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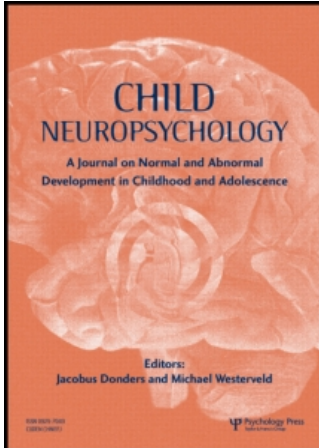
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EXECUTIVE FUNCTION IN CHARGE SYNDROME

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This study addressed the presence of executive dysfunction in children with CHARGE syndrome, a genetic disorder with multiple physical anomalies and severe challenging behaviors. Ninety-eight children were included in the study. More than half received clinically significant scores on the Behavior Rating Inventory of Executive Function (BRIEF; Gioia et al., 2000) scales of Shift, Monitor, and the Behavioral Regulation Index, with additional high scores on Inhibit and the Global Executive Composite. Associations were found with the age the child first walked, scores on the Autism Behavior Checklist (ABC; Krug et al., 1993), and being classified as deafblind. Difficulties with making transitions and flexible problem solving, monitoring their work and their effect on others, and acting on impulse, may be related to the behavioral difficulties exhibited by children with CHARGE. Interventions targeting improved self-regulation may help to manage this challenging behavior.

CHARGE syndrome was first identified by Hall in 1979, based on the association of choanal atresia and multiple anomalies, i.e., coloboma, heart defects, growth delays, genital anomalies, and ear malformations. Individuals with CHARGE are frequently deafblind and have cranial nerve abnormalities that affect vestibular functioning, swallowing, sense of smell, facial palsy, and sensorineural hearing loss. The acronym CHARGE was introduced in 1981 by Pagon and her colleagues (Pagon, Graham, Zonana, & Yong, 1981). Although originally described as an association, it became increasingly clear to those geneticists who studied CHARGE that distinctive features exist that qualify clinically as a syndrome (Davenport, Hefner, & Mitchell, 1986, Lubinsky, 1994, Blake et al., 1998, Graham, 2001). Recently the gene *CHD7* was implicated in 10 of 17 individuals with CHARGE (Vissers et al., 2004).

Behavioral difficulties in CHARGE syndrome have long been of concern to parents. Hartshorne and Cypher (2004) created a list of behaviors associated with disorders that have been reported in children with CHARGE (autism, attention deficit disorder, obsessive-compulsive disorder, tic disorder, and deafblindness). Three behaviors emerged as the most typical in children with CHARGE: “extreme preference for certain toys, people, food, etc.,” “restricted range of interests and/or pre-occupation with one narrow interest,” and “significant difficulty in ability to make same age friendships.”

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Several studies have addressed the autistic-like behavior in CHARGE (Fernell, Olsson, Karlgren-Leitner, Norlin, Hagberg, & Gillberg, 1999; Hartshorne, Grialou, & Parker, 2005; Bernstein & Denno, 2005; Smith, Nichols, Issekutz, & Blake, 2005). Hartshorne, Grialou et al. found that 27% of their sample of 160 persons with CHARGE could be classified as autistic based on Autism Behavior Checklist scores (ABC; Krug, Arick, & Almond, 1993), although total scores were significantly lower than the autism norms and significantly higher than the deafblind norms on the ABC. Participants with CHARGE scored considerably lower than both the autism and deafblind groups on one subtest entitled *Relating to Social Stimuli*. This subtest includes items such as avoiding eye contact, avoiding touch, and looking through others, suggesting that this is an area of strength.

Age of walking has been found in several studies to be related to behavior (Hartshorne & Cypher, 2004; Hartshorne, Grialou et al., 2005; Salem-Hartshorne & Jacob, 2004, 2005). Walking is typically delayed in CHARGE because of a combination of absent or malformed semicircular canals affecting the vestibular, visual, and motor functioning of these individuals (Williams & Hartshorne, 2005). The source for the relationship between age of walking and behavior is not known, but it provides a potentially useful marker for behavioral outcomes.

Nicholas (2005) speculated that executive dysfunction may contribute to the behavioral difficulties in CHARGE. He presented the case study of a 12-year-old girl with CHARGE who showed considerable executive dysfunction as measured by the Behavior Rating Inventory of Executive Function (BRIEF; Gioia, Isquith, Guy, & Kenworthy, 2000).

