure 1).

All publications were analysed for signs of regression, stasis or delay. Regression was defined by the authors as any sign of a loss of earlier acquired skills, stasis as a lack of change in developmental level, and delay as acquiring skills at later ages than peers with
visual impairments. In cases without clear data about regression, stasis or delay but with some mention of difficulties in development or atypical developmental paths, the classification ‘different developmental progress’ was assigned to the results. If possible, the setback was classified as transient or permanent. The principal outcome and summary measures were delay in months or, when given, in developmental quotient. If no exact numbers were given a description of setback or regression was noted.

Results

The literature search yielded three articles with direct mention of a DS: Cass, Sonksen, and McConachie (1994), Waugh, Chong, and Sonksen (1998), and Dale and Sonksen (2002). All of these authors were connected to the Developmental Vision Clinic at the Wolfson Centre of the Institute of Child Health in London. They made use of a retrospective case study design and used overlapping databases. An additional 25 articles describing studies on longitudinal development fulfilled the inclusion criteria. Table 1 shows the results.
### Table 1. Longitudinal studies on children with blindness and visual impairment.

<table>
<thead>
<tr>
<th>#</th>
<th>Reference, country</th>
<th>Participants, ages and length of follow-up</th>
<th>Design and methods</th>
<th>Subject</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Part A: Developmental setback studies</strong></td>
<td><strong>N = 102, Normal development at first assessment with no additional disabilities</strong>&lt;br&gt;– Blind (n = 32)&lt;br&gt;– Blind at first but visual improvement later (n = 21)&lt;br&gt;– Better vision throughout (n = 49)&lt;br&gt;Start ≤16 months until at least 2,5 years</td>
<td>Retrospective case study&lt;br&gt;– Reynell-Zinkin Scales (RZS): Sensorimotor Understanding (SMU); Verbal Comprehension (VC); Expressive Language Structure (ELS)&lt;br&gt;– Parental reports</td>
<td>Cognition, motor, social and language development</td>
<td>Developmental setback (DS) found in SMU, VC and ELS in 11 (11%) children. DS in 31% of children in blind group.</td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>Cass et al. (1994), UK</td>
<td>N = 79&lt;br&gt;– Profound VI (PVI) (n = 45)&lt;br&gt;– Severe VI (SVI) (n = 34)&lt;br&gt;Age at first assessment ≤ 15 months</td>
<td>Retrospective case study&lt;br&gt;Review of MRI (n = 21) and CT (n = 51) scans or both (n = 7)</td>
<td>Brain anatomy</td>
<td>Brain abnormalities found in 50% of the cases. The 7 (9%) of children with DS had significantly more abnormalities.</td>
</tr>
<tr>
<td><strong>Part B: General longitudinal studies</strong></td>
<td></td>
<td>N = 66, 85% were born prematurely. Blind but a large percentage recovered to partially sightedness.&lt;br&gt;Age at onset younger than 15 months up to 6 years &lt;br&gt;3 months intervals up to 27 months of age, 6 months intervals up to the age of 6 years.</td>
<td>Longitudinal, descriptive&lt;br&gt;Qualitative study: opportunity for learning scale: 3 ratings: 1&lt;sup&gt;st&lt;/sup&gt; at onset of study, 2&lt;sup&gt;nd&lt;/sup&gt; after 2 years, 3&lt;sup&gt;rd&lt;/sup&gt; at evaluation.&lt;br&gt;Developmental tests: Cattell, Hayes-Binet, Maxfield Fjeld Social Maturity scale</td>
<td>General development</td>
<td>Regression*</td>
</tr>
<tr>
<td>#</td>
<td>Reference, country</td>
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</tr>
<tr>
<td>5</td>
<td>Fraiberg (1968), USA</td>
<td>Blind (N= 8)</td>
<td>Longitudinal, observational Biweekly video analyses, Developmental tests: Cattell, Vineland Adaptive Behavior Scales, Maxfield Buchholz scale</td>
<td>Mother–child interaction</td>
<td>Regression (transient)</td>
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<td></td>
<td></td>
<td>Age: 3 days to 6 months until 18 months to 6 years. No additional impairments</td>
<td></td>
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<tr>
<td>6</td>
<td>Fraiberg and Adelson (1973), USA</td>
<td>Blind (N= 4), 3 born preterm</td>
<td>Longitudinal, descriptive Biweekly narrative descriptions and video analyses</td>
<td>Mother–child interaction</td>
<td>Regression</td>
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<td></td>
<td></td>
<td>Age: 9 months – 5 years</td>
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</tr>
<tr>
<td>7</td>
<td>Fraiberg (1975), USA</td>
<td>Blind (N= 10), 3 born preterm</td>
<td>Longitudinal, observational Bimonthly video analyses</td>
<td>Characteristics of attachment behaviour</td>
<td>Regression (transient)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Age: 1–11 months until 24 months –6 years</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>Als, Tronick &amp; Brazelton (1980), USA</td>
<td>Blind (N= 1)</td>
<td>Longitudinal, descriptive Biweekly video observation</td>
<td>Face to face interactions and object play</td>
<td>Regression (transient)</td>
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<tr>
<td></td>
<td></td>
<td>Age: 10 days until 15.5 months</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>Gerhart (1982), USA</td>
<td>Blind (N= 1)</td>
<td>Longitudinal, descriptive Video analyses of play sessions with 8 different set of tasks</td>
<td>Development of object play and classificatory skills</td>
<td>Different developmental progress**</td>
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<td></td>
<td></td>
<td>Longitudinal data at 14, 16, and 18 months</td>
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<tr>
<td>10</td>
<td>Andersen, Dunlea &amp; Kekelis (1984), USA</td>
<td>Blind (N= 4)</td>
<td>Longitudinal, descriptive Video/audio recording, diaries of vocabulary development, individualized experiments and developmental assessments</td>
<td>Language development</td>
<td>Delay</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Sighted (N= 2)</td>
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<td></td>
<td>Age: 9 months until 3;4 years</td>
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</tr>
<tr>
<td>11</td>
<td>Kekelis and Andersen (1984), USA</td>
<td>Blind (N= 2)</td>
<td>Longitudinal, descriptive with measurement at two time points</td>
<td>Parent–child interaction</td>
<td>Different developmental progress</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Visually impaired (N= 2)</td>
<td>Analysis of video and audio transcriptions</td>
<td></td>
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<td>Sighted (N= 2)</td>
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<td>Age: 1–3 years, No additional impairments</td>
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<td>Age: 11 until 32 months</td>
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<td>4 children had additional impairments</td>
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Table 1. (Continued).

