Short-term memory in children with sickle cell disease: Executive versus modality-specific processing deficits

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Accepted 7 June 2005

Abstract

Prior research has identified a number of areas of cognitive deficit among children with sickle cell disease (SCD), including decrements in memory span and working memory. The present study examined short-term memory span and working memory performance among children with SCD (n = 25) and demographically matched comparison children (n = 25) using digit span, spatial span, and the self-ordered pointing test. Children with SCD showed difficulties only for digit span-backward. Additional cognitive ability measures administered indicated auditory processing was an area of deficit related to digit span-backward performance. The study suggests that modality specific deficits are one factor in short-term memory span for children with SCD. The cause of this deficit is unclear, but may involve both central and peripheral components of auditory processing.

Keywords: Sickle cell disease; Cognitive; Memory

Sickle cell disease (SCD) is an inherited hemoglobinopathy resulting in a variety of complications including hemolytic anemia, decreased oxygen carrying capacity of the blood supply, and susceptibility to vaso-occlusion (for review see Ris & Greunich, 1999). Cerebral infarction is a recognized cause of cognitive deficits in SCD (Armstrong et al., 1996; Bernaudin et al., 2000; Brown et al., 2000; Craft, Schatz, Glauser, Lee, & DeBaun, 1993). Two recognized forms of cerebral infarction are: (a) overt strokes, which typically involve large vessel disease and more tissue injury and (b) silent cerebral infarcts, which typically involve a smaller amount
of tissue injury typically in subcortical white matter and/or the striatum (Moser et al., 1996). Silent cerebral infarcts are a more recently recognized form of brain injury in SCD (Armstrong et al., 1996; Craft et al., 1993). Silent cerebral infarcts are abnormal neuroimaging findings that are consistent with cerebral infarction in an individual that otherwise has a normal neurologic history and normal neurologic exam (Glauser, Siegel, Lee, & DeBaun, 1995).

Recent studies have indicated that children with SCD that have no evidence of cerebral infarction show at least small decrements (typical effect size of $r = .15$) on IQ measures relative to comparison groups, and medium-size decrements (typical effect size of $r = .34-.43$) on tests of specific cognitive functions (Schatz, Finke, Kellett, & Kramer, 2002; Steen et al., 2003b, 2005). These findings suggest that there are more diffuse neurocognitive effects of SCD beyond the presence of focal cerebral vascular injury. Studies to date have indicated similarities in the profile of cognitive deficits across children with overt stroke, silent cerebral infarctions, and no visible tissue injury. Verbal ability, higher-level attention, and executive skills are common areas of deficit across all three of these groups (Brown et al., 2000; DeBaun et al., 1998; Schatz et al., 1999, 2002). The severity of the deficits varies among these groups with overt stroke groups showing the largest deficits and groups with SCD and normal brain magnetic resonance imaging (MRI) showing the smallest deficits (Armstrong et al., 1996; Bernaudin et al., 2000; Wang et al., 2001).

Frontal brain injury in SCD, which is the most common location of injury (Brown et al., 2000), has been related to reduced efficiency in rehearsing verbal information in working memory, difficulties with manipulating verbal information in working memory, and poorer retrieval of verbal information on memory recall trials (Brandling-Bennett, White, Armstrong, Christ, & DeBaun, 2003; White, Saloria, Schatz, & DeBaun, 2000). These data suggest specific learning mechanisms associated with frontal injury that would impact academic learning and other functional outcomes for most children with stroke from SCD. Prior studies have provided mixed results, however, as to whether and how memory functions are impacted by SCD in the absence of stroke. Studies assessing specific measures of short-term or working memory in children with SCD and no known cerebral infarcts have reported mixed outcomes with both decrements in children with SCD (Gilbert, 1970; Knight, Singhal, Thomas, & Serjeant, 1995; Noll et al., 2001; Schatz, Finke, & Roberts, 2004; Swift et al., 1989; Wasserman, Wilimas, Fairclough, Mulhem, & Wang, 1991) and null findings (Bernaudin et al., 2000; Brown et al., 1993; Craft et al., 1993; Fowler et al., 1988).

