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Factors Contributing to Executive Functioning in Children with Neurofibromatosis Type 1

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FACTORS CONTRIBUTING TO EXECUTIVE FUNCTIONING IN CHILDREN WITH NEUROFIBROMATOSIS TYPE 1

by

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ABSTRACT

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Despite variability in the neurofibromatosis type 1 (NF1) cognitive phenotype, attention and executive functioning (EF) difficulties are often described, and high rates of attention-deficit/hyperactivity disorder (ADHD) have long been associated with NF1. Despite the known clinical relation between NF1 and ADHD, there is a paucity of research exploring potential factors that contribute to ADHD vulnerabilities in children and adolescents with NF1. Furthermore, recent research suggests that impairment in EF, a construct highly associated with ADHD, occurs in children with NF1 independent of ADHD diagnosis suggesting that the presence of EF impairment in children with NF1 may not be uniquely associated with ADHD. Given the complexity of EF and the relative lack of literature about factors that might contribute to EF performance in children with NF1, further research is warranted. The current study aims to characterize EF in children with NF1, compare EF from performance-based and functional measures, and explore potential neuropsychological, sociodemographic, and psychosocial factors that contribute to EF in children with NF1. Overall, results confirmed that children with NF1 demonstrate difficulty on performance and functional EF measures, and difficulties were more evident based on functional parent report of behavior. Over one-third of children with NF1 met diagnostic criteria for ADHD; however, children with NF1 as a group demonstrated similar EF

profiles on performance measures, independent of ADHD diagnosis. On functional parent reported measures of EF, children with NF1 and ADHD demonstrated significantly higher levels of executive dysfunction compared to children with NF1 without ADHD. Relations between performance-based working memory and general cognitive functioning were found for children with NF1, as a group. Parent report of internalizing problems were related to parent report of functional emotional control, shifting/cognitive flexibility, and overall behavioral regulation. As hypothesized, parent reported sleep difficulties were related to functional EF. In addition, slower reaction times on a working memory task were related to parent report of snoring, and parent report of restlessness during sleep was related to functional EF. Group differences between children with NF1 who met cut-off criteria for a sleep-related breathing disorder and those that did not were apparent when examining parent report of functional inhibition, working memory, and self-monitoring difficulties. Overall, results highlight the utility of a multi-method assessment of EF and provide evidence for contributing factors of overall cognition, attention, internalizing problems, and sleep on various aspects of EF in children and adolescents with NF1.

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Factors Contributing to Executive Functioning in Children with Neurofibromatosis type 1

Neurofibromatosis type 1 (NF1) is a common, highly variable autosomal dominant neurodevelopmental disorder that presents in approximately 1 in 3,500 individuals. Over half of children with NF1 manifest specific cognitive impairments, including language problems, learning disabilities, visual-motor impairment, and visuospatial deficits (Hyman et al., 2005). In addition, nearly half of children with NF1 also display attention and executive functioning (EF) impairments, and high rates of attention-deficit/hyperactivity disorder (ADHD) are associated with NF1 (Templer et al., 2013; Koth et al., 2000). Recent research suggests EF impairment occurs in children with NF1 independent of ADHD diagnosis (Roy et al., 2014). As such, the presence of EF impairments in children with NF1 may not be uniquely associated with ADHD. Despite evidence that EF impairments are prevalent in children with NF1, previous studies have rarely utilized functional measures in conjunction with more common performance-based EF measures. Furthermore, there is a lack of literature examining potential contributions to EF performance in children with NF1. Examination of neuropsychological, sociodemographic, and psychosocial factors known to contribute to EF is warranted. The aims of the current study are threefold. First, this study aims to characterize EF performance in our sample of children with NF1. Second, this study aims to compare and contrast EF from performance-based and functional measures; and third, this study aims to examine potential contributing variables to EF performance in children with NF1.

In this introduction, the current literature on EF deficits and ADHD symptomatology often reported in children with NF1 will be examined. First, I will provide general background information about NF1, in which I will briefly describe medical features and the common

cognitive and behavioral characteristics of children with NF1. Second, I will review research examining EF in children with NF1 and discuss factors that may contribute to EF impairments in children with NF1. Third, I will provide general background information about EF, including conceptualization, development of specific EF processes, and common assessment tools used to examine EF. Lastly, I will provide a summary and rationale for the current study.

Genetics and Medical Presentation of NF1

Neurofibromatosis type 1 (NF1) is one of the most common autosomal dominant neurodevelopmental disorders, with an estimated incidence of 1 in 3,500 individuals. NF1 is caused by a mutation in a gene located on chromosome 17q11.2, which codes for neurofibromin. Neurofibromin regulates activity of ras, a protein that promotes cell division. Due to the mutation present in NF1, the neurofibromin protein is unable to bind to ras or regulate its activity, thus causing the ras protein to be more active. This over activity in ras protein leads to development of tumors over time. Therefore, NF1 is progressive and complications relating to central nervous system dysfunction worsen over time (Friedman, 1999; North, 1998).

Despite variability in the NF1 presentation, the most common physical manifestations of NF1 include café-au-lait skin patches, cuteaneous neurofibromas, axillary freckling, and Lisch nodules (North, 1998). Café-au-lait skin patches are present in more than 95% of individuals with NF1 and can be the earliest sign of NF1, often presenting before 2 years of age. NF1 is considered a multisystem disorder with many potential medical complications, including tumors, malformations, neuropathy, neurovascular disease, and epilepsy (Friedman, 1999). While medical abnormalities are indeed problematic for many children with NF1, the most common complaints from parents of children with NF1 are not medical in nature, but rather are related to neuropsychological, behavioral, and emotional functioning.

NF1 Neuropsychological Phenotype

General Phenotype. It has been well documented that over half of children with NF1 manifest specific cognitive impairments, including language problems, math and reading disabilities, attention and executive function (EF) deficits, and visuospatial deficits (Hyman et al., 2005; Mautner et al., 2002; Hyman et al., 2006). Generally, there is evidence for a slight downward shift of the normal distribution with regards to mean IQ in individuals with NF1 relative to general populations, with their performance typically falling in the low average to average range of functioning relative to the general population and compared to sibling contrast groups (Cutting et al., 2000; Ferner et al., 1996; Hyman et al., 2005; Sangster et al., 2011). The presence of intellectual disability remains relatively rare in the NF1 population; however, reported rates (4-8%) are higher than that seen in the general population (1-2%) (North et al., 1997; Maulik et al., 2011).

NF1 research over the past several years has focused largely on the presence of attention and learning deficits in NF1. It has been well established that children with NF1 are at a higher risk for learning and attention difficulties compared to unaffected siblings (Vogel et al., 2017; Ferner et al, 1996; Cutting et al, 2000; Mautner et al, 2002; Hyman et al., 2005). Difficulties with attention affect approximately 30-50% of children with NF1, making attention difficulties a seemingly characteristic feature of the overall cognitive profile (Hofman et al., 1994; Hyman et al., 2005; Koth et al., 2000). Nearly half of children with NF1 meet criteria for ADHD (Hyman et al., 2005; Mautner et al., 2002), which is significantly higher than the 4-7% diagnosed with ADHD in the general population (Thomas et al., 2015). Attention abilities have been systematically examined and have been discussed in the context of delineating the common behavioral features of NF1 and to partially account for the high rates of learning deficits (Hyman

et al., 2006; Potvin et al., 2015). Deficits related to inattention and vigilance with sustained attention have been reported to be more prevalent in children with NF1, as opposed to hyperactive and impulsive symptoms (Hyman et al., 2005; Templer et al., 2013). Hyman and colleagues (2005) found deficits in sustained attention in children with NF1, even when controlling for intellectual functioning. Furthermore, the ADHD prevalence ratio of males to females was observed to be equal in the NF1 population, whereas the prevalence ratio of males to females for ADHD in the general population is 2:1-3:1 (Pastor et al., 2015). It has been suggested that ADHD represents a prevalent neuropsychiatric phenotype of NF1 due to the pervasive nature of inattention in individuals with NF1 (Huijbregts, 2012); however, others have suggested the possibility of an ADHD comorbidity in NF1 (Lidzba et al., 2012).

Executive Functioning in NF1. Many children with ADHD display EF deficits. As such, it is not unexpected that early descriptions of the NF1 cognitive phenotype literature included anecdotal observations of EF deficits through descriptions of poor performance on visual-perceptual tasks which were partially explained by impulsivity (Eliason, 1986). In one of the first studies to examine EF performance in children and adults with NF1, Ferner and colleagues (1996) compared the performance of individuals with and without NF1 on performance-based measures, revealing that individuals with NF1 demonstrated more difficulty inhibiting responses on automated performance tests compared to unaffected individuals. Deficits in working memory and flexible set-shifting were also reported in the NF1 group, and according to Ferner (1996) and colleagues, performance by children with NF1 resembled the performance of individuals with frontal lobe disorders.

Since these first descriptions of EF impairments in NF1, evidence from performance-based and functional behavior measures has emerged suggesting that EF, like attention,

represents a core deficit in NF1 (see Table 1 for review of studies). Inhibition is the most frequently reported EF impairment in children with NF1 (Ferner et al., 1996; Rowbotham et al., 2009; Gilboa et al., 2011; Isenberg et al., 2013; Pride et al., 2017; Casnar & Klein-Tasman, 2016; Mazzocco et al., 1995; Payne et al., 2011; Mautner et al., 2002; Plasschaert et al., 2015). Working memory deficits are also frequently reported in children with NF1 (Casnar & Klein-Tasman, 2016; Champion et al., 2014; Ferner et al., 1996; Gilboa et al., 2014; Huijbregts et al., 2010; Payne et al., 2011; Plasschaert et al., 2015; Rowbotham et al., 2009; Sangster et al., 2011; Ulrich et al., 2010). Cognitive flexibility and shifting deficits have also been observed in children with NF1 (Casnar & Klein-Tasman, 2016; Descheemaeker et al., 2005; Hofman et al., 1994; Lion-Francois et al., 2017; Payne et al., 2011; Plasschaert et al., 2015; Pride et al., 2010; Rowbotham et al., 2009; Roy et al., 2014). In addition, planning and organization deficits have been reported in children with NF1 (Galasso et al., 2014; Gilboa et al., 2014; Hofman et al., 1994; Hyman et al., 2005; Payne et al., 2011; Plasschaert et al., 2015; Pride et al., 2010; Roy et al., 2010). Plasschaert and colleagues (2016) conducted a multi-method study which included an extended battery of tests to study inhibition, cognitive flexibility, working memory, and planning in children ages 8 -18 years with NF1. Compared to an unaffected sibling group, children with NF1 exhibited deficits on all EF domains, even after including IQ as a covariate, suggesting problems are not merely due to lower level of cognitive functioning. In addition, all functional behavior ratings of EF were significantly elevated, reflecting more EF difficulties, in comparison to the contrast group. In the first study to investigate the functional correlates of response inhibition in children with NF1, Pride and colleagues (2017) reported that children with NF1 had significantly less activation than age-matched controls in the pre-supplementary motor area, inferior frontal gyrus, inferior occipital gyrus and the fusiform gyrus/posterior cerebellum, a

network previously established as crucial to the Go-No-Go processing. It was also noted that this abnormality was associated with faster reaction times, a reflection of impulsivity, and deficits in sustained attention.

It has also been demonstrated that EF deficits exist in children with NF1 with and without ADHD (Heimgärtner et al., 2019; Huijbregts et al., 2012; Lion-Francois et al., 2017; Payne et al., 2012), suggesting that ADHD alone cannot account for the EF impairments observed. Hyman and colleagues (2005) assessed the planning and abilities of 81 children and adolescents with NF1 and compared performance to 49 unaffected siblings using performance-based measures. Children with NF1 performed significantly lower on all measures of planning, and children with comorbid ADHD generally did not perform worse than children with NF1 without ADHD. However, when controlling for IQ, the differences in performance between children with NF1 and unaffected siblings were no longer significant. These results have been confirmed by other researchers, such as Roy and colleagues (2010), who reported that children with and without comorbid ADHD exhibit planning deficits above and beyond the role of cognitive functioning. Galasso and colleagues (2014) examined performance of 18 children with NF1, 18 children with ADHD, and 18 typically developing children on functional reports of inattention and Tower of London (Krikorian et al., 1994) performance. Significantly elevated inattention scores were evident on the parent-reported functional measure for the NF1 and ADHD groups. Compared to typically developing children, children with NF1 and children with ADHD showed significant impairment on planning and problem solving on the Tower of London. When examining relations between report of inattention and planning and problem-solving deficits, there were no significant relations, which indicates that the deficits in problem and planning solving was not

related to inattention. Despite using a small sample size, this study lends further support for a core deficit in EF for children with NF1.

In summary, the medical, cognitive, and behavioral profile of NF1, while highly variable, is notable for significant difficulties in the areas of learning, attention, and EF. There is increasing evidence for a core deficit in EF in children with NF1. While inhibition is the most commonly reported EF impairment in children and adolescents with NF1, impairments in working memory, cognitive flexibility, shifting, and planning/organization have also been described. Evidence for significant EF impairments, independent of ADHD diagnosis, has recently accumulated. If EF impairments cannot be accounted for by ADHD diagnosis alone, then examination of potential contributions to EF in children with NF1 is warranted.

Contributions to Executive Functioning

Examination of the relations between clinical variables of NF1 and the degree of specific cognitive impairments in NF1 have produced little to no explanation as to why children with NF1 display EF impairments. Previous studies have examined factors such as gender, mode of inheritance (familial or sporadic), the presence of macrocephaly, clinical severity, and the degree of cognitive impairment and have found no consistent significant predictors of cognitive dysfunction (Ferner et al., 1996; Hyman et al., 2005; North et al., 1995).