Executive functions are the abilities needed to monitor, to control and to regulate thought and action (Carlson, Davis, & Leach, 2005). They include processes that serve to coordinate the execution of cognitive abilities such as attention and memory, and that play a role in typical and atypical social and cognitive development (Carlson et al.). Executive functions include strategic planning, decision making, judgment, self-perception, and self-monitoring that direct and regulate behavior (Nicholas, 2005). Many behavioral problems arise from impaired executive functions such as poor impulse and inhibitory control (Barkley, 1997), impaired judgment and problem solving (Zelazo, Carter, Reznick, & Frye, 1997), difficulties in making mental or behavioral shifts (Denckla, 1996), inability to apprehend errors in performance, reduced self-regulation, or organized strategic behaviors (Graham & Harris, 1996), low mood and motivation, poor communication skills, low self-esteem, and social isolation (Anderson, Anderson, Northam, Jacobs, & Mikiewicz, 2002). Disturbances of executive functioning are most likely to be evident when the person is required to cope with novel and/or complex tasks and situations (Shallice & Burgess, 1998; Godefroy & Rousseaux, 1997).

Shallice and colleagues have developed a model of executive function disorders based on damage to a higher level supervisory system (Norman & Shallice, 1986; Shallice, 1982), which most likely contains a number of separable subsystems (Shallice & Burgess, 1998). Impairment of the supervisory system results in over reliance on routine control through a contention scheduling system, leading to higher rates of over learned responses to environmental contingencies. From this model, we would predict that children with executive dysfunction might 1) engage in more routinized behaviors and 2) express less flexibility in their behaviors, both of which seem consistent with the behavioral profile of children with CHARGE (Hartshorne, Hefner, & Davenport, 2005).

Executive functions are presumed to be a function of the prefrontal cortex (Denckla, 1996) as well as a distributed network of reciprocal connections in other areas of the brain such as the basal ganglia, the limbic system, the thalamus, and the posterior cortex

(Pennington, Bennetto, McAleer, & Roberts, 1996). Of interest is whether executive functions comprise multiple subdomains so that distinct profiles of executive dysfunction may help to differentiate different developmental disorders. Gioia, Isquith, Kenworthy, and Barton (2002) used the Behavior Rating Inventory of Executive Function (BRIEF) to examine the profiles of groups of children with reading disorders, attention deficit/hyperactivity disorder, autism spectrum disorder, and traumatic brain injury and found some distinct differences in the patterns of their executive dysfunctions.

The behavioral characteristics seen in CHARGE syndrome can have many causes including sensory impairments, a history of medical illness and intervention, and communication difficulties (Hartshorne, Hefner et al., 2005), but central nervous system disturbances may be a contributory factor. This is especially true when neurological and other disturbances coexist, as is frequently the case in persons with CHARGE syndrome. Variability in cognitive functioning is often related to specific brain dysmorphologies. Central nervous system anomalies in CHARGE syndrome were reported by Lin, Siebert, and Graham (1990), including holoprosencephaly with and without arhinencephaly. Raqbi et al. (2003) found central imaging to be normal for only 4 of 21 cases, with asymmetrical ventriculomegaly, nonspecific cortico-subcortical atrophy, agenesis of the corpus callosum, brainstem hypoplasia, and anomalies of the cerebellum among the findings.

Measurement of executive functions has been complicated by the general lack of an operational definition or universally accepted set of executive function tasks (Hughes & Graham, 2002). In addition, some standardized neuropsychological measures may not reflect daily life experiences (Anderson et al. 2002). The child's everyday environments at home and school can serve as important environments for observing the essence of executive functions (Gioia et al., 2000), and recently developed rating scales, such as the BRIEF, have tried to capture these.

The goal of this study was to investigate the executive functions in persons with CHARGE syndrome, and how these may be related to their challenging behavior. This is the first examination of executive function in CHARGE syndrome.

METHODS

Participants

Materials for this and a related study were mailed to parents of 159 children with CHARGE syndrome who had previously participated in a study of autistic-like behavior in CHARGE (Hartshorne, Grialou et al., 2005). All of the children were estimated to be five years of age or older at the time of the present study. The participants were obtained through the CHARGE Syndrome Foundation. Materials were returned by 124 parents for a 78% response rate. However, 7 were not yet 5 years old and an additional 19 were older than 18. These were eliminated from the analyses for this study, leaving a sample size of 98.

Materials

CHARGE history questionnaire. This 13-item form (based upon one developed by Salem-Hartshorne and Jacob, 2004) gathered information on basic demographics, the child's CHARGE features, communication skills, age of crawling and walking, information on vision and hearing, other diagnoses, and medication.