<table>
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<tr>
<th>#</th>
<th>Reference, country</th>
<th>Participants, ages and length of follow-up</th>
<th>Design and methods</th>
<th>Subject</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>13</td>
<td>Rogers &amp; Puchalsky (1988), USA</td>
<td>Blind (N= 11), Visually impaired (N= 9)</td>
<td>Longitudinal, descriptive</td>
<td>Object permanence Attachment</td>
<td>Different developmental progress</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Age: 4–25 months, 10 born preterm</td>
<td>Bayley Scales of Infant Development, Reynell-Zinkin Scales, Strange situation and measures for object permanence</td>
<td></td>
<td></td>
</tr>
<tr>
<td>14</td>
<td>Preisler (1991), Sweden</td>
<td>Blind (N= 7), 1 born prematurely</td>
<td>Longitudinal, observational and descriptive</td>
<td>Mother–child interaction</td>
<td>Delay</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Visually impaired (N= 3)</td>
<td>Video analyses</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Age: 3–12 months, no additional impairments</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>15</td>
<td>Preisler (1993), Sweden</td>
<td>Blind (N= 9)</td>
<td>Longitudinal, observational</td>
<td>Child’s social activities in a group of sighted children</td>
<td>Regression</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Age: 2–3 years until 6–7 years, 4 born preterm</td>
<td>Detailed descriptions</td>
<td></td>
<td></td>
</tr>
<tr>
<td>16</td>
<td>Tröster et al. (1994), Germany</td>
<td>Blind (N= 10)</td>
<td>Longitudinal descriptive</td>
<td>Gross motor development</td>
<td>Delay</td>
</tr>
<tr>
<td></td>
<td></td>
<td>5 full term, 5 preterm (age corrected for prematurity)</td>
<td>Biweekly visits until the age of 36 months and monthly visits thereafter</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>7.5/16 months to 48 months</td>
<td>Checklist for gross motor development</td>
<td></td>
<td></td>
</tr>
<tr>
<td>17</td>
<td>Ferguson and Buultjens (1995), UK</td>
<td>Blind (N= 16), 5 born premature</td>
<td>Longitudinal, exploratory and descriptive</td>
<td>Play and general development</td>
<td>Different developmental progress</td>
</tr>
<tr>
<td></td>
<td></td>
<td>1.4–6.2 years, No other impairments</td>
<td>During 2 years monthly video analyses of play and Reynell-Zinkin Scales (RZS)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>18</td>
<td>Preisler (1995), Sweden</td>
<td>Blind (N= 7), Deaf (N= 7)</td>
<td>Longitudinal, observational</td>
<td>Parent–child interaction, pre-verbal abilities, exploration of toys, social and symbolic play, communicative intent and sharing of experiences</td>
<td>Delay</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Age: 3–9 months until 6 years, No additional impairments</td>
<td>Transcribed video analyses</td>
<td></td>
<td></td>
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<tr>
<td></td>
<td></td>
<td>No additional impairments, 1 mother was deaf</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>19</td>
<td>Peters (1996), USA</td>
<td>Blind (N= 1)</td>
<td>Longitudinal, descriptive</td>
<td>Collaborative oral story-telling</td>
<td>Different developmental progress</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Age: 23–31 months</td>
<td>Qualitative and quantitative analyses of audio recordings</td>
<td></td>
<td></td>
</tr>
<tr>
<td>#</td>
<td>Reference, country</td>
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</tr>
<tr>
<td>20</td>
<td>Preisler (1997), Sweden</td>
<td>Blind (N= 8), Age: 3 months – 6 years, follow-up at 10 years, No additional impairments, 3 born premature</td>
<td>Longitudinal, descriptive 0–6 years: video recordings in natural interactional settings at home and in (pre) school, Interviews with parents and teachers, Age 10: observations in school, interviews with children, parents, teachers and others involved</td>
<td>Social and emotional development</td>
<td>Regression</td>
</tr>
<tr>
<td>21</td>
<td>Hatton et al. (1997), USA</td>
<td>N= 186, of which 113 (61%) children had only VI; 73 (39%) children had VI + co-occurring Intellectual or Developmental Disability (ID/DD), Vision: 20/70 or worse, 54 (29%) children had light perception at best, Age: 12–73 months</td>
<td>Longitudinal, descriptive Batelle Developmental Inventory (BDI), Individual and group growth curves were drawn with hierarchical linear models (HLM).</td>
<td>Personal-social, adaptive, motor, and cognitive behaviour and communication</td>
<td>Delay</td>
</tr>
<tr>
<td>22</td>
<td>Gosch et al. (1997), Germany</td>
<td>Blind (N= 10), 5 full term, 5 preterm Extremely Low Birthweight (ELBW), 10 months to 58 months</td>
<td>Longitudinal descriptive Annual neurological assessments Every 6 months Bielefeld Developmental Test for Blind infants and pre-schoolers (BDTB)</td>
<td>Neurological status, motor and socio-emotional development, cognition, language, orientation &amp; mobility, social maturity</td>
<td>Delay in 5 ELBW children, Regression in 4 children</td>
</tr>
<tr>
<td>23</td>
<td>Bigelow (2003), Canada</td>
<td>Blind (N= 2) with no additional impairments, Age 1: 13–23 months, Age 2: 21–30 months</td>
<td>Longitudinal, observational Monthly video-analyses, Object search tasks in relation to stages 3–5 of object permanence</td>
<td>Joint attention</td>
<td>Delay</td>
</tr>
<tr>
<td>24</td>
<td>Rattray and Zeedijk (2005), UK</td>
<td>N= 5 dyads, Age child: 6–18 months – Sighted mother and child (n= 1) – Visually impaired mother and sighted child (n= 1) – Sighted mother and visually impaired child (n= 1) – Visually impaired mother and child (n= 2)</td>
<td>Longitudinal, observational Bimonthly video analyses of parent–child interactions during free play.</td>
<td>Parent–child interaction, with a focus on early dyadic interactions (touch, vocalizations and facial orientation)</td>
<td>Different developmental progress</td>
</tr>
</tbody>
</table>