There are at least two factors that may account for this discrepancy across studies. First, the methods of identifying cerebral infarction have varied across studies. Most of the studies identified above have used a history of overt stroke as the primary method for excluding children with cerebral infarction. This method misses a substantial number of children with SCD and silent cerebral infarction (Moser et al., 1996). This factor may account for the mixed pattern of results with studies including different numbers of children with silent cerebral infarcts in the sample believed to be without focal brain injury. Steen et al. (2003b) examined both anemia severity (hematocrit) and the presence of cerebral infarction on structural MRI as factors predicting cognitive functioning. This work found that hematocrit was an independent factor that predicted lower performance on measures of short-term and working memory from the Wechsler Intelligence Scales, third edition (i.e., digit Span, arithmetic subtests). The relationship between hematocrit and short-term memory performance is consistent with the
view that diffuse effects of anemia may be causing these cognitive effects rather than the effect solely occurring because of silent cerebral infarction (Steen et al., 2003b). A second factor that may be impacting the findings in this area has been the types of measures used. Studies have predominantly used auditory–verbal measures to assess short-term and working memory functions. There has been a predominant use of tests such as digit span, word span, and word list learning tests to assess memory functions in SCD with very few attempts to use comparable visual-nonverbal measures. Few studies have attempted to measure auditory processing in a more isolated manner, but there is some suggestion that this may be an area of deficit for some children with SCD (Steen, Hu, Elliott, Miles, Jones, et al., 2002). In addition, studies using the digit span test have typically reported the overall score rather than reporting digit span-forward and digit span-backward separately.

The purpose of the present study was to assess short-term memory span and working memory functions in children with SCD using tests that require both auditory and visual processing. Digit span and spatial span measures were used to assess short-term memory span using comparable auditory–verbal and visual-spatial tests. In addition, the Self-ordered Pointing Test (SOPT) was used to assess short-term and working memory for representational (easy to encode verbally) and nonrepresentational (difficult to encode verbally) stimuli (Petrides & Milner, 1982; Spreen & Strauss, 1998). The SOPT assesses working memory, organizational strategy, and response monitoring; it has been shown to be dependent on intact prefrontal cortex and basal ganglia circuits (Petrides and Milner, 1982; Spreen and Strauss, 1998). We hypothesized that children with SCD would show greater difficulties on tasks requiring more executive control (i.e., backward span tasks, interference effects on the SOPT). The relationship between short-term memory performance and other areas of cognitive ability was also explored to better understand the nature of short-term memory difficulties in children with SCD.

1. Methods

1.1. Participants and recruitment

Twenty-five children with SCD (20 with HbSS, 3 with HbSC, 2 with HbS beta-thalassemia) from regional pediatric hematology clinics for SCD were recruited for this study as part of an ongoing screening program to provide baseline cognitive assessment information for children with SCD. Hematocrit levels from the most recent hematology clinic visit (within 8 weeks of the testing session) ranged from 20.3 to 37.3% with a mean of 27.5% and a standard deviation of 5.0. Twelve of the 25 SCD participants had been admitted to the hospital at least once in the past year due to SCD-related complications. None of the children with SCD were receiving transfusion therapy or oral hydroxyurea treatment at the time of testing, reported feeling well on the day of testing, and had normal neurologic screening exams at their most recent clinic visit. A review of medical records and a parent interview conducted after the cognitive testing sessions indicated nine of the children with SCD had a history of neurologic symptoms. Eight had a history of severe headache. Three had a history of a transient ischemic attack with subsequent normal brain MRI and magnetic resonance angiography (MRA). Among the six children with a history of severe headache five of the six had normal neuroimaging exams
reported in their medical records and the remaining child had not received any neuroimaging exams.

Twenty-five children without a history of a severe or chronic health condition (per parent report) were recruited to match the SCD group for age, household income, and parental education levels. These children were recruited through elementary schools and after-school programs in the same local communities (see Table 1). All parents and children participating in the study identified themselves as being of African–American ethnicity. All parents provided informed consent for research and all children provided assent for participation. Each child’s history of birth complications, neurologic symptoms, and learning difficulties in school was assessed via parent report. Learning difficulties were defined by either needing special services at school for learning problems or a previous diagnosis of a learning disability by a professional. Rates of learning problems in each sample were comparable to that expected in each population according to previous population estimates (see Table 1; Reschly, 1996; Schatz, 2004).