Behavior Rating Inventory of Executive Function (BRIEF) – parent version. The BRIEF (Gioia et al., 2000) consists of two rating forms – a parent questionnaire and a teacher questionnaire – designed to assess executive functioning in home and school environments in children ages 5 to 18. Each BRIEF questionnaire contains 86 items within eight theoretically and empirically derived, nonoverlapping clinical scales that measure different aspects of executive functioning (Inhibit, Shift, Emotional Control, Initiate, Working Memory, Plan/Organize, Organization of Materials, and Monitor) and two validity scales (Inconsistency and Negativity). The clinical scales form two broader indexes, Behavioral Regulation and Metacognition, and an overall score, the Global Executive Composite. Norms are based on a normal population, although limited geographically. Reliability and validity indicators are acceptable (Baron, 2000). Initial factor analytic studies and structural equation modeling provided support for the two-factor model of executive functioning as encompassed by the two indexes, although recent confirmatory factory analysis suggests a three-factor solution (Gioia, Isquith, Retzlaff, & Espy, 2002).

To date, clinical populations that have been studied using the BRIEF include children with autism spectrum disorders (Gilotty, Kenworthy, Sirian, Black, & Wagner, 2002), ADHD (Kenealy, 2002; Blake-Greenberg, 2003; Palencia, 2003; Pratt, 2000), traumatic brain injury (Mangeot, Armstrong, Colvin, Yeates, & Taylor, 2002; Vriezen & Pigott, 2002; Gioia & Isquith, 2004), Tourette's disorder (with and without ADHD) (Mahone, Cirino, Cutting, Cerrone, Hagelthorn, Hiermenz et al., 2002), 22q11 Deletion Syndrome (Kiley-Brabeck, 2004), spina bifida and hydrocephalus (Burmeister, Hannay, Copeland, Fletcher, Boudousquie, & Dennis, 2005), and children with documented brain disease (Anderson et al., 2002). While Nicholas (2005) presented a case study of one child with CHARGE using the BRIEF, this is the first large scale study of CHARGE using this instrument.

Autism Behavior Checklist (ABC). The ABC is a component of the Autism Screening Instrument for Educational Planning, Second Edition (Krug et al., 1993). The Checklist (ABC) consists of a list of 57 behaviors that are checked if present. These are then coded onto five scales, Sensory, Relating, Body and Object Use, Language, and Social and Self-Help, as well as a total score. Total scores of 68 and above are considered indicative of autism. Norms are also provided for normal, deafblind, severely mentally retarded, and severely emotionally disturbed. Psychometric studies support the use of the ABC as a screening instrument (Eaves, Campbell, & Chambers, 2000). The participants' scores on the ABC were collected for a previous study, approximately one year prior to the present study.

RESULTS

Participants

Of the 98 returned, 92.9% were completed by the mother, 59.2% of the children were male, and ages ranged from 5 to 18, with a median of 10 and a mean of 10.63, $SD = 3.73$. The percentage of children with various CHARGE characteristics is shown in Table 1.

Executive Function

Standard scores on the BRIEF are *T*-scores (mean of 50 and standard deviation of 10), with a score 65 or higher considered potentially clinically significant. Three protocols

Table 1 Medical Problems in the Sample.

Condition	Percent (<i>N</i> = 98)
Delayed Motor Milestones	98
Vestibular Problems	88
Coloboma	85
Sensorineural Hearing Loss	83
Characteristic CHARGE ear	82
Growth Deficiency	82
Frequent Middle Ear Infections	80
Heart Defect	77
Swallowing Problems	74
Choanal Atresia or Stenosis	61
Genital Hypoplasia	55
Facial Palsy	46
Anosmia	38
Spine Anomalies	37
Renal Problems	36
Cleft Lip or Palate	28
Tracheoesophageal Fistula	24
Hand Anomalies	19
Abdominal Defects	15

were invalid because of the omission of more than 14 items. A number of the clinical scales could not be scored due to the omission of more than two items. The number of scales that could be scored ranged from 80 (Plan/Organize) to 94 (Emotional Control). Usable index scores included 87 (Behavioral Regulation), 80 (Metacognition), and 79 (Global Executive Composite). As shown in Table 2, the mean score on the Shift scale reached clinical significance for this sample. Mean scores for the Inhibit and Monitor scales were over 64. To examine the extent of the difference found for children with CHARGE, one-sample *t*-tests were calculated against the standardized mean of 50. All of the scales of the BRIEF were significantly different except for Organization of Materials, even with a step-up Bonferroni correction (Hochberg, 1988). Effect sizes (Cohen's *d*) were large for all scales except plan/organize that was medium, and organization of materials, which while not significant, did obtain a small effect size of .20. Table 2 also shows

Table 2 BRIEF Mean Standardized T-Scores, One-Sample *t*-Tests, and Percent Clinical.