Variability in EF performance can arise due to numerous factors given the gradual maturation of the prefrontal cortex, the area of the brain most commonly related to EF and given the vast interconnectedness of other brain regions to the prefrontal cortex. In the typically developing literature, important contributing factors to the cognitive and behavioral profile of a child's functioning are the presence of salient factors known to influence EF, such as early life conditions, including socioeconomic status (SES), internalizing problems, attention, and sleep

(Noble et al., 2007; Rhoades et al., 2011; Blunden et al., 2005; Wagner et al., 2015b; Craske, 2012).

It has been well studied and documented how central the role of experience plays in the development of the brain beginning early in infancy and continuing through critical developmental periods into young adulthood. Evidence clearly suggests that early adversity negatively affect the development of a child's ability to efficiently process cognitive information and regulate behavior (Hackman & Farah, 2009; Hackman et al., 2015). Children and adolescents with limited SES resources and those from minority populations have disadvantages in their EF skills compared to same-aged peers from higher SES backgrounds and those from majority populations (Finn & Rock, 1997; Noble, Farah, & McCandliss, 2006; Roy & Raver, 2014). Furthermore, studies have shown a positive relationship between SES and EF (Mezzacappa, 2004; Noble, McCandliss, & Farah, 2007; Noble, Norman, & Farah, 2005), which is also consistent with research demonstrating positive relations between adverse environmental experiences and frontal lobe deficits (Hackman & Farah, 2009). Notable differences with regards to development of working memory and inhibitory control processes have been observed (Noble et al., 2007; Hackman & Farah, 2009). In a recent study by Berthelsen and colleagues (2017), contributions of child and family factors in early childhood, including SES, was examined in relation to development of EF during adolescence using longitudinal data of two cohorts of approximately 5,000 children. Results indicated that children who already exhibited a behavioral risk, had sleep problems, displayed emotional dysregulation and hyperactivity/impulsivity, and whose families had lower SES, poorer maternal mental health and poorer parenting, had significant self-regulation deficits at young ages, which were directly associated with global EF deficits in adolescence. Overall, the culmination of factors relating to behavior in early

childhood, paired with low SES, provides important information about the predictive utility of these early factors to later functioning. Despite this evidence in the typically developing literature, SES has not been systematically examined in relation to EF performance in children and adolescents with NF1.

There is evidence in the typically developing literature that children and adolescents with EF impairments and ADHD have high rates of internalizing problems, such as anxiety and depression (see Wagner et al., 2015b and Craske, 2012 for review). Impairments in inhibition (Brooks et al., 2010; Maalouf et al., 2011), working memory (Brooks et al., 2010; Klimkeith et al., 2011), shifting (Günther et al., 2011; Micco et al., 2009), and sustained attention (Cataldo et al., 2005; Günther et al., 2011; Micco et al., 2009) have all been described in children and adolescents with internalizing problems. In children and adolescents with NF1, there is evidence for increased anxiety and depressive symptoms based on caregiver report (Johnson et al., 1999; Graf et al., 2006; Barton and North, 2007); however, results from self-report in older children and adolescents is more mixed. In a study of children and adolescents, maternal report of withdrawal symptoms was the only clinically significant elevation, and paternal and child self-reports did not show significant differences between participants with and without NF1 (Noll et al., 2007). As such, continued examination of the influence of anxiety and depression symptoms on EF performance in children and adolescents with NF1 using multiple methods and informants is necessary.

It has long been known that sleep plays an integral role in the growth, development, learning, and behavior of children, and is essential for the developing brain and learning process (Dahl, 1996; Blunden et al., 2005; Cirelli & Tononi, 2008; Harvey & McGlinchey, 2015). Sleep disorders are generally classified into difficulties falling asleep, disorders of arousal, excessive

daytime somnolence, sleep initiation and maintenance, sleep breathing disorders, sleep-wake transition disorders, and sleep hyperhidrosis. Prevalence rates of childhood sleep problems in the general population are 25-40%, with problems including difficulty maintaining sleep, sleepwalking, night terrors, nightmares, teeth grinding, hyperhidrosis, insomnia, and insufficient sleep (Jenni & O'Connor, 2005). Evidence suggests clear associations between children's sleep, learning, attention, and behavior functions across development (Gozal, 1998; Gozal & Pope, 2001; Ravid et al, 2009), and have been shown to exacerbate psychosocial and neuropsychological functioning, such as depression, anxiety, attention, EF, academic function, and social development (Beebe, 2011). Children presenting to clinicians with sleep disturbances frequently score significantly lower on tests of intelligence, academic performance, and EF compared to children without sleep disturbances (Gozal, 1998; Wolfson & Carskadon, 1998; Bourke et al, 2011). Williams and colleagues (2017) examined the associations between sleep and self-regulation in a longitudinal study using caregiver report data collected from infancy to age 9. Results suggest relations between early problem behaviors and self-regulation and EF development over time. Overall, given that a substantial proportion of children exhibit sleep problems at some point during childhood, there is a great need to consider sleep-related factors within the context of a child's cognitive and behavioral functioning.

Sleep problems characterized by difficulty initiating and maintaining sleep are reported in approximately 25-50% of children and adolescents with ADHD (Marcotte et al, 1998; Corkum et al., 1998). Furthermore, as aforementioned, there is well established evidence that both insufficient and poor-quality sleep results in behavioral dysregulation that affects a range of neuropsychological functions, but especially attention and EF (Fallone et al., 2002). This seemingly bidirectional relationship between sleep problems and ADHD has vast implications

for the manifestation of ADHD symptomatology; however, given the complexity of this association, it is not clear whether difficulties arising from sleep problems make existing ADHD symptoms worse in all children, or only in a subset of children with ADHD. What is clear from this literature is that the high rates of comorbidity between children with ADHD and sleep problems warrants regular assessment of every child with ADHD for sleep problems, and that children who present with both ADHD and sleep problems be systemically evaluated.

Research has also suggested that children with neurodevelopmental disorders, including children with NF1, are at an increased risk for sleep-related problems (Johnson et al, 2007; Marcotte et al, 1998; Licis et al, 2013). Only two studies have assessed sleep-related problems in children with NF1, which is surprising given the clear evidence that high rates of comorbidity between ADHD and sleep problems exist, and children with NF1, as a group, have higher rates of ADHD. Results from previous studies of children with NF1 indicate that children with NF1 have increased sleep-related problems (Johnson et al, 2007; Licis et al, 2013). In the first study to examine sleep in children with NF1, Johnson and colleagues (2007) examined parent report data from 64 children, ages 3-18 years, with NF1 and concluded that sleepwalking and sleep terrors were more prevalent in the children with NF1 compared to population norms. In addition, significant relations between sleep problems and parent reported problem behaviors were evident. More recently, these findings were replicated and extended in a larger sample of children with NF1 using a contrast group. Licis and colleagues (2013) examined parent report of sleep in 129 children with NF1 and 89 unaffected siblings between the ages of 2-17 and found that children with NF1 had increased difficulty with initiating and maintaining sleep, transitioning between sleep and wakefulness, arousal, and hyperhidrosis compared to unaffected siblings. Children with NF1 were reported to have a more disruptive sleep schedule,

characterized by reduced sleep duration, more night awakenings, and longer type to sleep onset. It was also reported that cognitive functioning, ADHD, and stimulant medications did not affect overall sleep scores.

Overall, while there are numerous factors that may contribute to EF performance in children, evidence from the typically developing literature points to a strong influence of home environment factors, such as SES to the development of EF in early childhood. Accumulating evidence suggests that internalizing problems, including depression and anxiety, and sleep problems also contribute to the development and performance of EF. While this has been clearly demonstrated in the typically developing population, there is a relatively less literature systematically examining these potential contributions to EF in children and adolescents with NF1. To our knowledge, there have been no studies that have systematically examined SES in relation to EF performance in children with NF1. While there is some evidence that children and adolescents with NF1 have increased rates of anxiety and depressive symptoms, the extent to which these symptoms contribute to ADHD and EF performance in children with NF1 is not yet understood. In the only two studies that have examined sleep problems in children with NF1, results indicate that children with NF1 demonstrate significantly more sleep problems when compared to the normative mean and data from unaffected siblings; however, these sleep problems have not been systematically examined in relations to their effect on EF in children with NF1.

Executive Functioning

The construct of EF has yet to be conceptualized using a single, widely accepted model, but rather, is the term used for the diverse set of cognitive processes that underlie goal-directed behavior (Miyake et al., 2000). The construct of EF has evolved over time into an umbrella term

encapsulating at least 30 different definitions. The current dominant theory of EF comes from Miyake and colleagues (2000), who utilized a latent variable approach to methodically examine overlap in task performance between related EF tasks of EF; and two major themes emerged. First, EF involves higher-order, complex cognitive processes; and second, EF involves a "central-executive" component (Miyake et al., 2000). The complex cognitive processes described in this theory include the ability to plan, problem solve, inhibit inappropriate responses through self-regulation, flexibly shift mental set, and effectively organize goal-directed behavior in both short-term and long-term timeframes. In addition, the theory notes that attention and memory processes that guide these cognitive processes, such as working memory, selective attention, and sustained attention, should also be incorporated within the definition of EF. Therefore, foundational cognitive processes include inhibition, working memory, and shifting, with closely related processes including planning, divided attention, self-monitoring, self-regulation, and initiation (Stuss & Alexander; Miyake et al., 2000; Best & Miller, 2010).

EF is strongly associated with the prefrontal cortex, which has been illustrated in studies describing patients with prefrontal cortex damage who display EF deficits, yet have average cognitive functioning (e.g. Stuss & Benson, 1984), and has been extensively studied (Goldstein et al., 2013; Brocki & Bohlin, 2004; Casey et al., 2000; Tranel et al., 1994; Welsh, Pennington & Groisser, 1991). However, given the extensive inter-connectivity of the prefrontal cortex with other regions of the brain, including the basal ganglia, anterior cingulate gyrus, cerebellum, and thalamus (Baddeley & Della Sala, 1998), EF is highly susceptible to disruptions in multiple brain regions that lead to significant variability in functioning.

Development of Executive Functioning

In typically developing children, EF abilities emerge and develop rapidly during the first year of life, and continue developing gradually in stages (Diamond, 1991). The processes related to EF within this first year of life include recognition of patterns in the environment, and the ability to spontaneously form categories of events and event sequences (Diamond, 2002). The next stage of rapid development of EF is in the preschool years (ages 4-6 years) and involves rapid advancement in logical thought processes, verbal mediation, working memory, and selective attention (Welsh et al., 1991). This second stage of rapid development is noteworthy for the maturation of several different EF processes (Best et al., 2009), and has been studied extensively for the past decade (see Isquith et al., 2005 for review). Additionally, it is during this stage of development that EF become more integrated and increasingly related to self-regulatory behaviors, which has been described as occurring partially due to the development of attention (Diamond, 1991; Garon et al., 2008; Welsh & Pennington, 1988). The last stage of rapid development of EF occur in adolescence and is characterized by further refinement of EF due to pruning of the frontal neural systems (Luna et al., 2010).

Integral work by Senn, Espy, and Kaufman (2004) showed that relations among EF processes change over the course of development. As part of a longitudinal study, children ages 2 to 6 years of age were assessed on measures of inhibition, working memory, and shifting. When scores on a performance-based task were split based on age (i.e. ages 2-4, ages 5-6) different patterns of relations were found. For the younger group, only inhibition predicted problem solving on a performance-based task. However, for older children, only working memory predicted problem solving on the same task. The authors concluded that these findings demonstrate the differential course of development for EF processes. This work also

demonstrates that various EF skills can be drawn on to solve complex problems depending on the current developmental stage.

The EF abilities of children have been of great interest over the past decade, and research examining EF in both typically developing and clinical groups in childhood and adolescence have been ample. However, despite the multitude of research, the literature has significant limitations that pose problems when determining the developmental trajectories of specific EF processes. Much of the EF literature has focused on that second, rapid stage of EF development in preschoolers, which not only paints an incomplete picture of the full developmental trajectory, but also limits the extent to which conclusions about the sequences and mechanisms of development across the trajectory occur. Despite inconsistencies in the literature with regards to these methodological and theoretical issues, it has been demonstrated that inhibition improves during the preschool years and shows significantly less change later in childhood (Romine & Reynolds, 2005). On the other hand, working memory and shifting tend to show a more gradual linear improvement throughout a child's development and into adolescence (Kwon et al., 2002; Luciana et al., 2005).

EF deficits have been observed across numerous pediatric medical and development disorders, which is not surprising given the maturation of EF spans the entire course of childhood and into young adulthood. Children with medical and/or developmental disorders are also particularly at risk due to the complexity and widespread neurological underpinnings spanning the cortical and subcortical structures (Makris et al., 2007; Miller & Cummings, 2007), such that the very nature of the inter-connectivity of the prefrontal cortex with the rest of the brain increases the likelihood that any insult to the brain is likely to result in poorer EF. For example, despite controlling for IQ, EF deficits have been associated with several genetic disorders, such

as Turner syndrome (Romans, 1997), NF1 (Remigereau et al., 2017), Fragile X syndrome (Mazzocco et al., 1993), as well as acquired disorders, such as traumatic brain injury (Sykes et al., 1997) and frontal lobe lesions (Eslinger et al., 1999). As previously indicated, EF deficits are also common in certain developmental disorders, such as ADHD (Pennington & Ozonoff, 1996; Barkley et al., 1997) and ASD (Pennington & Ozonoff, 1996; Bishop, 1993).