1.2. Cognitive measures

1.2.1. Short-term memory span

The digit span and spatial span subtests of the Wechsler Intelligence Scale for Children-Processing Instrument (WISC-PI; Kaplan, Fein, Kramer, et al., 1999) were administered to examine short-term memory span for auditory–verbal and visual-spatial material. Separate age-

<table>
<thead>
<tr>
<th>Demographic variable</th>
<th>SCD group</th>
<th>Comparison group</th>
<th>Statistic</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (years)</td>
<td>M = 11.4; S.D. = 2.8 (range 6.9–16.3)</td>
<td>M = 11.1; S.D. = 1.7 (range 7.0–16.0)</td>
<td>t = 0.43</td>
</tr>
<tr>
<td>Gender ratio (male:female)</td>
<td>14:11</td>
<td>11:14</td>
<td>χ² = 0.72</td>
</tr>
<tr>
<td>Household income</td>
<td></td>
<td></td>
<td>χ² = 0.84</td>
</tr>
<tr>
<td>$10,000–20,000</td>
<td>10 (40%)</td>
<td>8 (32%)</td>
<td></td>
</tr>
<tr>
<td>$20,000–30,000</td>
<td>7 (28%)</td>
<td>10 (40%)</td>
<td></td>
</tr>
<tr>
<td>$30,000–40,000</td>
<td>6 (24%)</td>
<td>5 (20%)</td>
<td></td>
</tr>
<tr>
<td>$40,000–50,000</td>
<td>1 (4%)</td>
<td>1 (4%)</td>
<td></td>
</tr>
<tr>
<td>More than $50,000</td>
<td>1 (4%)</td>
<td>1 (4%)</td>
<td></td>
</tr>
<tr>
<td>Parental education level</td>
<td></td>
<td></td>
<td>χ² = 0.94</td>
</tr>
<tr>
<td>Less than 12 years</td>
<td>6 (24%)</td>
<td>5 (20%)</td>
<td></td>
</tr>
<tr>
<td>High school diploma</td>
<td>10 (40%)</td>
<td>11 (44%)</td>
<td></td>
</tr>
<tr>
<td>Some post secondary</td>
<td>6 (24%)</td>
<td>7 (28%)</td>
<td></td>
</tr>
<tr>
<td>College degree</td>
<td>3 (12%)</td>
<td>2 (8%)</td>
<td></td>
</tr>
<tr>
<td>History of pre-term birth or birth complications</td>
<td>4 (16%)</td>
<td>3 (12%)</td>
<td>χ² = 0.17</td>
</tr>
<tr>
<td>History of neurologic symptoms</td>
<td>9 (36%)</td>
<td>3 (12%)</td>
<td>χ² = 5.95</td>
</tr>
<tr>
<td>History of learning problems in school</td>
<td>8 (32%)</td>
<td>3 (12%)</td>
<td>χ² = 2.91</td>
</tr>
</tbody>
</table>

Note: see Section 1 for definitions of demographic variables.

* P < .05.
adjusted norms for the forward and backward recall conditions of these subtests are provided in the WISC-PI norms.

1.2.2. Working memory and strategic memory

A version of the Self-Ordered Pointing Test was administered to assess additional aspects of short-term memory. Three sets of visual stimuli were used with each set administered two times in succession to create interference effects on the second administration of the set. The first two sets of stimuli were designed to be easy to name (easy to encode verbally) and the third set was designed to be difficult to name (difficult to encode verbally). The first set of stimuli consisted of six line drawings of common objects (e.g., light bulb, car). The second set consisted of eight colored rectangles approximately 3 cm square (e.g., dark blue, light blue, dark green, light green, etc.). The third set of stimuli consisted of ten nonrepresentational patterns of lines and dots within 3 cm square rectangles. Instructions were provided to select each item in the stimulus set only once during a series of trials. Item location on the page of stimuli changed with each trial. Instructions were repeated with additional demonstration if any errors were made during the first series of trials. The total number of errors (i.e., selecting a previously chosen item within a series of trials) was the variable of interest.

1.2.3. Other measures

A battery of tests to assess different domains of cognitive ability based on the Cattell–Horn Gf-Gc model was administered for descriptive purposes (for review see McGrew & Flanagan, 1998). The tests were selected based on their empirical support as measures of distinct, theoretically-derived areas of cognitive abilities (based on the Cattell–Horn Gf-Gc model) and the appropriateness of the measures for this population based on cultural and linguistic factors (McGrew & Flanagan, 1998; Schatz, 2004; Schatz et al., 2004). The majority of these measures were from the Woodcock–Johnson Psycho-Educational Battery-Revised (WJ-R; Woodcock & Johnson, 1989). The WJ-R measures were Spatial Relations, Oral Vocabulary, Visual Closure, Incomplete Words, and Visual Matching. The Category Fluency test from the Delis–Kaplan Executive Functions Scale was also administered (D-KEFS; Delis, Kaplan, & Kramer, 2001). WJ-R Spatial Relations is a test that taps into both fluid ability and visual processing (McGrew & Flanagan, 1998). We used this as a measure of fluid ability because we have found that the measure works well with a wider age range of children than alternative measures on the WJ-R. The remaining measures load more specifically on single Gf-Gc domains and tap into crystallized ability, visual processing, auditory processing, processing speed, and long-term retrieval, respectively (McGrew and Flanagan, 1998).