(Continued)
<table>
<thead>
<tr>
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<th>Subject</th>
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</tr>
</thead>
<tbody>
<tr>
<td>25</td>
<td>Brambring (2005), Germany</td>
<td>Blind (N= 4) Included were 4 full term children without additional disabilities from the sample of Tröster et al. (1994)</td>
<td>Longitudinal observations during home visits every two weeks (age 1–3 years) and every four weeks (age 4–6 years) Bielefeld Observation Scales (Brambring, 1999)</td>
<td>Social interactive skills (for gross motor, fine motor/manual skills and verbal development see Brambring, 2006, 2007ab)</td>
<td>Delay</td>
</tr>
<tr>
<td>26</td>
<td>Brambring (2006), Germany</td>
<td>Blind (N= 4) Included 4 full term children without additional disabilities from the sample of Tröster et al. (1994)</td>
<td>Longitudinal observations during home visits every two weeks (age 1–3 years) and every four weeks (age 4–6 years) Bielefeld Observation Scales (Brambring, 1999)</td>
<td>Gross motor development</td>
<td>Delay</td>
</tr>
<tr>
<td>27</td>
<td>Brambring (2007a), Germany</td>
<td>Blind (N= 4) Included 4 full term children without additional disabilities from the sample of Tröster et al. (1994)</td>
<td>Longitudinal observations during home visits every two weeks (age 1–3 years) and every four weeks (age 4–6 years). Bielefeld Observation Scales (Brambring, 1999)</td>
<td>Fine motor/manual/daily living skills</td>
<td>Delay</td>
</tr>
<tr>
<td>28</td>
<td>Brambring (2007b), Germany</td>
<td>Blind (N= 4) Included 4 full term children without additional disabilities from the sample of Tröster et al. (1994)</td>
<td>Longitudinal, observations during home visits every two weeks (age 1–3 years) and every four weeks (age 4–6 years) Bielefeld Observation Scales (Brambring, 1999)</td>
<td>Verbal skills</td>
<td>Delay</td>
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</table>