Scale	Mean (<i>SD</i>)	<i>N</i>	<i>t</i> -test	<i>d</i>	%>64
Inhibit	64.02 (14.66)	88	8.97***	.96	46.6
Shift	65.42 (14.00)	93	10.62***	1.10	57
Emotional Control	58.44 (13.06)	94	6.26***	.65	31.9
Initiate	60.93 (12.64)	87	8.07***	.86	40.2
Working Memory	62.67 (12.90)	90	9.31***	.98	12.3
Plan/Organize	60.66 (13.89)	80	6.87***	.77	41.2
Organization of Materials	52.1 (10.54)	90	1.89	.20	12.3
Monitor	64.44 (13.40)	91	10.28***	1.07	54.8
Behavioral Regulation Index	63.79 (13.58)	87	9.47***	1.01	50.6
Metacognition Index	61.64 (13.00)	80	8.00***	.90	45
Global Executive Composite	63.00 (12.91)	79	8.94***	1.01	49.4

****p* < .001.

the percentage of those children who achieved a score 65 or higher on each scale. More than half received significant scores on Shift, Monitor, and the Behavioral Regulation Index (BRI). Of course it is also important to note that for most scales, a majority of children with CHARGE did not receive clinically significant scores.

Age of Walking

In this study the average age of walking was 3.08 ($SD = 1.74$) and the average age of crawling was 1.64 ($SD = 0.94$). Two children, ages five and six, were not yet walking, but they were both crawling. Parents were also asked to describe their child's walking ability, and this was converted to a three-point scale. Correlations between age of walking and crawling and walking ability with the three indexes on the BRIEF (Behavioral Regulation Index, Metacognition Index, Global Executive Composite) are shown in Table 3. Age of walking was significantly associated with all three indexes, indicating that as age of walking increases, scores on the BRIEF indexes are more clinically significant.

Deafblindness

Hartshorne and Cypher (2004) found that those persons with CHARGE who were deafblind had more behavior problems. In the present study, 55 of the 98 persons with CHARGE were identified by their parents as deafblind. The relationship between being deafblind and scores on the BRIEF was investigated by independent t -tests. Results are shown in Table 4. Shift, Emotional Control, and the Behavior Regulation Index (BRI) were all significantly higher for those who were identified as deafblind. However, when the step-up Bonferroni procedure was applied (Hochberg, 1988), only the Behavior Regulation Index continued to be significant. The effect size for the BRI was medium (Cohen's $d = .50$).

Autistic-like Behavior

Scores on the BRIEF were correlated with total score on the ABC. There is clearly a strong association (Table 5). With the exception of Organization of Materials, all correlations were significant at the .01 level.

Logistic Regression

In order to learn more about those participants who received clinically significant scores on the BRIEF, logistic regression was used to explore variables that might predict clinical scores. Total score on the ABC, age of walking, deafblindness, age, and gender

Table 3 Correlations Between Walking and BRIEF.

	BRI	MI	GEC
Walking Ability	.11	.12	.13
Age Walked	.23*	.29*	.27*
Age Crawled	.07	.14	.10

* $p < .05$.

Table 4 Performance on the BRIEF for Those Identified as Deafblind) versus not Deafblind.

Scale	Deafblind		Not Deafblind		<i>t</i> -test (df)
	<i>N</i>	Mean (<i>SD</i>)	<i>N</i>	Mean (<i>SD</i>)	
Inhibit	51	65.69 (14.86)	37	61.73 (14.27)	1.25 (86)
Shift	54	68.06 (13.02)	39	61.77 (14.65)	2.18 (91)*
Emotional Control	54	60.96 (13.50)	40	55.03 (11.75)	2.23 (92)*
Initiate	50	61.32 (11.67)	37	60.41 (13.99)	.33 (85)
Working Memory	51	62.71 (12.06)	39	62.62 (14.09)	.03 (88)
Plan/Organize	46	60.00 (13.63)	34	61.56 (14.39)	-.49 (78)
Organ. of Materials	51	51.69 (9.89)	39	52.64 (11.44)	-.42 (88)
Monitor	52	64.94 (12.95)	39	63.77 (14.12)	.41 (89)
Beh. Regulation Ind.	50	66.62 (13.21)	37	59.97 (13.30)	2.31 (85)*
Metacognition Index	46	61.61 (11.79)	34	61.68 (14.67)	-.02 (78)
Global Exe. Comp.	45	64.11 (11.92)	34	61.50 (14.17)	.89 (77)

p* < .05.Table 5** Correlations of BRIEF Scales with Autism Behavior Checklist.