Developmental differences in EF have been proposed as central deficits in ADHD. ADHD is one of the most common developmental disorders of childhood, with a prevalence estimate of 7.2% (Thomas et al., 2015). Characterized by symptoms of inattention, hyperactivity, and impulsivity, ADHD is thought to be sustained by excessive and inappropriate situational motor behavior, limited inhibitory control of responses, and an inability to focus, sustain, and switch attention (Barkley, 1997; Frank 1996; Biederman et al., 2006). Numerous authors have proposed the notion that ADHD symptomatology arise from a primary deficit in EF (Barkley, 1997; Castellanos and Tannock, 2002; Pennington and Ozonoff, 1996). Behavioral inhibition, in particular, has been defined as the primary deficit in ADHD (Barkley, 1997). Barkley's (1997) model refers to behavioral inhibition as interrelated processes of inhibiting a prepotent response, stopping an ongoing response, and interference control. Deficits in behavioral inhibition are related to secondary impairments in working memory, self-regulation, internalization of speech, and reconstitution (Barkley, 1997, p. 68). In their formative meta-analytic review of the neuropsychological correlates of ADHD, Pennington and Ozonoff (1996) found that children with ADHD performed significantly worse than three comparison groups comprised of individuals with conduct disorder, ASD, and Tourette syndrome on over two-thirds of the EF measures administered. Specific weaknesses for individuals with ADHD were apparent on measures of vigilance, processing speed, and motor inhibition. These areas of specific

weaknesses have continued to be confirmed by other researchers in additional studies of children with ADHD (Corbett et al., 2009; Shallice et al., 2002; Kasper et al., 2012), which is further evidence that ADHD is characterized by deficits in vigilance and inhibitory control. However, more global EF deficits in the areas of working memory, cognitive flexibility, and planning have also been reported in children and adolescents with ADHD (Sergeant et al., 2002). In a comprehensive meta-analytic review, Willcutt and colleagues (2005) determined that EF impairment in the areas of inhibition, planning, vigilance, and working memory play an important role in the neuropsychology of ADHD; however, EF weaknesses were neither necessary nor sufficient to cause all cases of ADHD. While these results clearly reiterate the notion that EF impairment is associated with ADHD and remain an important component to the ADHD behavioral profile, the hypothesis that EF deficits alone are sufficient to cause ADHD in all individuals is unsupported.

Assessment of Executive Functioning

Results from EF assessment can provide information that could not otherwise be obtained relating to a child's overall ability, motivation, and potential; and this is particularly true for children and adolescents. Despite the vast utility of EF assessment in children with NF1, selection of appropriate tools to assess EF can be challenging given the complex mechanisms of EF across age groups. Furthermore, not only is the assessment of EF plagued with the issue of finding appropriate measures to assess EF across a wide age range and span of development, but it is also overwhelmed with validity issues. Both performance-based measures and functional behavior rating measures are commonly used in clinical and neuropsychological assessments and are intended to measure the same underlying construct of EF. Despite both being commonly utilized, the most conventional method for assessing EF is through use of performance-based

measures (Pennington & Ozonoff, 1996). Performance-based measures are typically conducted in a highly structured one-on-one setting, making them advantageous for their controlled and structured environment. Performance on these measures are typically based on the examinee's accuracy, response time, and processing speed under a time constraint.

One of the greatest advantages to using performance-based measures of EF can also be considered a limitation. The highly structured, one-on-one setting that performance-based measures are administered allows for the possibility of one to perform better than they would in more realistic, less structured environments. The structured nature of a typical assessment does not place high demands on EF, thus reducing the opportunity for observing behavior related to more everyday EF (Holmes-Bernstein & Waber, 1997). Furthermore, it makes it even more challenging to hone in on difficulties that are observed in less structured environments (Salthouse et al., 2003). It has been posited that the highly controlled environment, itself, acts as its own frontal lobe, allowing for more optimal performance than in a less structured environment (Salthouse et al., 2003). A second issue with using these measures is involves task impurity. EF measurement involves invoking cognitive processes within other domains during tasks that assess EF due to the very nature of EF as a construct. In fact, it has been demonstrated that many measures of EF involve non-executive processes as part of the task, such as color naming in the Stroop task (Miyake & Friedman, 2012). Lastly, a third issue with using performance-based measures involves the idea that novelty is a key characteristic of many EF tasks; and therefore, the degree to which various EF tasks are novel to an individual varies significantly depending on that individual's personal experience. Different experiences likely lend themselves to different strategies for the same task (Hughes, 2002). As such, this is likely the reason that EF measures have been found to be weakly correlated among themselves,

particularly over time or with one another (Hughes, 2002; Miyake et al., 2000). For these reasons, a child's everyday environment, such as in the home or at school, may provide more useful venues for observing the true essence of aggregate everyday EF, making caregivers' report of behaviors in these environments highly valuable.

Functional behavior rating scales are also commonly used to assess EF and are thought to prevent some of the methodological issues that often accompany performance-based measures. Within the past decade, several studies examining child populations have noted the clinical utility of the Behavior Rating Inventory of Executive Function (BRIEF; Gioia et al., 2000) and its ability to distinguish between clinical groups, such as children with ADHD, traumatic brain injury, frontal lesions, and ASD (Toplak et al., 2012). Functional measures of EF are assumed to measure behaviors that are significantly related with the processes that are assessed by performance-based measures of EF. Functional measures can be completed by observers, such as parents and/or teachers, and can also be completed by the child depending on his/her age. The BRIEF (Gioia et al., 2000) is the most commonly utilized functional measure of EF. It is composed of eight individual scales and three composite scores assessing behaviors on scales of inhibition, shifting, emotional control, initiation, working memory, planning/organization, organization of materials, and monitoring. The BRIEF has been demonstrated to adequately capture the expected patterns of EF in diverse clinical populations and have correlated with biological markers associated with executive function, thus, providing evidence of relations between EF and real-world behavior (Isquith, Roth, & Gioia, 2013).

While ratings of everyday functional behavior provide useful insight into a child's typical, everyday behavior, there are issues that exist when relying on these measures alone. First, raters completing ratings have limited control of environmental influences that could affect

ratings. An individual who is described as having EF impairments in a highly demanding environment might be described as having sufficient EF abilities in a less demanding environment, for example. Second, given the integrative nature of EF, particularly within an everyday context, functional rating scales make it difficult to parse out impairments in specific EF processes compared to relatively narrower focused performance measures (Gioia et al., 2016). Third, the observant/rater's perspective requires consideration when interpreting functional behavior, which lead to inconsistencies in reports from different informants (Dibartolo & Grills, 2006).

Performance-based measures of EF and functional behavior rating scales are presumed to assess the same construct, and thus, should be significantly positively correlated with each other; however, results from several studies point to a different conclusion. Studies that have examined relations between performance-based and functional measures of EF have generally reported minimal to no convergence. As reported in a review by Toplak and colleagues (2013), of the 12 studies that have examined relations between the BRIEF with several commonly used EF performance measures in childhood, there were "extremely weak" to no relations between measures. Indeed, it has been argued that these two different types of measurements may assess different underlying constructs of EF (Chevignard et al., 2012; Toplak et al., 2013).

Summary and Study Rationale

For the past decade, the presence of ADHD has increasingly emerged as a characteristic feature of the NF1 neuropsychological phenotype. ADHD and EF abilities are closely interrelated across many levels, skillsets, and tasks. While ADHD is characterized by EF deficits, it is important to highlight that not every EF deficit stems from problems with attention or

impulse control. This has been demonstrated through findings that children with NF1 evidence EF deficits independent of a comorbid ADHD diagnosis. However, less is understood about what factors contribute to EF performance in children with NF1. While ADHD is likely to explain some of the difficulties in EF in children with NF1, evidence suggests that ADHD does not explain all the variance. Examination of the relations between EF performance and attention, cognitive functioning, anxiety and depression, various sleep factors, and SES, in children with NF1 is warranted. In addition, while performance-based measures are useful in providing information regarding a child's ability under optimal conditions, functional behavior measures can provide us with aggregate information on how the child typically functions day-to-day. Relatedly, most NF1 studies examining EF in children have done so using performance-based measures of EF, which may limit the extent to which difficulties with EF are detected. As such, assessment of EF in children with NF1 using multiple methods is warranted.

Methods

Study Aims & Hypotheses

Aim 1: Characterize EF performance in our sample of children with NF1. Results from both performance-based and functional measures of EF will be described. Specifically, the number of children who display performance-based EF impairment (at least 1 standard deviation below the mean) will be reported. The number of children with behavior in the "at-risk" and "clinically elevated" range on the functional parent-reported EF measure will also be described. In addition, the mean scores for all EF measures will be reported. It is hypothesized that children with NF1 will display EF impairment on standardized performance-based measures. It is also expected that functional executive dysfunction across EF scales on the BRIEF will be commonly reported by parents.

Aim 2: Correspondence between performance and functional measures. Relations between scores obtained from EF performance-based measures and functional parent-reported measure will be examined. Mean performance scores and the percentage of children who display EF impairment (at least 1 standard deviation below the mean) on performance-based measures will also be compared and contrasted with the number of children reported to demonstrate behavior in the "at-risk" and "clinically elevated" range on the functional parent-reported BRIEF to determine whether children with NF1 display impairment more on performance-based or functional parent reported measures of EF. It is hypothesized that scores on EF performance-based and on functional parent reported measures will show small interrelations. It is also expected that our sample of children with NF1 will display more difficulties based on functional parent report compared to performance-based measures.

Aim 3: Examine potential neuropsychological, sociodemographic, and psychosocial factors that contribute to EF performance.

Attention Problems. To assess whether attention problems are related to EF in our sample of children with NF1, performance-based and functional EF measures will be examined for children with NF1 with and without ADHD separately. In addition to examination of EF performance based on ADHD diagnosis, the contribution of ADHD symptomatology will also be examined using dimensional scales from a parent-reported measure. It is hypothesized that children with NF1 will exhibit EF impairment, independent of ADHD diagnosis; however, it is expected that EF performance and report of dimensional ADHD symptoms will be significantly related.

General Cognitive Functioning. To examine the role of general cognitive functioning to EF, general cognitive ability scores will be examined in relation to performance-based and

functional EF. It is expected that general cognitive functioning and EF will be significantly related in our sample of children with NF1.

Socioeconomic status and Maternal Education. Examination of SES and maternal education in relation to EF on performance-based and functional EF measures will be conducted. While exploratory in nature, given previous work in the typically developing population, it is expected that SES and maternal education and EF performance will be significantly related in our sample of children with NF1.

Anxiety and Depression. To examine the contributing role of internalizing problems to EF performance, parent-reported anxiety and depression will be examined in relation to performance-based and functional EF. While exploratory in nature, it is expected that anxiety and depression symptoms will be significantly related to EF performance in our sample of children with NF1.

Sleep. The role of sleep and the presence of sleep problems will be examined. Performance-based and functional EF will be examined to determine whether children who meet cut-off criteria for SRBD demonstrate significantly different EF performance compared to children without SRBD. Specific sleep-related problems from the PSQ will also be examined in relation to performance-based and functional EF. It is hypothesized that children who meet cut-off criteria for SRBD will display significantly more EF impairments and that sleep-related problems will be related to EF performance.

Relative Contributions. Lastly, to get a sense of the degree to which ADHD, SES, anxiety/depression, and/or the presence of sleep problems predict EF impairment in our sample of children with NF1, two multiple regression analyses will be conducted. The first regression will include the performance-based Toolbox Flanker as the dependent variable, given extensive

literature supporting the notion that inhibitory processes are primary EF constructs and other EF processes are highly associated to inhibition. The second regression will include functional parent reported measure BRIEF GEC as the dependent variable. However, if high intercorrelations among EF measures are present ($r \geq .7$), a composite performance-based EF score may be calculated and used in the regression analysis as the dependent variable in place of the Toolbox Flanker and BRIEF GEC. Specific predictors of the models will be determined based on results of aforementioned analyses. This aim is exploratory in nature.

Participants

Participants included 40 children between the ages of 9 and 13 diagnosed with NF1 and one parent of each child participant. Only children whose first and main language spoken in the home was English were included in the study. Children whose first and main language were not English were excluded given study measures and instructions were standardized and normed using English-speaking populations. Children who had significant surgery requiring general anesthesia within the 6 months prior to screening were excluded from this study, given that the effects of general anesthesia could have an impact on performance on study measures.

Procedures

Recruitment of participants involved three methods. The first method involved sending fliers describing the current study to participants who have participated in prior research and who consented to be informed of future studies in the lab. Second, participants were recruited through several Midwestern Neurofibromatosis Clinics. NF1 clinic directors were asked to share a description of the study to families with children between the ages of 9 and 13 with a confirmed diagnosis of NF1. Parents who were interested in participating or finding out more about the study were provided with a flier and encouraged to contact the PI or study coordinator. The third

method involved recruitment through the national Neurofibromatosis Research Registry. Families within driving distance who had noted their interest in being contacted about possible research opportunities on the registry were emailed a description of the study and a flier.

Participants who met eligibility requirements were scheduled for an evaluation at the Child Neurodevelopment Research Lab (CNRL) at the University of Wisconsin-Milwaukee or in a quiet hotel conference room near their home. Participants were consented and were given an opportunity to ask questions or express concerns before agreeing to participate. Prior to the assessment appointment, the consent form and questionnaires were mailed to the family for parental completion. The questionnaires of interest for the study were designed to examine ADHD symptoms, everyday EF abilities, sleep-related difficulties, and anxiety/depression symptoms. Each child was administered an age-appropriate neuropsychological battery by a trained member of the study team. Assessment sessions lasted approximately 4 hours for all children, including time for breaks to minimize fatigue. All assessment measures were administered to children in the same order. Parents were interviewed about their child's behavior during their child's assessment in an adjacent room.