*Regression = loss of earlier acquired skills; **Different developmental progress = no clear description of delay or regression but a different developmental path is mentioned; ***Delay = acquiring skill at a later age than sighted peers.*
The overall picture arising from Table 1 is that delay in development is far more common than regression, especially in play, language, cognition, adaptive behaviour and motor development. The picture is mixed with regard to parent–child interaction, attachment and social and emotional development. Here both regression and delay are mentioned.

**Developmental Setback Studies**

Developmental setback was defined by Cass et al. (1994) as a stasis or regression of raw scores in one or more of three scales of the Reynell-Zinkin Scales (RZS): Sensorimotor Understanding, Verbal Comprehension and Expressive Language; sustained over at least two consecutive assessments and/or an increasing disorder in social communication (no assessment schedules mentioned). Cass's sample included 102 children with visual impairment and blindness. Dale and Sonksen (2002) were more specific about DS. At least two scales of the RZS, including the sensorimotor understanding scale, had to be in the normal range at first assessment between 10 and 16 months of age. The term DS was used when at time 2 (between the ages 27 and 54 months) there was a loss of 30 or more Developmental Quotient (DQ) points to a DQ level of 70 or lower on one or more scales. The setback in behaviour and social communication was not included in their definition. The sample of Dale and Sonksen included 69 children with 'simple' CDPVS. Waugh et al. (1998) looked at the same scales of the RZS as Cass et al. (1994) and defined DS as stasis or regression of raw scores on one or more scales sustained over two or more consecutive assessments during a time period of 15 or more months.

For the total group of children who were blind or had visual impairment the number of children with DS ranged from 9% (n = 7) in Waugh et al. (1998) to 11% (n = 11) in Cass et al. (1994), and 22% (n = 15) in Dale and Sonksen (2002). Children who were blind or had PVI dominated this group. In Cass et al. 10 out of 11 were blind throughout, that is their visual status did not change (i.e. improve) during the study (which is 31% of the children with blindness). In the sample of Dale and Sonksen (2002) nine children had profound visual impairments and DS (33% of all PVI children), while five children progressed from PVI to SVI during the measurement time and only one child had SVI. In the sample of Waugh et al. (1998) all children with DS had PVI (25% of all PVI children). These data suggest that the degree of visual impairments seems to be associated with setback. Most children with DS are blind or have PVI. The range of visual diagnoses in children with developmental setback showed this group to be clinically and pathogenically heterogeneous. Therefore, conclusions about the relationship between aetiology and DS cannot be made.

Setback was reported in sensorimotor and language development (both verbal comprehension and expression), explorative and manipulative play, social communication and behaviour in general. The greatest decelerations were in language comprehension and sensorimotor skills. The syntactical and structural aspects of language expression were superficially preserved. Partial recovery of the setback was however mentioned by Cass (1994): four out of the eleven children recovered cognitively. However, in three of these four children social and communication problems persisted. Cass et al. (1994) and Dale and Sonksen (2002) reported only two measurements on the RZS, so no empirical statements about reversibility can be made.
Although children had no obvious additional disabilities, neurological abnormalities were reported by Waugh et al. (1998). MRI or CT scans of 79 children were available and 40 (50%) showed abnormalities. The seven children with DS showed significantly more brain abnormalities than the children without setback. Social adversities were described as risk factors. Cass et al. (1994) mentioned that hospitalisation, marital discord, maternal depression, financial problems were found in 60% of the children with a developmental setback, in contrast to only 23% of the children with typical outcome.