Behavioral Regulation Index	.49**
Metacognition Index	.37**
Global Executive Composite	.45**
Inhibit	.44**
Shift	.37**
Emotional Control	.42**
Initiate	.36**
Working Memory	.45**
Planning	.29**
Organization of Materials	.11
Monitor	.35**

***p* < .01.

were entered stepwise into the logistic equation for Global Executive Composite (GEC) scores on the BRIEF. Only ABC scores met the criteria for entry and provided a significant model chi square (1, *N* = 77) = 9.34, *p* = .002. A Nagelkerke R Square was .15, indicating that only about 15% of the variance is accounted for. Ability to correctly predict membership (clinical vs. nonclinical) improved from 50.6% to 66.2%. Thus, knowing that a child had a high score on the ABC would be a predictor of a clinically significant score on the BRIEF, but not a strong predictor. The other variables did not help to discriminate clinical from nonclinical participants.

DISCUSSION

This study supports the presence of considerable executive dysfunction in children with CHARGE syndrome as measured by the BRIEF. Problems with being able to flexibly respond to situational demands (Shift), to track one's own behavior in relation to task attainment (Monitor), and to control impulses and to terminate behaviors as required (Inhibit) are evident from the results reported. Deficiencies in these particular executive

functions seem consistent with some of the challenging behavior exhibited by many children with CHARGE.

The Shift scale measures how flexibly a person can move from one activity or focus to another. Problems with shifting may be seen in perseverative behavior such as asking the same question over and over. Bernstein and Denno (2005) looked at 29 children with CHARGE and found that repetitive behaviors in that group significantly interfered with daily routine. Furthermore, redirection commonly elicited aggression, and traditional behavioral techniques were not beneficial.

The Monitor scale measures the child's ability to review their own behavior in terms of task accomplishment as well as the effect their behavior has on others. Although Hartshorne, Grialou et al., (2005) found that children with CHARGE had a relative strength on the social engagement (Relating) subtest of the Autism Behavior Checklist compared to the norms for children who were autistic or deafblind, despite this interest they tend to have limited social success. Hartshorne and Cypher (2004) found that difficulty in establishing same-age friendships was one of the top three behavioral concerns. Souriau et al. (2005) found difficulty in waiting one's turn and in following social rules to be problems in children with CHARGE. These findings are consistent with someone who tries to engage socially but is unsuccessful because of a problem with monitoring how they are impacting the relationship. Sensory impairments are likely to be a major contributor to this difficulty. However, parents report that even those children with good communication skills, and whose vision and hearing are not affected or are only mildly affected, nevertheless have problems with social relationships, supporting the possible role of executive dysfunction.

Because ADHD is a diagnosis often given to children with CHARGE (Hartshorne and Cypher, 2004), the finding of difficulty on the Inhibit scale was expected. This scale measures problems with impulse control. Souriau et al. (2005) found that 54% of 71 children seemed permanently on the move. Hartshorne and Cypher (2004) found at least a third of the 100 children in their sample had difficulty with 9 of 10 behaviors commonly associated with ADHD. The most common of these was frequently interrupting others and fidgeting with objects.

Among the executive functions measured by the BRIEF, Organization of Materials was a relative strength for this sample of children with CHARGE. This scale measures the ability to manage and to organize one's space, for example one's desk or closet. Souriau et al. (2005) found that more than half of their participants (55%) liked things to be in the same place. Of course orderliness is a benefit to individuals with sensory impairments.

Based on previous research on executive dysfunction in children with autism (e.g., Gilotty, Kenworthy, Sirian, Black, & Wagner, 2002), the finding of an association between scores on the BRIEF and the ABC was expected. Only Organization of Materials was not significantly correlated with total scores on the ABC. In addition, scores on the ABC helped to differentiate children with clinically significant scores on the General Executive Composite from those who did not. While there may be overlap in the content of these two measures, the correlation coefficients were moderate suggesting that they were measuring somewhat distinct constructs, and that those children with CHARGE who have more executive dysfunction also tend to be those who have more autistic-like behaviors.