1.3. Statistical methods

Descriptive data for demographics and broad areas of cognitive ability were evaluated with independent samples t-tests or chi-square tests to assess for group differences. Performance on the span tests was evaluated with a mixed factor repeated measures multivariate analysis of variance (MANOVA) procedure. Age-adjusted span scores were organized according to two within subjects factors (stimulus type: digits vs. spatial locations; stimulus manipulation: forward vs. backward span) and group was the between subjects factor (SCD group; demo-
Table 2

Mean scores (standard deviation) for descriptive measures of cognitive abilities

<table>
<thead>
<tr>
<th>Test (Gf-Gc domain)</th>
<th>SCD group (n = 25)</th>
<th>Comparison group (n = 25)</th>
<th>t(48)</th>
</tr>
</thead>
<tbody>
<tr>
<td>WJ-R Spatial Relations (fluid ability/visual processing)</td>
<td>93.8 (10.8)</td>
<td>99.2 (13.4)</td>
<td>1.55</td>
</tr>
<tr>
<td>WJ-R Oral Vocabulary (crystallized ability)</td>
<td>87.7 (13.4)</td>
<td>96.2 (15.7)</td>
<td>2.07</td>
</tr>
<tr>
<td>WJ-R Visual Closure (visual processing)</td>
<td>97.2 (16.9)</td>
<td>100.6 (14.0)</td>
<td>0.78</td>
</tr>
<tr>
<td>WJ-R Incomplete Words (auditory processing)</td>
<td>84.9 (16.0)</td>
<td>95.6 (12.8)</td>
<td>2.62 *</td>
</tr>
<tr>
<td>D-KEFS Category Fluency (long-term retrieval)</td>
<td>102.1 (13.3)</td>
<td>103.0 (16.6)</td>
<td>0.20</td>
</tr>
<tr>
<td>WJ-R Visual Matching (processing speed)</td>
<td>90.4 (15.5)</td>
<td>102.1 (16.0)</td>
<td>2.61 *</td>
</tr>
</tbody>
</table>


* P < .05.

Descriptive statistics for demographic characteristics are shown in Table 1 and the data for the broader cognitive profile of the participants are shown in Table 2. The two groups appeared to be well matched on a range of background variables. The group with SCD showed lower scores on the Oral Vocabulary, Incomplete Words, and Visual Matching tests.

For the span test the analyses indicated a three-way interaction between stimulus type, stimulus manipulation, and group, F(1, 48) = 4.59, P < .05. There were also lower order effects for the two-way interaction between stimulus manipulation and group, F(1, 48) = 4.22, P < .05, and a main effect for stimulus type, F(1, 48) = 7.26, P < .05. There was also a nonsignificant trend for a main effect for group, F(1, 48) = 3.12, P < .09. All other effects were not significant (Fs ranging from 0.11 to 1.67, all Ps > .2). The three-way interaction is shown in Table 3 and Figure 1. The SCD group showed significantly lower scaled scores on the digit span-backward test, but did not differ from the comparison group on any of the other three span tests. The two-way interaction was due to a larger discrepancy between the SCD group relative to the comparison group in the backward conditions (M = 8.52 for SCD vs. 9.98 for controls) compared to the forward conditions (M = 9.30 for SCD vs. 9.56 for controls). The one-way effect for stimulus type was due to higher scaled scores for spatial locations (M = 9.82) than for digits (M = 8.86).

For the SOPT the analyses indicated main effects for stimulus set, F(2, 47) = 45.49, P < .01, and trial, F(1, 48) = 11.18, P < .01. All other effects were not significant (Fs ranging from 0.74 to 2.07, all Ps > .13). The main effect for stimulus set is shown in Table 3. Both groups showed similar increases in error rates as the number of stimuli in the set increased. The main effect for trial is shown in Figure 2. Both groups demonstrated relatively similar amounts of interference.
Table 3
Mean scores (standard deviation) for measures of short-term and working memory