Measures

All measures chosen for this study were developed for use with children 9 to 13 years and are widely used in pediatric assessment and research both in typically developing populations and children with a variety of developmental disorders. All neuropsychological measures are norm-referenced and have demonstrated strong psychometric properties, including good reliability and validity. The measures selected were chosen to provide information about children's attention and EF abilities, as well as sociodemographic and psychosocial factors.

These measures were selected to pick up on both obvious impairments, as well as more subtle difficulties that are commonly found in children with NF1. A detailed description of each measure is provided below, and a summary of the measures is provided in Table 2.

Differential Ability Scales-Second Edition: School-Age Form (DAS-II; Elliot, 1990). The DAS-II is a commonly used, comprehensive measure of cognitive abilities for children ages 7-0 to 17-11. The DAS-II is empirically derived and demonstrates excellent internal consistency, test re-test reliability and correlates highly with other commonly used measures of cognitive abilities (Elliot, 1990). The DAS-II provides normative data collected on a large representative national sample and contains excellent floor and ceiling levels, making it appropriate for children with neurodevelopmental disorders. This measure yields an overall composite score called the General Conceptual Ability (GCA) standard score (M = 100, SD = 15) that is equivalent to a full-scale IQ score. The GCA is broken down into three cluster scores, including Verbal Ability, Nonverbal Reasoning Ability, and Spatial Ability. In this study, participants completed the core subtests for the School-Age Form (including Word Definitions, Verbal Comprehension, Matrices, Sequential and Quantitative Reasoning, Recall of Designs, and Pattern Construction) to yield a GCA. In addition, subtests for the Working Memory cluster (Recall of Sequential Order, Recall of Digits Backward) were administered.

NEPSY – Second Edition: Auditory Attention/Response Set (NEPSY-II; Korkman, Kirk, & Kemp, 2007). The NEPSY-II is a widely-used measure that assesses children's performance in areas of six theoretically derived domains, including Attention and Executive Functioning, Language, Memory and Learning, Sensorimotor, Social Perception, and Visuospatial function. Administration of selected subtests takes approximately 5-10 minutes and is designed for children 3-16 years old. The Auditory Attention/Response Set (AA/RS) subtest

from the Attention domain has two parts; Auditory Attention which was designed to assess sustained, selective auditory attention and Response Set which was designed to assess shifting and sustained attention skills. Both subtest parts yield an overall scaled score (M = 10, SD = 3).

Cogstate Research Battery (Cogstate; http://www.cogstate.com), selected subtests. The Cogstate battery is a commercially available, computerized cognitive testing system designed specifically for the use in research studies. Cogstate tasks have been shown to be highly reliable and sensitive. The entire Cogstate testing battery targets a wide range of cognitive domains, including processing speed, attention, EF, and social-emotional cognition. The Visual Attention/Vigilance and Attention/Working Memory tasks were administered for the current study, including the Identification Task (ID).The ID task, designed to assess simple visual attention and vigilance and takes approximately 2 minutes to complete. The primary outcome measure of ID is log transformed reaction time, in which lower times indicate better performance. The One Back Task (ONB), designed to assess working memory and sustained visual attention, was also completed. The ONB takes approximately 4 minutes to complete. The primary outcome measure for ONB is Arcsine transformed accuracy, in which higher scores indicate better performance. The ONB task also provides log transformed reaction time, in which lower times indicate better performance. All Cogstate outcome scores were converted to z-scores (M = 0, SD = 1) for analyses.

NIH Toolbox (http://www.nihtoolbox.org), selected subtests. The NIH Toolbox is a comprehensive set of neurobehavioral measurements that quickly assess cognitive, emotional, sensory, and motor functions. NIH Toolbox is based on a nationally representative sample and has been validated to be psychometrically sound. For the current study, the List Sorting Working Memory (LSWM), Dimensional Change Card Sort Test (DCCS), and Flanker Inhibitory Control

and Attention Test (Flanker) tasks were administered. LSWM is a 7-minute working memory task where participants recall and sequence different visually and orally presented stimuli. The DCCS is a 4-minute measure of cognitive flexibility and attention. Pictures are presented varying along two dimensions (e.g. shape and color) and participants are required to sort based on a cue word on the screen. Flanker is a 3-minute attention and inhibitory control task where participants are required to focus on a given stimulus while inhibiting attention to nearby stimuli. For the Flanker, if accuracy levels were less than or equal to 80%, the final "total" computed score was equal to the accuracy score. If accuracy levels for the participant reached more than 80%, the reaction time score and accuracy score were combined (Zelazo et al., 2013). All NIH Toolbox outcome scores were standard scores (M = 100, SD = 15).

Spence Children's Anxiety Scale (SCAS; Spence, 1998). The SCAS is a parent-reported measure designed to assess the severity of childhood anxiety symptoms. The SCAS assesses six domains of anxiety including generalized anxiety, panic/agoraphobia, social phobia, separation anxiety, obsessive compulsive disorder, and physical injury fears; and provides an overall total anxiety score. The SCAS has been widely used in research and clinical contexts for assessment purposes. While not a diagnostic measure, the SCAS was designed to provide an indication of the nature and extent of anxiety symptoms to assist in the diagnostic process. Total SCAS raw scores will be used for correlational analyses.

Pediatric Sleep Questionnaire (PSQ; Chervin et al., 2000). The PSQ is a parent-reported measure of pediatric sleep disorder symptoms. The PSQ was designed as a broad clinical screen for research purposes and has demonstrated good reliability and validity. The presence of various sleep-related problems, as well as the overall Sleep-Related Breathing Disorder (SRBD) scale was examined. The SRBD scale consists of 22 symptoms items from the

PSQ that inquire about snoring frequency, loud snoring, observed apneas, difficulty breathing during sleep, sleepiness, daytime behavior, and other pediatric obstructive sleep apnea features. The SRBD scale was developed for clinical research purposes and has been validated against polysomnography (Chervin et al., 2006). The number of 22 symptom-items endorsed positively is divided by the number of items answered positively or negatively, so that missing responses are excluded. The result is a proportion that ranges from 0.0 to 1.0. Scores >.33 are considered positive and are suggestive of high risk for a pediatric sleep-related breathing disorder. This threshold is based on a validity study (Chervin et al., 2000) that suggested optimal sensitivity and specificity at the 0.33 cut-off.

Kiddie Schedule for Affective Disorders and Schizophrenia – Present and Lifetime (KSADS-PL; Kaufman et al., 1997)- ADHD Section. The KSADS-PL is a semi-structured clinical interview administered to parents. The ADHD section was administered to parents to assess ADHD symptomatology, including inattention and hyperactivity/impulsivity symptoms.

Behavior Assessment System for Children – Second Edition (BASC-2; Reynolds & Kamphaus, 2004). The BASC-2 is a commonly used screener of childhood problem behaviors. The BASC-2 was administered to parents. Scales of interest for the current study include the Anxiety, Depression, Somatization, and Withdrawal content scales, as well as the Internalizing Problems composite scale. The BASC-2 yields t-scores (M = 50, SD = 10).

Behavior Rating Inventory of Executive Function (BRIEF; Gioia et al., 2000). The BRIEF is a parent rating scale developed to provide a glimpse into everyday behaviors associated with EF in the home and school environments. The BRIEF yields 8 clinical scales: Inhibit, Shift, Emotional Control, Initiate, Working Memory, Plan/Organize, Organization of Materials, and Monitor. These clinical scales form two broader indexes, the Behavioral

Regulation Index (BRI) and the Metacognition Index (MI), and an overall score, the Global Executive Composite (GEC). The BRIEF yields t-scores (M = 50, SD = 10). Normal scores include t-scores ≤ 59, at-risk scores include t-scores 60-64, and clinically elevated scores include t-scores ≥ 65).

Conners Parent Short Form, Third Edition (Conners-3 Short; Conners, 2008). The Conners-3 Short is a parent reported measure that assesses ADHD and its most common comorbid problems and disorders for children and adolescents 6-18 years. The Conners-3 Short is meant to assess children's behavior across multiple settings. For the purposes of this study, scales assessed include the ADHD Inattentive and ADHD Hyperactive-Impulsive scales. The Conners-3 Short yields t-scores (M = 50, SD = 10).

Hollingshead Four-Factor Index of Socioeconomic Status (Hollingshead; Hollingshead, 1975). The Hollingshead is a survey designed to measure the SES of an individual based on four domains: marital status, employment status, educational attainment, and occupation. Information gathered from children's parent(s) based on these domains were coded by examiners to calculate a total parental SES score. The Hollingshead is widely used in psychosocial research and has demonstrate adequate internal consistency and strong cross-sectional convergent validity based on 1970 census data.

Background Questionnaire. The CNRL Background Questionnaire is a comprehensive parent-completed questionnaire used to collect demographic information, which may aid in analysis of data (examining differences based on, for example, parental education or child medical history).

Results

The data were analyzed using IBM SPSS for Mac, version 26. Spearman's rho was used when correlational analyses were conducted and interpretations of correlation effect size (Cohen, 1988) are as follows: small = 0.1 – 0.3; medium = 0.3 - 0.5; large = 0.5 – 1. Given the number of comparisons made, the False Discovery Rate approach (Benjamini & Hochberg, 1995; Pike, 2011) was used to determine a q-value adjusted for the number of comparisons within each set of analyses with multiple comparisons, and these q-values were compared with alpha = .05 to determine statistical significance.

Demographics and Individual Differences. See Table 3 for complete participant demographics. No significant differences between sporadic and familial etiology of NF1 were evident when examining SES, $t(39) = -1.84$, $p = .089$; mother education, $t(39) = -1.98$, $p = .844$; general cognitive functioning (DAS-II GCA), $t(39) = .462$, $p = .616$, or ADHD diagnosis, $t(39) = .771$, $p = .253$. There were also no significant differences in scores on any performance-based EF measure between sporadic and familial etiology of NF1, $t(39) = .045 – 1.42$, $p = .163 - .965$; nor on any functional EF BRIEF scale or index, $t(39) = .170 – 1.62$, $p = .113 - .866$.

EF in Children with NF1.

Aim 1: Characterize EF performance in our sample of children with NF1. A summary of parent-reported BRIEF scores sampling functional EF is detailed in Table 4. Group mean scores fell in the average range for all domain scores; however, independent one-sample t-tests indicated significantly higher scores than the normative mean on the Inhibit, Initiate, Working Memory, Plan/Organize, Organization of Materials, and Monitor domains; and on the Behavioral Regulation, Metacognition, and GEC indices. No significant differences in scores

from the normative mean were evident when examining the Shift and Emotional Control domains. Figure 1 details the distribution of normal (≤ 59), at-risk (60-64), and clinically elevated (≥ 65) problems reported by parents on the BRIEF. On the BRIEF GEC, 6 children (15%) were rated as demonstrating EF difficulties in the "at-risk" range; and 12 children (30%) were rated as demonstrating EF difficulties in the clinically elevated range. There were no significant relation between BRIEF scale scores and age, *rho*(40) = -.130 - .194, *p* = .961 - .234, and no significant effect of sex, *t*(39) = .033 – 1.23, *p* = .216 - .974.

A summary of scores on EF performance-based tasks is presented in Table 5. Group mean scores fell in the broadly average range across measures. Independent one-sample t-tests revealed significantly lower scores than the normative mean on DAS-II RSO, DAS-II RDB, Toolbox DCCS, Toolbox Flanker, NEPSY-II RS, and NEPSY-II AA/RS. Figure 2 details the distribution of performance-based EF scores 2 SD above the mean, 1 SD above the mean, within 1 SD of the mean, 1 SD below the mean, and 2 SD below the mean. Based on frequency of difficulties, children with NF1 demonstrated the most difficulty on Toolbox Flanker, with 14 children (39%) with scores 1 SD below the mean; and 3 children (8%) with scores 2 SD below the mean. Performance on Cogstate ID had moderate significant associations with age, *rho*(39) = -3.72, *p* = .020. No additional relations between age and performance-based measures were evident. No sex difference in scores on performance-based EF tasks was evident, *t*(36-39) = .172 – 2.06, *p* = .058 - .865.

When comparing and contrasting rates of difficulties between performance-based and functional EF, the percentage of children showing performance one standard deviation or more below the mean on tasks of inhibition, working memory, and cognitive flexibility/shifting across

measures was examined. Results revealed higher rates of impairment on the BRIEF compared to performance-based measures. Specifically, a chi-square goodness-of-fit test indicated that there was a statistically significant differences in the proportion of identified EF difficulties across measures, χ^2 (2, n = 40) = 42.23, p = <.001, when examining rates of impairments (1 SD below the mean) on the BRIEF Inhibit scale (48%) compared to NIH Toolbox Flanker (44.4%); the BRIEF Working Memory scale (37.5%) compared to NIH Toolbox LSWM (18.9%); and the BRIEF Shift scale (25%) compared to NIH Toolbox DCCS (21.6%).

Aim 2: Correspondence between performance and functional measures. To compare EF scores from performance-based and functional measures, bivariate Spearman correlations were conducted. See Table 6. Results revealed medium, negative correlations between performance on the Toolbox DCCS and the BRIEF Monitor scale, *rho*(37) = -.519, p =.044. No additional significant relations between performance-based and functional measures of EF were evident.

Aim 3: Examine potential neuropsychological, sociodemographic, and psychosocial factors that contribute to EF performance.