Because age of walking has been found in previous research to be correlated with ABC scores (Hartshorne, Grialou et al., 2005) as well as adaptive behavior scores (Salem-Hartshorne & Jacob, 2004, 2005), it is not surprising that they were also correlated with all

three indexes on the BRIEF. Salem-Hartshorne and Jacob (2005) found support for a mediational model where vision severity and medical involvement affect age at walking, which in turn affects adaptive behavior. Medical involvement was measured by summing the various CHARGE features and so is not a pure measure of the severity of CHARGE. Severity is likely influenced by neurological factors.

Hartshorne, Grialou et al. (2005) found average total scores on the ABC to be significantly higher than the reported norms for children who are deafblind. It is evident from the present study that being deafblind as a part of CHARGE is generally related to higher scores on the BRIEF. On the Behavioral Regulation Index (BRI), which is a global measure of the child's ability to self-regulate behavior, those who were deafblind had statistically higher scores with a moderate effect size. Many children with CHARGE have cranial nerve dysfunction including optic nerve coloboma and sensorineural hearing loss that suggests the possibility of neurological problems that could in turn be associated with executive dysfunction. The use of the BRIEF with other populations with deafblindness would be useful.

Limitations and Future Directions

This study was based on parent-report that has been typical of much of the research on behavioral issues in CHARGE. This has been necessitated by the rarity of the condition and the lack of large clusters of persons with the syndrome. An exception was the work by Bernstein and Denno (2005) at Perkins School for the Blind where there is a concentration of children with CHARGE. The report of autism in CHARGE by Fernell et al. (1999) was based on three case histories. More research based on direct observation or teacher/professional report would be useful.

CHARGE has a highly variable phenotype. Due to the multisensory impairments it is very difficult to measure cognitive ability in many individuals with CHARGE. Since some young adults with CHARGE are able to attend college, it is clear that cognitive impairment is not an inevitable component of the syndrome. Salem-Hartshorne and Jacob (2004, 2005) investigated adaptive behavior skills in children with CHARGE and found more than half to have scores in the normal range. However, multisensory impairments and communication difficulties, along with some cognitive impairment, made it difficult for parents to answer all of the questions on the BRIEF. For example, the item "Has good ideas but cannot get them on paper" is difficult if the child does not write or perhaps even communicate well. Future studies might consider using other measures of executive function that better tap the abilities of this group.

While descriptions of executive function profiles can be useful (Gioia, Isquith, Kenworthy et al., 2002), there is a need to move beyond the level of description and address behavioral symptoms at the deeper, causal level. This will require more understanding of the neurological impairments that are frequent in CHARGE, as well as more research on the neurology of executive functions (Anderson et al., 2002). While the profile provided by the BRIEF is a good beginning, it will be helpful to first identify situations during which children with CHARGE exhibit problems with inhibition, shifting, and monitoring and, second, to identify when these problems increase their reliance on routine control (Norman & Shallice, 1986; Shallice, 1982). Such work could begin to generate hypotheses for improving the executive functioning of individuals with CHARGE.

An improvement in executive functioning by the facilitation of the supervisory-executive control process is based on a sound framework of neuropsychological function and intervention (Cicerone, 2002; Eslinger, 2002). Such intervention is directed toward

the internalization of self-regulatory processes, and therefore is by nature accessible in a variety of situations. Two types of changes in specific behaviors seen in CHARGE syndrome might be achieved through this type of intervention: first, changes in the enhanced performance on tasks requiring problem solving and error correction; and second, changes in the improved ability to inhibit the release of inappropriate responses. Research on specific interventions for the behavioral difficulties of children with CHARGE that take into account the executive dysfunction is needed.

It should be noted that, as is typical of samples of individuals with CHARGE due to their considerable variability, the standard deviations on all of the scales were fairly large. Thus a diagnosis of CHARGE should not be presumed to indicate that the child will measure as having executive dysfunction.

CHARGE syndrome is a complex disorder affecting multiple organs and processes, and individuals with CHARGE frequently engage in challenging behaviors. The finding that children with CHARGE were assessed as having considerable executive dysfunction as measured by the BRIEF is consistent with the theory that executive dysfunction is linked to the observed challenging behavior. Utilizing the profile created by the BRIEF will be useful in better understanding sources of the behavior difficulties, and ultimately the development of specific interventions.

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