**Developmental Setback in General Longitudinal Studies**

The longitudinal studies in Table 1, part B were checked for descriptions of a setback in the development of children with congenital blindness or visual impairment. Regression or delay was described in the 25 studies, but none of the studies described stasis. Eight studies, of five different groups, mentioned a regression in the children’s development (Als, Tronick, & Brazelton, 1980; Fraiberg, 1968, 1975; Fraiberg & Adelson, 1973; Gosch, Brambring, Gennat, & Rohlmann, 1997; Norris, Spaulding, & Brody, 1957; Preisler, 1993, 1997) and developmental delays were mentioned in 12 studies. Seven of these studies mentioned a delay in acquiring specific social interaction skills, such as communication, language, and joint attention (Andersen, Dunlea, & Kekelis, 1984; Bigelow, 2003; Brambring, 2005, 2007b; Preisler, 1991, 1995; Rowland, 1984). Nine studies described delays that were adaptations to the blindness, such as a different developmental progress in parent–child interaction, play, gross and fine motor skills (Gerhardt, 1982; Brambring, 2006, 2007a; Ferguson & Buultjens, 1995; Kekelis & Andersen, 1984; Peters, 1996; Rattray & Zeedijk, 2005; Rogers & Puchalski, 1988; Tröster, Hecker, & Brambring, 1994). Lastly, Hatton et al. (1997) and Brambring (2005) described an overall delay in development for all children with blindness. Due to the research design of these latter two studies, only group means or median scores were mentioned and not data on individual children with delays. The number of individual children with delays was therefore not known.

The eight studies that found regression followed the children over a period of months or several years. These studies, mostly detailed observations and analyses of bi-weekly video recordings, were descriptive and broad in their focus, covering all aspects of development. Regression was described extensively but was not clearly defined in most studies nor operationalised as loss of (raw) scores on a developmental scale. The authors did not use the term DS but instead called it ‘regression in behaviour’ that may refer to the same phenomenon.

In the studies of Als et al. (1980), Fraiberg (1968), and Fraiberg and Adelson (1973) regression was defined from a psychodynamic theoretical perspective, referring to a temporary psychological regression to an earlier stage of ego development. For instance, Fraiberg (1968) described periods of desperate clinging to the mother and panic during actual or anticipated separation in ‘each of the blind children as they moved into the second year (p. 288)’. Although this behaviour can also be seen in sighted children of this age she also described other examples of regression in five children that looked more alarming, ‘a transient pathological state’ (p.288), which expressed itself in different forms, for instance: stuporous sleep or other disturbances in the circadian rhythm, screaming fits which lasted for hours, a change in personality, a regression in all aspects of development, the onset of stereotyped behaviours. According to Fraiberg (1968) this period of regression in children ‘who have
shown good overall development’ (p. 277) is due to a delay in young children with blindness in the formation of object and person permanence. Fraiberg and Adelson (1973) described four children with blindness of seemingly normal development. One boy was described with a disordered personality with frequent regressions to echolalic speech. Because there was no evidence of neurological damage, Fraiberg presumed that blindness itself imposes extraordinary impediments on the development of a valid self-image in this child.

Als et al. (1980) described a crisis in a two-month-old baby who was blind. The baby was suddenly restless, upset and difficult to console. For this child, periods of consolidation and balance were followed by periods of disorganisation and seeming regression. According to Als et al. (1980), this happened when the child was adapting to a new developmental stage and reorganising his behaviour. From a dynamic systems perspective, this would be viewed as a period of disorganised behaviour preceding a sudden jump in development, common in all children, and nowadays seen as completely normal.

Norris et al. (1957) confirmed the impression that for children who are blind development in the second and third year of life is at risk. Although at first very optimistic about the developmental possibilities of children with blindness, they described the period during the second and third year of life as ‘one of the most difficult periods for a blind child and his family: a so-called negativistic period’ (p. 55), when problems intensify.

Preisler (1993) observed nine children with blindness in a nursery with sighted children. Two children (both 2 years of age) displayed marked physical and mental regression during their first year at the nursery. They started banging and twisting toys in a stereotypical way and showed a particular interest in auditory stimulation (e.g. the sound of the washing machine). It is unknown whether this regression was permanent or not. In a later study Preisler (1997) studied eight children with blindness between 3 months and 10 years of age. Of the eight children, diagnosed as cognitively typically developing during their first 18 months, three were later diagnosed with autism or identified as displaying behaviours associated with autism. The other five children displayed considerable variation in emotional, social and cognitive development (Preisler, 1997). Gosch et al. (1997) reported that two preterm and two full-term children out of a group of 10 children with blindness showed deterioration of neurological status between 10 and 58 months of age. The problems in language and communication, the occurrence of stereotyped behaviour, and the behavioural problems of the children with blindness that had a setback had parallels with ASD. This is described by, amongst others: Preisler (1993, 1997), Sonksen and Dale (2002), Dale (2005), Dale and Salt (2008), and Dale, Tadic, and Sonksen (2013).