Attention. Bivariate Spearman correlations were also used to examine relations between parent-reported dimensional ADHD symptoms on the Conners-3 Short and functional parent-reported EF. Results revealed medium to large positive relations between dimensional inattentive symptoms and BRIEF GEC, (*rho*(40) = .628, p = < .001); BRIEF BRI and MI, (*rho*(40) = .437 - .493, p = .002 - .009); Monitor, (*rho*(40) = .596 8, p = < .001); Working Memory, (*rho*(40) = .558, p = < .001); Inhibit, (*rho*(40) = .626, p = < .001); Plan/Organize, (*rho*(40) = .423, p = .009); and Initiate, (*rho*(40) = .373, p = .024) scales. When examining parent report of inattentive symptoms, examination of relations on performance-based EF tasks revealed a

negative, moderate correlation with NEPSY RS, (*rho*(40) = -.407, *p* = .049). No additional relations were apparent. Parent-reported hyperactive/impulsive symptoms from the Conners-3 Short were also explored in relation to parent-reported functional EF. Results revealed medium to large positive correlations between hyperactive/impulsive symptoms and BRIEF GEC,(*rho*(40) = .389, *p* = .027); BRI,(*rho*(40) = .382, *p* = .027); MI, (*rho*(40) = .455, *p* =.008); Inhibit, (*rho*(40) = .545, *p* = .001); Working Memory, (*rho*(40) = .491, *p* = .005); Monitor, (*rho*(40) = .473, *p* = .007); and Plan/Organize (*rho*(40) = .368, *p* = .031). No significant relations were evident when examining associations between performance-based EF and parent report of hyperactive/impulsive symptoms.

To further examine the extent to which ADHD symptomatology is related to EF in our sample of children with NF1, group differences between children diagnosed with NF1 + ADHD and children with NF1 without ADHD were explored using multivariate analysis of variance. See Table 7. No significant effect of ADHD diagnosis was observed when examining scores on performance-based EF tasks (*F*(11, 24) =.910; *p* = .545; Wilks' λ = .706). When examining functional EF on the BRIEF, group differences were evident between children with and without ADHD, (*F*(11, 28) = 3.89; *p* = .002; Wilks' λ = .396). See Table 8 for group differences on the BRIEF. The distribution of BRIEF scores by group is illustrated in Figure 3.

General Cognitive Functioning. To better understand what neuropsychological variables could contribute to EF in children with NF1, relations between DAS-II GCA and EF variables were investigated using bivariate Spearman correlations. Results revealed medium to large positive correlations between DAS-II GCA and Cogstate ONB Accuracy, *rho*(39) = .548, *p* = <.001; LSWM, *rho*(37) = .581, *p* = < .001; DAS-II RDB, *rho*(40) = .535, *p* = <.001; Flanker, *rho*(36) = .506, *p* = .004; and DAS-II RSO, *rho*(40) = .481, *p* = .004. Examination of relations

between DAS-II GCA and parent-reported EF on the BRIEF did not reveal any significant associations (*rho* = .001 - .197, *p* = .222 - .996).

SES & Maternal Education. To better understand the extent to which sociodemographic variables contribute to EF in children with NF1, relations between SES and level of maternal education were examined in relation to EF using Bivariate Spearman correlations. When examining performance-based EF, no significant associations were present with SES, (*rho* = .030 - .227, *p* = .679 - .857), nor maternal level of education, (*rho* = .005 - .161, *p* = .812 - .976). In examination of parent-reported functional EF on the BRIEF, no significant associations were apparent in relation to SES, (*rho* = .017 - .272, *p* = .531 - .917), nor maternal level of education (*rho* = .040 - .236, *p* = .687 - .805).

Depression and Anxiety. To better understand the extent to which psychosocial variables contribute to EF in children with NF1, parent-reported depression symptoms on the BASC-2 were examined. Bivariate Spearman correlations were conducted examining relations between the BASC-2 Internalizing Problems scale and performance-based and functional EF. No significant relations were evident on performance-based EF; however, medium to large positive correlations were evident on the BRIEF Emotional Control (*rho*(40) = .557, *p* = <.001); Shift (*rho*(40) = .502, *p* = .005) scales; and the BRI (*rho*(40) = .481, *p* = .007). Bivariate Spearman correlations were also conducted to examine relations between the BASC-2 Depression scale and performance-based and functional EF. No significant relations were evident on performance-based EF when examining the BASC-2 Depression scale; however, medium to large positive correlations were evident for the BRIEF Emotional Control (*rho*(40) = .653, *p* = <.001); Shift (*rho*(40) = .460, *p* = .011) scales; and the BRI (*rho*(40) = .514, *p* = .005) and GEC (*rho*(40) = .408, *p* = .024).

Bivariate Spearman correlations were also conducted to examine parent-reported anxiety symptoms on the SCAS in relation to EF. Results revealed no significant relations between anxiety on the SCAS and performance-based EF; however, medium to large positive correlations were evident for the BRIEF Emotional Control (*rho*(40) = .535, *p* = <.001) and Shift (*rho*(40) = .683, *p* = <.001) scales; BRI (*rho*(40) = .572, *p* = <.001); and GEC (*rho*(40) = .429, *p* = .016).

Sleep. To better understand the extent to which sleep difficulties contribute to EF in children with NF1, parent report of various sleep-related difficulties on the PSQ were examined. In examination of relations to performance-based EF, parent report of restlessness during sleep did not reveal any significant correlations. Bivariate Spearman correlations revealed medium to large correlations between restlessness and functional parent-reported EF on the BRIEF Emotional Control (*rho*(40) = .543, *p* = <.001); Monitor (*rho*(40) = .478, *p* = .007); Shift (*rho*(40) = .407, *p* = .018); Inhibit (*rho*(40) = .409, *p* = .018) scales; and the BRI (*rho*(40) = .509, *p* = .005) and GEC (*rho*(40) = .416, *p* = .018). Examination of relations between difficulty falling asleep at night and both performance-based and functional EF did not reveal any significant correlations (*rho*(40) =.008 - .323, *p* = .143 - .981). For performance-based EF, bivariate Spearman correlations revealed a negative medium correlation between snoring during sleep and performance on NIH Toolbox ONB Speed, (*rho*(36) = -.496, *p* = .025). For functional EF using the BRIEF, snoring during sleep was significantly related to the BRIEF Monitor scale (*rho*(40) = .501, *p* = .022), with a medium positive correlation. No additional relations to snoring during sleep were evident.

To further explore the extent to which sleep-related difficulties are related to EF in our sample of children with NF1, group differences between children who met cut-off criteria for a SRBD on the PSQ and children who did not were explored. A total of 14 children (35%) met

cut-off criteria for SRBD based on parent report of sleep and daytime behavior. No significant effect of group was observed when examining scores on performance-based EF tasks ($F(11, 24) = .635$; $p = .782$; Wilks' $\lambda = .775$). There was also no significant effect of group when examining functional EF on the BRIEF, ($F(11, 28) = 1.87$; $p = .088$; Wilks' $\lambda = .576$); however, when dependent variables were considerately separately, significant differences between groups were evident on the BRIEF Inhibit, Working Memory, and Monitor scales; and MI. See Table 9. The distribution of BRIEF scores by group is illustrated in Figure 4.

Relative Contributions. To further explore the extent to which various factors contribute to EF in children with NF1, standard multiple regressions were conducted. First, a regression was conducted to investigate predictive variables to functional, parent-reported EF on the BRIEF GEC. Predictors of the model included overall cognitive functioning (DAS-II GCA), SES, ADHD diagnosis, sleep-related breathing difficulties (SRBD cut-off criteria from the PSQ), anxiety (SCAS Total Anxiety), and depression symptomatology (BASC-2 Depression scale). Results indicated that these variables were significant predictors of parent-reported, functional EF on the BRIEF GEC; $F(6, 33) = 5.51$, $p = <.001$. Upon examination of the unique contribution of each variable in predicting BRIEF GEC scores, BASC-II Depression scores ($\beta = .408$; $t = 2.79$; $p = .009$) and ADHD diagnosis ($\beta = .312$; $t = 2.21$; $p = .034$) uniquely contributed to BRIEF GEC.

Next, a standard multiple regression was conducted to investigate predictive variables to performance-based scores on the NIH Toolbox Flanker. Predictors of the model were the same as for functional EF and included overall cognitive functioning (DAS-II GCA), SES, ADHD diagnosis, sleep-related breathing difficulties (SRBD cut-off criteria from the PSQ), anxiety (SCAS Total Anxiety), and depression symptomatology (BASC-2 Depression scale). Results

indicated that these variables were not significant predictors of performance on the NIH Toolbox Flanker task; $F(6, 29) = 1.34$, $p = .273$. Upon examination of the unique contribution of each variable in predicting performance-based scores on the Flanker, DAS-II GCA scores uniquely contributed, ($\beta = .397$; $t = 2.40$; $p = .023$)

Discussion

Despite well-documented rates of executive dysfunction in children and adolescents with NF1, to date there exists only a preliminary understanding of factors that may contribute to executive functioning difficulties in children and adolescents with NF1. ADHD has increasingly emerged as a characteristic feature of the NF1 neuropsychological phenotype, and while ADHD typically includes deficits in certain aspects of EF, prior research has lent support to the notion that children with NF1 display EF impairments independent of a comorbid ADHD diagnosis (Huijbregts et al., 2012; Lion-Francois et al., 2017; Payne et al., 2012). In the current study, we sought to characterize EF across performance-based and functional measures and compare and contrast rates of difficulties across the two measurement methods. This multi-method design allowed for investigation into the utility of parent-reported information on how children with NF1 typically function on a daily basis, in conjunction with formalized performance-based measures of EF. As hypothesized, children with NF1, as a group, displayed EF impairment on standardized performance-based measures, and functional EF difficulties were commonly reported by parents. While EF difficulties were seen across performance-based and functional EF measures, only minimal associations were found between the two measure types, consistent with previous literature examining performance-based and functional EF measures. As expected, our sample of children with NF1 displayed more difficulties based on functional parent report compared to performance-based measures. Higher rates of difficulties were apparent based on

parent report of functional inhibition, working memory, and cognitive flexibility/shifting, compared to the level of difficulty observed on performance-based EF measures.

In addition, the present study examined potential contributing effects of SES, general cognitive functioning, attention, internalizing problems, and sleep to EF scores across functional and performance-based measures of EF. Contrary to our hypothesis, no significant relations between any EF measure, performance-based or functional, were evident when examining SES. The present study found that general cognitive functioning was associated with performance-based working memory tasks and an inhibition task. No additional relations to general cognitive functioning were evident when examining other performance-based EF tasks. No relations to any functional EF scales was evident. Over one-third of children with NF1 met diagnostic criteria for ADHD. Similar to findings from previous studies, the present study found that children with NF1 as a group demonstrate similar EF profiles on performance-based measures, independent of ADHD diagnosis. However, as expected, on functional parent reported measures of EF, children with NF1 and ADHD demonstrated significantly higher levels of executive dysfunction compared to children with NF1 without ADHD. Contrary to our original hypothesis, parent report of internalizing problems was not related to EF on performance-based measures; however, relations between depression and anxiety symptomatology and parent report of functional emotional control, shifting/cognitive flexibility, and overall behavioral regulation were found. As hypothesized, specific sleep-related difficulties were significantly related to EF in the present study. Parent report of restlessness and snoring was associated with functional EF in our sample of children with NF1. Children who were reported to snore regularly had slower reaction times on a performance-based working memory task. Group differences between children with NF1 who met criteria for SRBD and those who did not were apparent when examining parent report

of functional inhibition, working memory, and self-monitoring difficulties. Potential explanations for findings will be provided below, along with limitations, future directions, and implications.

Characterization of EF. In line with previous research, the present study found that children with NF1, as a group, demonstrated difficulty on performance-based and functional EF measures. Specifically, functional difficulties compared to the normative mean were apparent on scales sampling inhibition, initiation, working memory, planning/organization, organization of materials, and self-monitoring. Half of parents reported functional difficulties related to organization of materials; 45% reported self-monitoring difficulties; 42.5% reported planning/organization difficulties; 37.5% reported working memory and initiation difficulties, and 32.5% reported inhibition difficulties. The rates of functional EF difficulties in the present study are somewhat lower than previously reported in the NF1 population. Payne and colleagues (2011) found rates of working memory, self-monitoring, and planning/organization difficulties to be nearly 60% in their sample of children and adolescents with NF1. The difference in reported rates of functional difficulties between the present study and Payne and colleagues (2011) may be due to differences in sample size and age ranges. The present study, which used a relatively smaller sample, was designed to focus on the school-age years spanning 4 years (ages 9-13). Payne and colleagues (2011) examined the functional EF of children spanning 10 years (ages 6-16) and had a significantly larger sample size. It is possible that these differences explain the difference in rates of functional difficulties. The present study found particularly high rates of parent-reported difficulties with organization of materials. This finding differs from previous research in preschool children with NF1 that has demonstrated working memory deficits to be the most prevalent and notable parent-reported difficulty (Casnar & Klein-Tasman, 2017;

Sangster et al., 2011); however, it is important to consider these findings from a developmental perspective. Given that working memory is considered a foundational EF process, and rapid development typically occurs in the preschool-years (Welsh et al., 1991), it is reasonable to consider that working memory abilities may be particularly vulnerable in children with NF1 during the preschool years. In school-aged children, as task demands increase, secondary EF processes such as organization, remain in a developmental phase and thus, may be more susceptible to day-to-day difficulties. As children progress through various developmental stages, EF becomes increasingly differentiated into a dissociable, but related set of skills.