<table>
<thead>
<tr>
<th>Variable</th>
<th>SCD group (n=25)</th>
<th>Comparison group (n=25)</th>
<th>t (48)</th>
</tr>
</thead>
<tbody>
<tr>
<td>WISC-PI digit span-forward s.s.</td>
<td>9.0 (3.2)</td>
<td>9.0 (2.2)</td>
<td>0.10</td>
</tr>
<tr>
<td>WISC-PI digit span-backward s.s.</td>
<td>7.4 (2.9)</td>
<td>10.0 (2.8)</td>
<td>3.19 *</td>
</tr>
<tr>
<td>WISC-PI spatial span-forward s.s.</td>
<td>9.6 (2.7)</td>
<td>10.1 (2.4)</td>
<td>0.61</td>
</tr>
<tr>
<td>WISC-PI spatial span-backward s.s.</td>
<td>10.0 (2.6)</td>
<td>9.6 (1.7)</td>
<td>0.58</td>
</tr>
<tr>
<td>SOPT total errors on all trials</td>
<td>5.8 (2.9)</td>
<td>5.5 (3.2)</td>
<td>0.32</td>
</tr>
<tr>
<td>6 Line drawings of objects</td>
<td>0.8 (0.8)</td>
<td>0.9 (1.0)</td>
<td>0.63</td>
</tr>
<tr>
<td>8 Colored squares</td>
<td>1.7 (1.2)</td>
<td>1.8 (1.6)</td>
<td>0.30</td>
</tr>
<tr>
<td>10 Nonrepresentational patterns</td>
<td>3.3 (1.8)</td>
<td>2.8 (1.5)</td>
<td>−1.46</td>
</tr>
</tbody>
</table>

Note. s.s. = age-adjusted scaled score.

* P < .05.

on the second trial as demonstrated by an increased number of errors. In particular it is worth noting that the trial by group interaction, which could have signified greater difficulties with the executive components of memory for the SCD group, did not suggest any meaningful difference between the groups, F(2, 47) = 1.56, n.s., partial eta squared = .062.

Post hoc analyses were conducted to explore the finding of lower digit span-backward scores in the SCD group. Digit span-backward scaled scores were examined for their relationship with parent education (r = .03, n.s.), parent income (r = −.02, n.s.), hematocrit (r = .15, n.s.), and each of the six subtests of cognitive abilities administered: Spatial Relations (r = .52, P < .01), Oral Vocabulary (r = .42, P < .05), Visual Closure (r = .08, n.s.), Incomplete Words (r = .50, P < .05), Category Fluency (r = .06, n.s.), and Visual Matching (r = .24, n.s.). Spearman rank order correlations were used for these tests of association. A regression analysis was then run.
Fig. 2. predicting digit span-backward-scaled scores from the three cognitive tests correlated with the digit span-backward test. The overall prediction of digit span-backward scores was significant, $F(3, 21) = 7.68$, $P < .01$, $R^2 = .523$. Both Spatial Relations, beta = .47, $t = 2.86$, $P < .01$, and Incomplete Words, beta = 1.09, $t = 3.04$, $P < .01$, showed unique relationships with digit span-backward scores.

A final post hoc procedure was conducted in which the individual data for the group with SCD was examined to determine which children had scored in the bottom half of the sample on the digit span-backward subtest. Among the twelve lowest scores on this subtest eight of the 12 cases were children that had reported a history of TIA ($n = 2$) or severe headache ($n = 6$). Seven of these eight cases had a history of normal neuroimaging exams within 2 years of study participation and the remaining child had not received a neuroimaging exam. Thus, a history of neurologic symptoms appeared to be related to lower scores, but there was no evidence to indicate silent cerebral infarcts were present in these cases.

3. Discussion

The present study evaluated short-term and working memory abilities in children with SCD that had no known history of cerebral vascular injury. The modality of the stimuli testing these abilities was varied to assess both auditory and visual modalities. A prior meta-analysis had suggested attention and executive skills may be particular areas of difficulty for children with this condition (Schatz et al., 2002). Therefore, we had hypothesized that the SCD group would show difficulties with the executive components of short-term and working memory. Contrary to our hypothesis children with SCD only showed decrements on the digit span-backward
condition. Children with SCD were able to perform similarly to a demographically matched group without SCD on forward span measures, spatial span reversed, and all conditions of the SOPT (which involved visual stimuli). Descriptive measures of cognitive ability also showed decrements in the SCD group for measures of crystallized ability, auditory processing, and processing speed within the Cattell–Horn Gf-Gc model.