Age of Onset and Permanency of Developmental Setback

In the eight studies that mentioned regression, this occurred at various ages. Only three studies covered the age period from 16 to 27 months (Fraiberg, 1975; Fraiberg & Adelson, 1973; Preisler, 1997). Two studies described a regression before 16 months of age (Als et al., 1980; Fraiberg, 1968). Three studies mentioned a regression after 27 months of age (Gosch et al., 1997; Norris et al., 1957; Preisler, 1993). Hence, it appears that the presumed age range of the regression, 16–27 months, could actually be much longer.

The outcomes for the children in the studies of Norris et al. (1957), Fraiberg (1968, 1975), Als et al. (1980), and Preisler (1993) are unclear, but a transient regression is presumed given
the terms used by the authors: ‘periods of disorganisation’ (Als et al., 1980); ‘transient regressive state caused by panic during separation’ (Fraiberg, 1968, p. 290) ‘separation distress’ (Fraiberg, 1975, p. 330) and a ‘negativistic period’ (Norris et al., 1957, p. 55). Only two studies reported permanent regression (Fraiberg & Adelson, 1973; Preisler, 1997) in some children. It is unclear whether the regression in neurological status at 58 months mentioned by Gosch et al. (1997) started earlier or was permanent. Note that in cases of transient regression described in Table 1 mostly the children’s emotions and behaviours changed not their motor, language or cognitive skills.

Discussion

The main research question was whether there is empirical evidence for the existence of a setback in development in young children who are congenitally blind. Up till now only three articles, written by researchers from one institute, the Wolfson centre in London, showed DS in children who are blind. DS was found in 9–21% of the children, of which the children with PVI showed setbacks in 25–33% of cases. Although the three studies stemmed from only one setting, these researchers had access to substantially large databases and their research design allowed for individual developmental trajectories to be followed. The retrospective DS studies all used the Reynell-Zinkin Scales. A setback was found for cognition and verbal comprehension. This does not mean that DS is only present for language and cognition, since the Social Adaptation and Exploration of the Environment scales of the RZS were not administered.

The review of the longitudinal studies showed that in addition to problems with language and cognition a setback was found with regard to disorganised and stereotyped behaviour and other autistic-like behaviour. DS was not permanent for all children, since some of them recovered, also from the more autistic-like behaviours. Whether this is also the case for the studies done at the Wolfson centre is unknown, except for the children in the study of Cass et al. (1994). They found that 4 out of 11 children recovered but social and communication problems were more permanent.

The reviewed articles met with some methodological problems that force us to remain cautious in drawing firm conclusions. Firstly, existing samples of children and not random samples were drawn and the study samples were, with the exception of Cass et al. (1994), Dale and Sonksen (2002), and Hatton et al. (1997) rather small. The study with the largest sample (Hatton et al., 1997) mentioned group mean and median scores and showed no DS, but these data make it impossible to analyse individual developmental trajectories. It only shows that on average there is no DS. Secondly, most studies in the review used a retrospective design and were not set up to capture possible DS. As a result, the presumed time of onset of the setback – somewhere between 16 and 27 months of age – or any other time of onset could not be determined exactly. For that matter, Cass et al. (1994), Waugh et al. (1998), and Dale and Sonksen (2002) recognised the limitations of retrospective and applied clinical research designs and stressed the desirability of prospective research designs in future studies. Thirdly, there is some concern with the RZS as to whether these scales are valid and sensitive enough to measure behavioural change in the first years of life (see Cass et al., 1994; Cass, 1998; Waugh et al., 1998; Dale & Sonksen, 2002.; Vervloed, Hamers, van Mens-Weisz, & Timmer-van de Vosse, 2000). Lastly, the studies included in this literature review
were rather dated. The three studies that identified DS (part A in Table 1) were published 15–23 years ago (M = 19 years). The 25 longitudinal studies (part B in Table 1) were published 10–60 years ago (M = 26.5 years). Using such dated publications for a current literature review has certain risks. For instance, child rearing and caretaking practices, education, and medical care have changed considerably in the last 60 years, as well as our views on child development. The behaviours described by Fraiberg (1968) and Als et al. (1980) are now interpreted differently. Although the assessment practices of today did not change much with regard to the diagnostic instruments used, it is quite likely that other assessment practices and early intervention methods are very different from those utilised in practice and research 10–60 years ago. We believe this does not make the results reported in this literature review obsolete, since in clinical practice of today we still meet children who are blind or have visual impairment that show DS. The lack of recent data is a strong argument for conducting new research on the topic, but this time with prospective longitudinal designs, ample measurement points, and valid and sensitive assessment instruments.