While more difficulties were apparent based on parent functional report of day-to-day behavior, difficulties compared to the normative mean were also apparent on performance-based working memory, inhibition, and cognitive flexibility/shifting tasks. Twenty-two percent of children with NF1 demonstrated difficulty based on a performance-based working memory task; 21.6% demonstrated difficulty on a cognitive flexibility/shifting task; and 37.5% to 48.6% demonstrated difficulty on performance-based inhibition tasks. It is evident that our sample of children with NF1 demonstrated particularly high rates of inhibitory control difficulties, with nearly 50% of children demonstrating such difficulties. This finding suggests that the NIH Toolbox Flanker task is an especially sensitive measure in identifying performance-based EF difficulties. This finding is consistent with previous studies that have found that children with NF1 make significantly more inhibitory errors compared to control groups of typically developing children (Gilboa et al., 2011; Isenberg et al., 2013). Our results also mirror previous studies that have found greater incidence of impulsivity on Go/No-Go tasks compared to typically developing children (Ribeiro et al., 2015). Furthermore, findings from functional MRI (fMRI) studies in children with NF1 have revealed disturbances within neural networks

associated with working memory (Shilyansky et al., 2010) and inhibition (Pride et al., 2017), suggesting greater incidence of working memory and inhibition deficits in children with NF1.

Correspondence between performance and functional measures. In line with findings from several previous investigations (Toplak et al., 2009; Gioia et al., 2002, Bodnar et al., 2007), the current study found only minimal relations between scores on performance-based EF measures and parent reported, functional EF. Findings indicated that only parent report of functional self-monitoring was associated with performance-based cognitive flexibility/shifting on a standardized task, such that children with NF1 who demonstrated more difficulty on the performance-based cognitive flexibility/shifting measure were rated by their parents as experiencing more day-to-day functional self-monitoring difficulties. No additional relations between performance-based and functional EF were present in the current study. This finding lends further support to the suggestion that performance-based and parent-reported functional measures assess different underlying constructs of EF. Furthermore, the vastly different settings in which EF is assessed between performance-based and functional EF is noteworthy. Highly structured settings may allow for individuals to perform better than they would typically on a day-to-day basis in their typical environment, thus limiting the extent to which difficulties are observed (Holmes-Bernstein & Waber, 1997; Salthouse et al., 2003). As hypothesized and consistent with previous research, the present study found significantly higher rates of inhibition, working memory, and cognitive flexibility/shifting impairment on our functional, parent-reported measure in direct comparison to rates on performance -based measures. This is in line with the notion that performance-based EF tasks were designed to detect EF impairments in the low ability level and may not be helpful in differentiating among children's performances across

a full range of ability. Furthermore, the optimal setting in which performance-based EF tasks are conducted are likely to limit the extent to which more subtle deficits may exist in a functional capacity on a day-to-day basis.

Contributions to Performance and Functional EF

Attention: As expected and given the overlap in executive dysfunction and ADHD, the present study found that both dimensional and categorical ADHD symptomatology were related to EF. When examined dimensionally, both parent-reported inattentive and hyperactive/impulsive symptoms were related to parent report of functional difficulties with working memory, self-monitoring, inhibition, and planning/organization. Dimensional inattentive symptoms were also related to parent report of functional parent report of difficulties with initiation. In addition to functional EF, dimensional ADHD inattentive symptoms were also related to a performance-based inhibition task. Despite the strong theoretical overlap, the present study did not find any relations to dimensional ADHD hyperactive/impulsive symptoms and performance-based EF. This may be due to the optimal nature of the highly structured environment in which performance-based tasks are completed, with clear instructions and specific goals, thus reducing the opportunity to observe more typical behavior.

Thirty-seven percent of children with NF1 in the current study met diagnostic criteria for ADHD, which is consistent with previously reported frequencies of ADHD in NF1 (Hyman et al., 2005; Koth, Cutting, & Denckla, 2000). Group differences between children with NF1 + ADHD and children with NF1 without ADHD were evident when examining parent report of functional inhibition, emotional control, working memory, planning/organization, and overall behavioral regulation, suggesting that children with comorbid ADHD experience more day-to-

day, functional difficulties in these areas compared to children without ADHD. This finding is consistent with the very nature of a diagnosis of ADHD, which requires report of functional impairment across environments for diagnosis.

In the present study, there were no significant group differences in EF scores on performance-based tasks between children with NF1 + ADHD and children with NF1 without ADHD. These findings are in line with previous investigations that have demonstrated that children with NF1 with comorbid ADHD generally do not perform worse than children with NF1 without ADHD (Hyman et al., 2005; Roy et al., 2010). Again, given the highly structured environment in which performance-based tasks are completed, it may be that subtle difficulties, which are more apparent day-to-day, are not observable and limiting in an optimal setting.

General Cognitive Functioning: In the current study, overall general cognitive functioning was associated with performance on all three performance-based working memory tasks, as well as a performance-based inhibition task. No additional relations to any performance-based EF task or parent reported, functional EF were evident. In general, previous literature has been inconsistent as to the relations between IQ and EF. In both the adult and pediatric populations, there is evidence that some EF processes tend to be more related to overall cognitive functioning/IQ compared to others. Working memory, in particular, tends to be one of the EF processes that has had the most robust associations with overall IQ in both the adult and pediatric populations. Friedman and colleagues (2016) suggested that the processes involving in working memory, including sustaining attention to process relevant information, ignoring irrelevant information, and updating and reworking information, corresponds to previous definitions of intelligence, and thus explain why these two constructs are so strongly related. Still, other studies have found no correlation between IQ and several performance-based EF

tasks in school-aged children (Welsh, Pennington, Grossier, 1991). Other researchers have highlighted the moderating effect of IQ on EF, particularly at higher IQ levels (Baron, 2003; Mahone et al., 2002). The lack of relations between overall cognitive functioning and parent reported, functional EF may be explained by the differing nature of performance-based and functional EF measures. Performance-based measures are cognitive measures, whereas functional EF measures involve descriptions and observations of behaviors. In addition, in the present study, overall cognitive functioning was obtained on the same day performance-based EF was obtained. In contrast, parent-reported functional EF is considered aggregate information taking into account behavior over the past 6 months. With this in mind, it seems plausible that performance-based EF tasks would be more likely to be associated with overall cognitive functioning, compared to functional EF.

SES & Maternal Education: The present study found no relations between any EF measure, performance-based of functional, to SES or maternal education. To date there are no studies that examine contributions of SES or maternal education to EF in children with NF1; however, this finding is in contrast to the typically developing literature which has consistently demonstrated positive relations between SES and EF and has found SES to be predictive of EF performance (Lawson, Hook, & Farah, 2018). While much of the SES and EF literature has focused on emerging EF in preschool and young ages, findings from studies of EF development during the early to middle childhood years have suggested that SES disparity in EF remains consistent across these ages (Hackman et al., 2015). There a few potential explanations for the lack of relations between SES and EF in the current study. Studies that have examined relations between SES and cognitive and neuropsychological functioning in children with learning disabilities have found differing relations depending on the aspect of functioning examined

(Morrison & Hinshaw, 1988). Specifically, while SES was significantly related to intelligence and achievement scores, SES was not related to scores on measures assessing speed naming, visual and spatial sequencing, and visual motor integration, suggesting that socioeconomic factors are not able to account for the variability in neuropsychological performance. The finding that SES is not related to EF performance and ratings in our sample of children with NF1 may be indicative of the extent to which executive dysfunction impacts school-aged children with NF1 as a group, independent of SES. With this in mind, the potential for a lack of positive relations seems possible for children with NF1. It is also plausible that the present study did not have a broad enough SES range to pick up on relations between SES and EF and, paired with our relatively small sample size, may have hindered our ability to identify significant results.

Depression & Anxiety: Contrary to our hypothesis, parent report of overall internalizing problems, and individually, depression and anxiety symptomatology, were not related to performance-based EF in the current study. Previous investigations of typically developing children have found high rates of internalizing problems in children and adolescents with EF impairments and ADHD (see Craske, 2012 for review). Previous studies of children and adolescents with depression have found associated inhibition and working memory impairments using performance-based EF measures (Brooks et al., 2010; Maalouf et al., 2011). It has also been demonstrated that children and adolescents with anxiety perform worse on performance-based working memory tasks and on cognitive flexibility measures (Toren et al., 2000; Emerson, Mollet & Harrison, 2005). Furthermore, relations between poorer working memory abilities and anxiety have been found in samples of participants without clinical levels of anxiety (Ursache and Raver, 2014), suggesting that even subtle, more task-dependent anxiety impacts EF. Despite the lack of significant findings in relation to performance-based EF, results from the current

study revealed that parent report of internalizing symptoms, and individually, depression, and anxiety symptomatology were significantly related to parent report of affective aspects of EF on scales sampling emotional control, shifting/cognitive flexibility, and overall behavioral regulation. Depression was also significantly associated with EF related to behavioral regulation on the parent-reported, functional measure. Our findings mirror results from neuroimaging and brain injury research that has demonstrated overlapping brain structures and networks to be implicated in the regulation of behavior and in the regulation of emotion (Zelazo & Cunningham, 2007). Given the concurrent nature of the present study, it is not possible to determine whether EF difficulties in children with NF1 increase risk for internalizing problems, or if internalizing problems increase risk for EF difficulties. More likely, however, is that these difficulties are bidirectionally influenced, given the overlap in these processes and shared brain involvement in the prefrontal cortex. Regardless, given previous research suggesting increased risk for anxiety and depression in children and adolescents with NF1 (Johnson et al., 1999; Graf et al., 2006; Barton and North, 2007), and results from the current study that suggest associations with emotional control and self-regulation, children with NF1 may benefit from targeted prevention and intervention efforts aimed at supporting cognitive self-regulatory abilities (Riggs et al., 2006).

Sleep: As expected, the current study found that certain sleep-related difficulties, such as restlessness and snoring during sleep, as reported by parents, were significantly related to parent report of functional EF. While no previous studies have examined relations between specific sleep-related factors to EF in children with NF1, there is evidence to suggest increased hyperactivity and emotional challenges for children with NF1 who were reported to have high sleep disturbance (Johnson et al., 2005). Specifically, the present study found that children with NF1 who were rated by their parents as having difficulties on a daily basis related to emotional

control, self-monitoring, shifting/cognitive flexibility, inhibition, behavioral regulation, and overall EF were reported by their parents as struggling with restlessness during sleep. The present study also found that children with NF1 who were reported to have difficulty snoring during sleep were rated by their parents as having functional self-monitoring challenges on a daily basis. Snoring was also significantly related to speed of responses on a performance-based working memory task, such that children who were reported to struggle with snoring during sleep tended to demonstrate slower response times. This finding is consistent with research that has demonstrated that sleep disordered breathing involving snoring is associated with poor performance on cancellation tasks that require speed (Beebe, 2006). Despite this identified association to performance-based EF performance, the current study did not find any additional relations to restlessness, snoring, or difficulty falling asleep on any performance-based EF measures, suggesting that children with NF1 who may struggle with restless sleep are adequately able to inhibit impulses, monitor behavior, and think flexibly on tasks in a one-on-one environment under optimal conditions.

As hypothesized, and given previous findings by Licis and colleagues (2013) that revealed increased incidence of sleep difficulties in children and adolescents with NF1, the current study found that 35% of children with NF1 met cut-off criteria for a sleep-related breathing disorder (SRBD) based on parent-report of sleep and daytime behavior. SRBDs, characterized by abnormal respiration during sleep, include conditions such as obstructive sleep apnea, upper airway resistance syndrome, and obstructive hypopnea syndrome. It has clearly been demonstrated that SRBDs can have a profound impact on daytime behavior, particularly in the developing child. Group differences between children with NF1 who met criteria for SRBD and those that did not were apparent when examining parent report of functional inhibition,

working memory, and self-monitoring difficulties. The present study did not find differences in performance-based EF when comparing children with NF1 who met criteria for SRBD and those that did not, suggesting relatively limited impact of sleep related problems on EF performance under optional conditions in a one-on-one setting. Overall, our findings mirror results from several studies that have concluded there is evidence of worsened EF performance for children meeting criteria for a pediatric sleep disorder based on functional ratings, but less evidence to support poorer EF performance when using objective performance-based measures (see Mietchen et al., 2016 for review).

Implications

Overall, results from the present study confirm that EF difficulties are a characteristic feature of the NF1 cognitive phenotype, are not simply a consequence of comorbid ADHD, and interfere with the day-to-day functioning of children and adolescents with NF1. Results from the current study also demonstrate the need to consider EF difficulties in the context of co-occurring internalizing problems and sleep-related difficulties. While clinicians are likely to screen for the presence of depression or anxiety symptoms as part of a neuropsychological evaluation, they may be less likely to screen for sleep-related breathing difficulties, such as snoring or heavy daytime breathing. Our finding that children with NF1 who met criteria for a sleep-related breathing disorder experience more functional impairments in inhibition, working memory, and self-monitoring compared to children with NF1 without sleep-related breathing difficulties is noteworthy. Neurocognitive, behavioral, and emotional dysfunction, as well as reduced academic achievements are well-characterized comorbidities in children with sleep-related breathing disorders (Ali, Pitson, & Stradling, 1996; Gozal, 1998). Furthermore, research has demonstrated

that parent report of daytime sleepiness and hyperactivity can develop, though to a lesser extent, in children who habitually snore but do not have meet criteria for a sleep-related breathing disorder (O'Brien et al., 2004; Beebe, 2006; Montgomery-Downs et al., 2003). This finding has implications for a detailed screening of sleep-related difficulties for all pediatric populations, but particularly for those already at increased risk for EF dysfunction and sleep difficulties, such as children with NF1. Given the evidence that behavior and learning improve after effective treatment of sleep-related breathing disorders (Montgomery-Downs, Crabtree, & Gozal, 2005; Chervin et al., 2006), early screening and treatment of sleep difficulties will benefit multiple facets of functioning.