One implication of these findings for current practice in neuropsychological assessment is the potential pitfalls inherent in combining the forward and backward conditions of span tests (Ramsay & Reynolds, 1995; Reynolds, 1997). Prior research with the SCD population has typically reported Wechsler digit span performance for the combined score and found mixed outcomes (Schatz et al., 2002). These mixed findings may due in part to combining scores from measures with somewhat different processing components. The findings also have implications for the role of executive functions in working memory. According to Baddeley and Logie (1999), the digit span-forward test primarily represents the capacity of the phonological loop in working memory whereas the backward condition begins to tap central executive functions related to manipulating information within temporary storage. In Baddeley’s model, the central executive is a separate component from either modality specific rehearsal system. The pattern of data in the present study do not fit Baddely’s model well, as the model would predict if central executive deficits are present this would impact digit span-backward, spatial span backward, and the SOPT. The present pattern of data, however, suggests modality specific difficulties within the central executive. Factor analytic research has also suggested the backward condition of digit span may load heavily on a visual-spatial memory factor in addition to sequential and auditory processing (Ramsay and Reynolds, 1995; Reynolds, 1997). This model from factor analytic work also does not provide a compelling explanation for the present set of findings.

Working memory models that incorporate modality specific rehearsal systems, however, could explain the present set of findings. Modality specific models of inhibitory control and other executive functions in working memory have been suggested by previous authors and are more consistent with the data in the present study (Cornoldi & Vecchi, 2000; Daneman & Tardif, 1987; Palladino, Mammarella, & Vecchi, 2003; Shah & Miyake, 1996). Functional neuroimaging data also support the existence of modality specific rehearsal systems; however, the extent to which these separate systems are involved in executive functions (rather than just rehearsal) is unclear at this time (Courtney, Ungerleider, Keil, & Haxby, 1996; McCarthy, Puce, Constable, & Krystal, 1996).

Follow-up analyses of the present data set suggested difficulties with digit span-backward were related to both auditory processing and fluid ability. Auditory processing deficits, however, appeared to be a more ubiquitous area of difficulty for children with SCD. Most prior studies of cognitive functioning in SCD have not included specific measure to assess auditory processing as a component ability. The one study that has identified this area of functioning relied on a teacher administered preschool screening test that has less well-established psychometric properties (Steen et al., 2002). Auditory–verbal measures, however, have frequently been found as areas of deficit including the Wechsler Scales vocabulary, arithmetic, digit span subtests (Schatz et al., 2002). The present data suggest a need to evaluate auditory processing as a distinct area of ability for children with SCD so as to help document both more basic and higher level processing difficulties. Additional studies of role of auditory processing in higher level cognitive abilities in SCD are needed as the current data is based on a relatively
small sample, which creates issues of generalizability of the findings, and the current study was not designed to assess different components of auditory processing which might provide greater insight into the causal factors involved (e.g., central vs. peripheral processing deficits).

Based on the data available it is difficult to know if the difficulties in auditory processing are due to central or peripheral nervous system problems. Sensorineural hearing loss (SNHL) can occur in SCD, although prevalence rates have varied from 3.5 to 41% across studies (Crawford, Gould, Beckford, Gibson, et al., 1991; Friedman, Luban, Herer, & Williams, 1980; Gould, Crawford, Smith, Beckford, Gibson, et al., 1991; MacDonald, Bauer, Cox, & McMahon, 1999). More recent U.S. studies, however, have suggested prevalence rates at the lower end of these estimates (MacDonald et al., 1999). Both lower level processing abilities such as acoustic reflex thresholds and higher level speech discrimination have been identified as possible areas of deficit (Sharp & Orlichik, 1978). There is no one mechanism consistently identified with temporal bone pathology, effects on the organ of Corti due to ischemia or thrombocytosis, and transient brain stem anoxia as examples of the range of SCD-related effects that may cause SNHL. It is of note, however, that most of the cases of SCD-related SNHL are not of the severity that would typically lead to corrective amplification (MacDonald et al., 1999). Furthermore, the impact of minor hearing deficits on higher level speech and language development appears to be relatively small (Roberts, Rosenfeld, and Zeisel, 2004).

In summary, the current study suggested modality specific difficulties with short-term memory span in children with SCD. Clinicians assessing this population should include measures of higher-level cognitive domains that rely specifically on either visual or auditory modalities to insure that modality specific issues can be identified in this population. Prior research on cognition in SCD has frequently relied almost exclusively on auditory–verbal measures to assess a wide range of functions, including memory and learning abilities. The test selection in these studies may have led to an over-generalization of the areas of deficit in SCD. The precise source of auditory–verbal deficits in SCD requires further evaluation.
References