Degree of visual impairment, neurological abnormalities, home life, and social adversities were mentioned as risk factors. What is not mentioned in most of the reviewed articles is the possible effects of environmental factors such as child-rearing practices – both positive and negative-, early intervention programmes and VI-preschool programmes. Although a DS occurred in some children with severe visual impairments, the majority of children who showed a setback were congenitally blind or had a profound visual impairment, that is visual acuity insufficient for perceiving objects, forms or contours. Another factor warranting mention is prematurity, which is common in children with blindness. Prematurity was insufficiently specified and considered in most of the study samples. Studies of very preterm children have shown however that development and learning are at risk and that psychiatric problems are quite common (e.g., Johnson et al., 2010). Sequelae stemming from premature birth have therefore to be considered as an alternative explanation for DS.

Given the possibility that brain dysfunction is in part responsible for a DS it could also be that DS may be a manifestation of a phenomenon already mentioned by Warren in 1994: ‘It may be that incipient neurological problems can go undiagnosed in children in the midst of concerns about the visual impairments itself’ (p. 315). Given the difficulty of measuring small changes in development, the insensitivity of assessment instruments like the RZS, and the fact that most retrospective studies had insufficient measurement points before 16 months of age, one cannot rule out the possibility of undiagnosed signs of neurological dysfunction.

In addition to social adversity and neurological dysfunction, a delay or different trajectory in psychosocial development and social interaction could contribute to DS. Congenital blindness has an impact on the caregiver–child interaction; on the development of intersubjectivity, attachment, the separation-individuation process, and on joint attention (Warren, 1994; Webster & Roe, 1998). The above variables do have unique contributions to behaviour and the variation in the studies that were reviewed cannot take these specific variations into account, unless the children are studied in a more controlled and nurturing environment and in a longitudinal design, so one can control for these mitigating factors.
The observed setback and regression in behaviour and communication in the second and third year of life may have had silent precursors in the behaviour of the infant and/or the parent–child relationship. Delayed or atypical developmental trajectories can put children who are blind under stress at the age of 18 months. At this age, when sighted children start crawling and walking, and start to explore the world at greater distances from their parents, the child with blindness still has a strong need to be close to his parents for comfort, security and help (Fraiberg, 1968; Preisler, 1993; Preisler & Palmer, 1989). Visiting a nursery at this age can put too much pressure on the child to become self-sufficient. The child might find itself in situations that are unpredictable, confusing or overwhelming. Still, today most nurseries are ill-equipped to focus on the needs of a child who is blind. In the beginning, the child is overwhelmed by all the different sounds (other children playing, strange caretakers talking, sounds of the washing machine, opening and closing doors) which make the environment unsafe and unpredictable. These kinds of sensory issues cannot be understood by typical early childhood employees. They may not know how to explain background noises. It is important for caretakers to realize that it takes months for a child who is blind to get used to a nursery or preschool. On the rebound, the child shows a regression in development and behaviour in these circumstances; it may react with (sometimes very) disturbed behaviour that can be difficult to read while not showing this behaviour at home.

So, apart from explaining noises, textures and smells, adjusting the environment and the daily routines of the nursery to the needs of a blind child is crucial. It is important, for example, to give the child the opportunity to gradually get accustomed, initially in the presence of one of his parents who can ‘translate’ its behaviour and habits to the caretaker. It is also important to allocate one particular caretaker to the child, to decrease the size of the group of children and to limit the rooms it is in. These are all crucial prerequisites to help the child to cope with and adjust to its new environment. These kinds of issues, however, are not easily integrated in the daily practice of the nursery.