The results of the current study also highlight the importance of a comprehensive, multi-method assessment of EF for children with NF1. While assessment of EF is routine in neuropsychological evaluations, clinicians are likely to differ in their method of determining the integrity of EF skills. The present study is in line with previous research highlighting the need for EF to be considered a diverse and distinguishable, yet interconnected set of processes that underlie goal-directed behavior, rather than a single unitary construct. In addition to this crucial consideration, it is also important that clinicians be explicit about their interpretation of various EF processes, and steer away from generalizing deficits with one EF process to another. Incorrect categorization or descriptions of a child's EF performance may lead to an inaccurate depiction of a child's neuropsychological profile and may hinder access to and/or responses to appropriate interventions.

Limitations and Future Directions

While the present study provides clinically relevant information about the pattern of EF across performance-based and functional parent-reported EF measures, and identifies contributing factors to executive dysfunction in children with NF1, there are limitations in the study design that will be useful to address in future research. First, this study utilized a relatively small sample size that may have limited our ability to identify significant results. Future studies employing larger sample sizes would improve power and generalizability of results. Second, this study used published normative data as a comparison when examining EF across performance-based and parent-reported functional measures. Future research that utilizes a control group of unaffected children and a comparison group of children with ADHD (without NF1) from the community would help control for this limitation. Third, an additional limitation is that it utilized the BRIEF, in which there is currently an updated version available for administration (BRIEF-2; Gioia et al., 2015). While no new items were added to the clinical scales in the recent version of the measure, the BRIEF-2 has fewer items, a new index (Emotion Regulation Index), and re-arrangement of scale content occurred resulting in the creation of two scales (Self-Monitor and Task-Monitor) out of the BRIEF Monitor scale items. However, it is expected that the results of the current investigation translate to use with these updated measures as correlations between the parent-reported BRIEF and BRIEF-2 have been found to be generally high, with most coefficients greater than .80 (BRIEF-2; Gioia et al., 2015). However, as expected given the changes made between editions, the original BRIEF Monitor scale and the BRIEF-2 Task Monitor scale had lower coefficients (r = .69). Future projects utilizing parent-reported BRIEF data may benefit from re-scoring the data using BRIEF-2 scoring. Fourth, although it has clearly been elucidated that EF cannot be understood as constituting a single entity, and rather must be

considered as several different processes, the present study did not comprehensively examine all EF constructs. The most notable absence from our performance-based EF measures was a task assessing planning, problem-solving, and organization. Ideally, future research should include a comprehensive research battery that is exhaustive in its multi-method efforts for EF tasks that includes a task of planning, problem-solving, and organization.

Finally, while the current study utilized a multi-method approach using performance-based and functional EF measures to simultaneously examine EF, for psychosocial functioning and sleep-related difficulties, our investigation relied solely on parent report. Relying exclusively on parent report may introduce response bias for these constructs and does not provide insight into the child's own perception of these areas of functioning. It is important to note that method variance may play a role in the observed relations between functional EF and other parent-reported measures. For psychosocial functioning in particular, prior research has suggested that parent-child agreement in reporting of behavior is quite low, with correlations as low as 0.25 for some behaviors (Achenbach, McConaughy, & Howell, 1987). Low agreement has been found to be particularly evident when rating behaviors that are not observable, such as internalizing problems (March et al., 1997; Rey, Schrader, & Morris-Yates, 1992). Future research examining internalizing problems in school-aged children with NF1 would benefit from inclusion of a self-report measure of depression and anxiety. The current study also relied on parent report for assessing sleep characteristics. While the PSQ has been validated against polysomnography, it has been found that sleep diary estimates, actigraphy estimates, and total sleep times differed substantially in a study examining sleep in adolescents (Short et al., 2012). Future studies in which actigraphy measurements are collected in conjunction with parent reported data will enable a more comprehensive assessment of the presence of sleep-related difficulties in children

with NF1, and its contribution to overall functioning. In addition, while we examined EF from both a performance-based and functional parent-reported perspective, future studies would benefit from including teacher report when sampling EF difficulties in children with NF1. In some ways, teachers may be more reliable reporters of day-to-day functional EF due to the higher demand for EF skills in the school setting rather than home. This may be especially true for younger, preschool-aged children. Furthermore, given the very nature of their work, teachers may also be more reliable reporters compared to parents given they often have a better sense of age-typical behavior. Future research utilizing a multi-informant approach that includes teacher report would be useful in characterizing EF difficulties in different contexts and environments.

Table 1. *Evidence for executive functioning impairments in children with NF1*

Area of Impairment	Supporting Studies using Performance-based Measures	Supporting Studies using Functional Measures	Supporting Studies using Performance-based and Functional Measures
Inhibition	Ferner et al. (1996) Rowbotham et al. (2009) Huijbregts et al. (2010) Gilboa et al. (2011) Isenberg et al. (2013) Pride et al. (2017) Lion-Francois et al. (2017) Mazzocco et al. (1995) Casnar & Klein-Tasman (2017)	Payne et al. (2011)	Mautner et al. (2002) Plasschaert et al. (2015)
Working Memory	Huijbregts et al. (2010) Rowbotham et al. (2009) Ferner et al. (1996) Payne et al. (2012) Champion et al. (2014)	Payne et al. (2011) Sangster et al. (2011)	Ulrich et al. (2010) Gilboa et al. (2014) Casnar & Klein-Tasman (2016) Plasschaert et al. (2015)
Cognitive Flexibility/Shifting	Hofman et al. (1994) Descheemaeker et al. (2005) Rowbotham et al. (2009) Roy et al. (2014) Lion-Francois et al. (2017) Casnar & Klein-Tasman (2016)	Payne et al. (2011) Pride et al. (2010)	Plasschaert et al. (2015)
Planning/Organization	Hofman et al. (1994) Hyman et al. (2005) Pride et al. (2010) Roy et al. (2010) Galasso et al. (2014)	Payne et al. (2011)	Gilboa et al. (2014) Plasschaert et al. (2015)

Table 2. *Summary of measures for current study*

Construct	Measure	Measure Type
Executive Functioning		
Working Memory	NIH Toolbox List Sorting Working Memory (Toolbox LSWM)	Performance-based; Dimensional
Cognitive Flexibility/Shifting	NIH Toolbox Dimensional Change Card Sort (Toolbox DCCS)	Performance-based; Dimensional
Inhibition	NIH Toolbox Flanker Inhibitory Control and Attention (Toolbox Flanker)	Performance-based; Dimensional
Working Memory	Cogstate One Back Test (Cogstate OBT)	Performance-based; Dimensional
Inhibition	NEPSY-II Response Set (NEPSY-II RS)	Performance-based; Dimensional
Working Memory	DAS-II Recall of Sequential Order; Recall of Digits Backward (RSO; RDB)	Performance-based; Dimensional
Inhibition, Working Memory, Cognitive Flexibility/Shifting, Planning/Organization	Behavior Rating Inventory of Executive Function (BRIEF)- Parent	Parent-reported; Dimensional
ADHD Symptomaology		
Visual Attention	Cogstate Identification (Cogstate ID)	Performance-based; Dimensional
Visual Attention	NIH Toolbox Flanker Inhibitory Control and Attention (Toolbox Flanker)	Performance-based; Dimensional
Auditory Attention	NEPSY-II Auditory Attention/Response Set (NEPSY AA)	Performance-based; Dimensional
ADHD symptomatology	Kiddie Schedule for Affective Disorders and Schizophrenia (KSADS)-ADHD Section	Parent-reported; Examiner-rated; Categorical
ADHD symptomatology	Conners Short Form (Conners)- Parent	Parent-reported; Dimensional
SES		
Total Parental SES	Hollingshead Four-Factor Index of Socioeconomic Status (Hollingshead)	Parent-reported; Examiner-rated; Dimensional
Anxiety		
Anxiety symptomatology	Spence Children's Anxiety Scale (SCAS)	Parent-reported; Dimensional
Depression		
Depression symptomatology	Behavior Assessment System for Children, Second Edition (BASC-2)	Parent-reported; Dimensional
Sleep		
Sleep-Related Breathing Disorder	Pediatric Sleep Questionnaire (PSQ SRBD)	Parent-reported; Categorical
Sleep-related problems	Pediatric Sleep Questionnaire (PSQ)	Parent-reported; Categorical
Overall Cognitive Functioning		
General Conceptual Ability (GCA)	Differential Ability Scales, Second Edition (DAS-II GCA)	Performance-based; Dimensional

Table 3. *Participant Demographic Data* (n = 40)

Variable	**Score/Percent**
Mean Age (*SD*)	10.91 (1.57)
Sex (%)	
Males	22 (55)
Females	18 (45)
Ethnicity (%)	
Caucasian	33 (83)
African American	4 (10)
Asian	1 (2)
Biracial	2 (5)
Mean SES Index (*SD*)	41.22 (12.95)
Maternal Education (%)	
HS Diploma	7 (17)
Some College	11(28)
College Degree	8 (20)
Grad/Prof College	14 (35)
Mean GCA (*SD*)	93.90 (13.24)
Current Grade (*SD*)	4.93 (1.64)
Special Education Services (%)	
Yes	21 (53)
No	19 (47)
Comorbid Diagnoses (%)	
None	21 (52.5)
ADHD	15 (37.5)
Inattentive type	8 (20)
Hyperactive/Impulsive type	3 (7.5)
Combined type	4 (10)
Generalized Anxiety Disorder	1 (2.5)
MD, Language Disorder	1 (2.5)
RD, Language Disorder	1 (2.5)
MD, RD, Language Disorder	1 (2.5)

NF1 Diagnosis (%)	
Sporadic	27 (68)
Familial	13 (32)

Table 4: *BRIEF Parent Descriptive Statistics and Differences from Normative Mean (One-sample t-tests)*

Domain/Index	N	M (SD)	*t*	*p*	≥ 1 SD above mean
Inhibit	40	56.70 (10.76)	3.94	<.001	13 / 40 32.5%)
Shift	40	53.33 (13.48)	1.56	.127	10 / 40 (25%)
Emotional Control	40	52.75 (10.36)	1.68	.101	10 / 40 (25%)
BRI	40	55.15 (10.45)	3.12	.003	12 / 40 (30%)
Initiate	40	57.08 (9.15)	4.89	<.001	15 / 40 (37.5%)
Working Memory	40	58.98 (10.43)	5.44	<.001	15 / 40 (37.5%)
Plan/Organize	40	57.18 (9.98)	4.54	<.001	17 / 40 (42.5%)
Organization of Materials	40	58.23 (9.66)	5.38	<.001	20 / 40 (50%)
Monitor	40	59.55 (10.74)	5.62	<.001	18 / 40 (45%)
MI	40	59.10 (9.58)	6.00	<.001	18 / 40 (45%)
GEC	40	59.13 (10.58)	5.45	<.001	18 / 40 (45%)

Note: Higher BRIEF mean scores reflect more difficulty

BRI= Behavioral Regulation Index; MI=Metacognition Index, GEC= Global Executive Composite

Table 5. EF/Attention Performance Descriptive Statistics and Differences from Normative Mean (One-sample t-tests)

Measure/Subtest	N	M (SD)	*t*	*p*	≥ 1 SD below mean
DAS-II RSO	40	45.88 (7.25)	-3.59	.001	9 / 40 (22.5%)
DAS-II RDB	40	43.25 (8.23)	-5.18	<.001	9 / 40 (22.5)
Toolbox DCCS	37	94.14 (12.05)	-2.96	.005	8 / 37 (21.6%)
Toolbox LSWM	37	98.51 (14.04)	-0.64	.524	7 / 37 (18.9%)
Toolbox Flanker	36	87.99 (13.11)	-5.49	<.001	18 / 37 (48.6%)
Cogstate Identification	39	-.07 (1.02)	-.431	.669	11 / 39 (28.2%)
Cogstate OBT Speed	39	.47(.97)	3.01	.005	3 / 39 (7%)
Cogstate OBT Accuracy	39	.05 (1.00)	.315	.755	4 / 39 (10.2%)
NEPSY-II AA	40	9.13 (3.38)	-1.64	.110	12 / 40 (30%)
NEPSY-II RS	40	8.70 (3.47)	-2.37	.023	15 / 40 (37.5%)

Table 6. *Relations between Performance EF/Attention Measures and Functional EF*

	BRIEF Inhibit	BRIEF Shift	BRIEF EC	BRIEF BRI	BRIEF Initiate	BRIEF WM	BRIEF Plan/Org	BRIEF Org/Mat	BRIEF Monitor	BRIEF MI	BRIEF GEC
DAS-II RSO	-.037	-.007	-.245	-.110	-.086	-.151	-.154	.013	-.184	-.211	-.074
DAS-II DB	-.102	.109	.094	-.012	.144	-.047	-.004	.138	-.065	-.017	.048
Toolbox DCCS	-.354	-.198	-.097	-.290	-.328	-.325	-.162	-.226	**-.519***	-.243	-.286
Toolbox LSWM	.002	-.159	-.125	-.108	-.131	-.149	-.157	-.159	-.236	-.235	-.120
Toolbox Flanker	.093	-.037	.137	.097	.069	.028	.136	.066	.071	.063	.025
Cogstate Identification	.060	.132	.079	.157	-.066	.053	.000	-.012	-.027	.000	.102
Cogstate OBT Speed	-.229	-.120	-.222	-.176	-.176	-.140	-.043	.024	-.238	-.050	-.057
Cogstate OBT Accuracy	.016	-.039	.020	-.075	-.236	-.296	-.264	-.250	-.226	-.220	-.171
NEPSY-II AA	-.008	.190	.022	.075	.111	-.126	.012	-.085	-.020	-.104	-.032
NEPSY-II RS	-.398	.004	-.193	-.290	-.278	-.319	-.092	-.025	-.391	-.235	-.295