The relationship between DS and Autism Spectrum Disorder (ASD) was frequently discussed in the discussion sections of the reviewed DS studies and also in some of the longitudinal studies. The problems in language and communication and the behavioural problems of the children with blindness that had a setback seem to have clear parallels with ASD. This raises the question whether DS might be due to or a precursor of ASD. We speak of precursor, and not of ASD itself, because ASD is normally not diagnosed before the age of three. These aforementioned behaviours, however, are not exclusively seen in children who experienced a DS. ASD in children with blindness is a much-disputed subject (see Begeer, Dik, Voor de Wind, Asbrock, Brambring, & Kef, 2014; Brown, Hobson, Lee, & Stevenson, 1997; Cass, 1998; De Vaan et al., 2016a; 2018; Hobson, Brown, Minter, & Lee, 1997; Hobson, Lee & Brown., 1999; Hobson & Bishop, 2003; Pring, 2005; Hobson & Lee, 2010; Jure, Pogonza, & Rapin, 2016). All children with blindness show one or more stereotyped movements (Fazzi et al., 1999; Tröster, Brambring, & Beelmann, 1991), and language and communication characteristics associated with autism are observed in many children with blindness (Pérez Pereira & Conti-Ramsden, 1999). Note, however, behaviours like joint attention and tests for theory of mind rely heavily on visual behaviours on the part of the child; behaviours that are, for obvious reasons, at risk in children with visual impairment and blindness. Because of this, these children have often been (mis)diagnosed when showing (temporary) ASD features (De Vaan et al., 2016a, 2016b, 2018).
The question, whether a DS is a precursor for ASD, cannot be answered with the presently available knowledge. The studies on ASD and blindness, unfortunately, do not describe the history of ASD in relation to the children’s development and a potential DS. These studies are mainly cross-sectional, with one follow up measurement at the most, and often focus on children at ages older than the age period in which the presumed setback occurs (see, e.g., Hobson et al., 1999; Jure, Pogonza, & Rapin, 2016).

In conclusion, the period between the second and third year of life seems to be a vulnerable period for children with blindness. Out of 25 longitudinal studies, 8 (32%) reported regression in both development and behaviour, and the DS studies indicate DS in 30% of the children with blindness. A substantial overlap with ASD is described. Children with neurological abnormalities appear to be most at risk, presumably in interaction with social adversities and disturbances in psychosocial development. As yet individual variation in maturation and development and methodological issues complicate conclusive statements about the existence of a DS in children with blindness. Further research into this phenomenon and its moderating and mediating factors is crucial. To capture this phenomenon, children with congenital blindness need to be monitored individually and prospectively in a longitudinal research design from birth to 6 years of age. Psychosocial development needs to be closely monitored, as well as cognitive and physical development. Assessments need to be carried out at close intervals, using valid instruments that are sensitive to small changes in development, in order to be able to draw conclusions about the duration and reversibility of the setback. Samples must be large and representative of the total group of children with congenital blindness or visual impairment to prevent sampling bias, and to be able to generalise the results.

An atypical developmental path can be adaptive and functional, but we must not close our eyes for disrupting roadblocks. For clinical practice, it is very important that signs of disturbed behaviour and DS are taken seriously and analysed properly with the above-mentioned risk factors in mind. Given the fact that many children with VI and blindness receive early intervention, and it is unethical to refrain from early intervention, the intervention itself can be a means to study a possible setback as well. In order to do that the intervention should be part of a controlled programme in which caregivers and child care workers are helped in understanding the signals of the children and encourage them to learn about the sounds and smells, and objects around them on a daily basis, so they learn to comprehend the world. But also, by giving them psychomotor opportunities and offering them ordinary experiences daily so they can develop more closely to their typically developing peers. Which caregivers and children pick up these interventions and which do not might help us to understand which of the children will develop instances of DS. Visual impairment and blindness are low incidence disabilities, making it hard to set up studies with good designs. It would, therefore, be very helpful if clinicians would systematically gather developmental data, especially around the period of 18–30 months of age, and make these data available for researchers. At the same time, early interventionists working with children at risk in general in areas such as education, baby well clinics or health care might want to know how to interpret the behaviours of children who are possibly visually impaired. For instance, how one can read their emotions, how to tune in, and how to set up joint attention in the absence of visual behaviours?
Acknowledgements

We want to thank Anke de Jong and her colleagues of the library of Bartiméus for their help with the literature searches, Naomi Dale and Michele O'Reilly for sharing their findings with us, and Jan Rodney for editing the final text.

Disclosure statement

No potential conflict of interest was reported by the authors.

Funding

This work was supported by the novum foundation [NA];

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References


**Appendix 1: Glossary and Abbreviations**

**ASD** = Autism Spectrum Disorder

**CDPVS** = Congenital Disorders of Peripheral Visual System. Peripheral means including the eye globe, retina, or anterior optic pathway

**CDPVS pc** = potentially complicated and CDPVS ps = potentially ‘simple’ (not part of neuro-ophthalmic disorders)

**DQ** = Developmental Quotient

**DS** = Developmental Setback

**Form vision**: visual perception of visual objects or contours

**LP** = Light Perception, no form vision but perception of light sources

**PVI** = Profound Visual Impairment (no form vision, Light Perception or worse)

**RZS** = Reynell-Zinkin Scales, consisting of: Sensori-Motor Understanding, Exploration of the Environment, Social Adaptation, Verbal Comprehension, Expressive Language, Structure, Expressive language, Content

**SVI** = Severe Visual Impairment (form vision or better)

**VI** = visually impaired/visual impairment