Note: * $p < .05$; ** $p < .01$; significant difference determined based on q-value (FDR derived significance threshold)

EC= Emotional Control; BRI= Behavioral Regulation Index; WM= Working Memory; Plan/Org=Plan/Organize; OrgMat=Organization of Materials; MI=Metacognition Index, GEC= Global Executive Composite

Table 7. *Descriptive Statistics and Group Differences on Performance EF/Attention Measures*

	NF1			NF1 + ADHD				
Measure	*N*	*Mean*	*(SD)*	*N*	*Mean*	*(SD)*	*F*	*q-value*	*D*
DAS-II RSO	25	47.12	6.73	15	43.80	7.83	1.81	.689	.46
DAS-II DB	25	43.68	8.32	15	42.53	8.32	0.29	.746	.18
Toolbox DCCS	23	95.96	12.92	14	91.14	10.24	0.96	.739	.34
Toolbox LSWM	23	101.04	12.90	14	94.36	15.31	1.94	.689	.47
Toolbox Flanker	22	88.90	14.07	14	86.57	11.81	0.26	.746	.17
Costate ID	25	-0.13	1.13	14	0.03	0.81	0.0	.991	.00
Cogstate OBT Speed	25	0.66	1.04	14	0.13	0.74	3.05	.677	.59
Cogstate OBT Accuracy	25	0.04	0.88	14	0.07	1.21	0.06	.888	.08
NEPSY-II AA	25	9.32	2.96	15	8.80	4.07	0.45	.739	.23
NEPSY-II RS	25	9.20	3.23	15	7.87	3.82	0.62	.734	.27

Table 8. *BRIEF Descriptive Statistics and Group Differences based on ADHD*

	NF1			NF1 + ADHD					
Measure	*N*	*Mean*	*(SD)*	*N*	*Mean*	*(SD)*	*F*	*q-value*	*D*
Inhibit	25	51.28	6.68	15	65.73	10.29	29.12	.000	1.76
Shift	25	50.24	12.96	15	58.47	13.16	3.73	.067	.63
Emotional Control	25	49.60	8.69	15	58.00	11.03	7.13	.015	.87
BRI	25	50.64	7.79	15	62.67	10.17	17.74	.000	1.37
Initiate	25	54.88	8.95	15	60.73	8.53	4.15	.058	.66
Working Memory	25	55.16	8.43	15	65.33	10.57	11.26	.003	1.09
Plan/Organize	25	53.00	8.74	15	64.13	7.99	16.17	.000	1.31
Organization of Materials	25	56.44	10.54	15	61.20	7.38	2.35	.133	.50
Monitor	25	54.08	7.60	15	68.67	8.94	30.23	.000	1.79
MI	25	54.76	7.90	15	66.33	7.69	20.48	.000	1.48
GEC	25	54.96	9.58	15	66.07	8.49	13.66	.002	1.21

Note: Higher BRIEF mean scores reflect more difficulty

BRI= Behavioral Regulation Index; MI=Metacognition Index, GEC= Global Executive Composite

Table 9. *BRIEF Descriptive Statistics and Group Differences for based on Sleep-related Breathing Disorder*

	NF1			NF1 + SRBD					
Measure	*N*	*Mean*	*(SD)*	*N*	*Mean*	*(SD)*	*F*	*q-value*	*D*
Inhibit	26	53.28	8.46	14	63.14	11.85	9.38	.014	1.0
Shift	26	52.85	14.90	14	54.21	10.82	.091	.764	.10
Emotional Control	26	51.73	10.45	14	54.64	10.29	.714	.443	.28
BRI	26	53.19	10.16	14	58.79	10.35	2.72	.166	.55
Initiate	26	55.42	8.74	14	60.14	9.40	2.52	.166	.53
Working Memory	26	56.08	9.42	14	64.36	10.37	6.55	.015	.85
Plan/Organize	26	54.69	9.05	14	61.79	10.32	5.06	.066	.75
Organization of Materials	26	57.15	10.24	14	60.21	8.47	.911	.422	.32
Monitor	26	55.42	8.50	14	67.21	10.48	14.85	.000	1.3
MI	26	56.00	8.51	14	64.86	9.04	9.45	.014	1.0
GEC	26	56.69	10.44	14	63.64	9.65	4.24	.084	.68

Note: Higher BRIEF mean scores reflect more difficulty

BRI= Behavioral Regulation Index; MI=Metacognition Index, GEC= Global Executive Composite

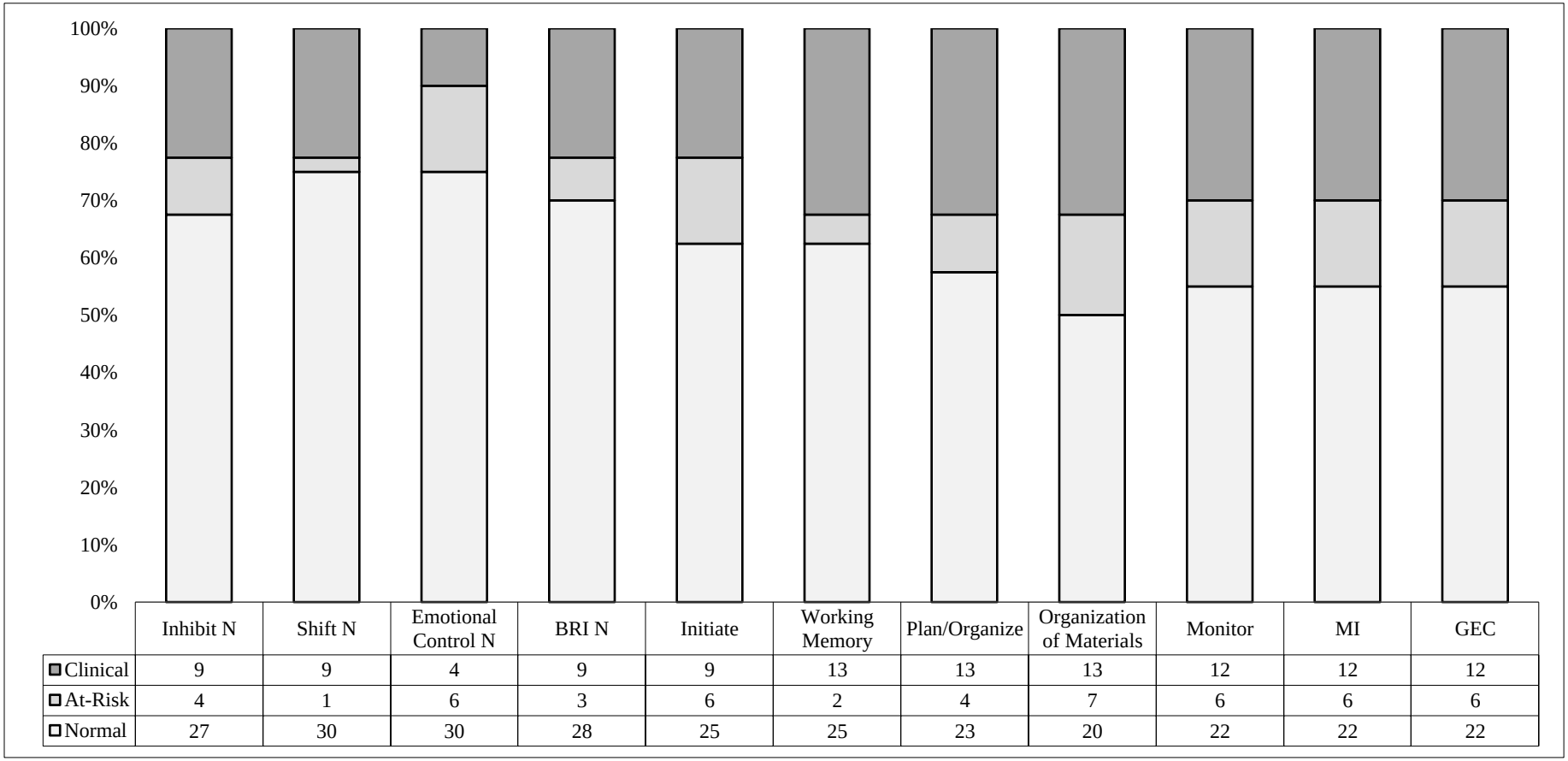
Table 10. *Regression Analyses: Contributions to Functional and Performance EF*

Functional Parent-Reported EF (BRIEF GEC)	*F*	*df*	*p*	R^2	*β*
Model	5.51	6, 33	.001	.500	
DAS-II GCA			.805		.031
SES			.536		.078
ADHD Diagnosis			.034		.312
SRBD Criteria			.067		.273
SCAS Total Anxiety			.284		.153
BASC-2 Depression scale			.009		.408

Performance-Based Inhibition (NIH Toolbox Flanker)	*F*	*df*	*p*	R^2	*β*
Model	1.34	6, 29	.273	.217	
DAS-II GCA			.023		.401
SES			.328		.166
ADHD Diagnosis			.701		-.073
SRBD Criteria			.729		-.067
SCAS Total Anxiety			.788		.051
BASC-2 Depression scale			.482		-.139

GEC= Global Executive Composite; GCA = General Conceptual Ability

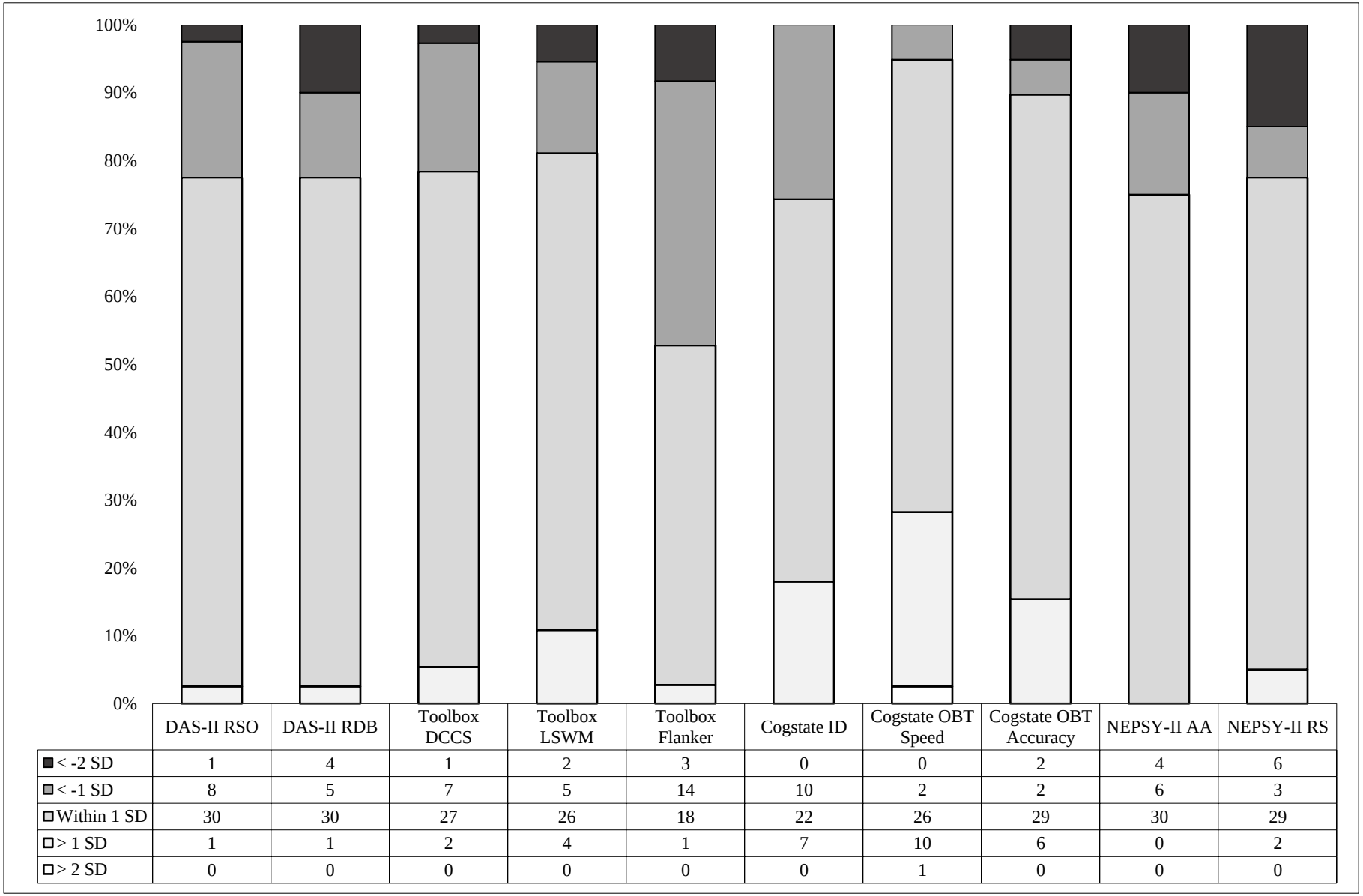
Figure 1. *Parent-reported level of difficulties on the BRIEF*



	Inhibit N	Shift N	Emotional Control N	BRI N	Initiate	Working Memory	Plan/Organize	Organization of Materials	Monitor	MI	GEC
Clinical	9	9	4	9	9	13	13	13	12	12	12
At-Risk	4	1	6	3	6	2	4	7	6	6	6
Normal	27	30	30	28	25	25	23	20	22	22	22

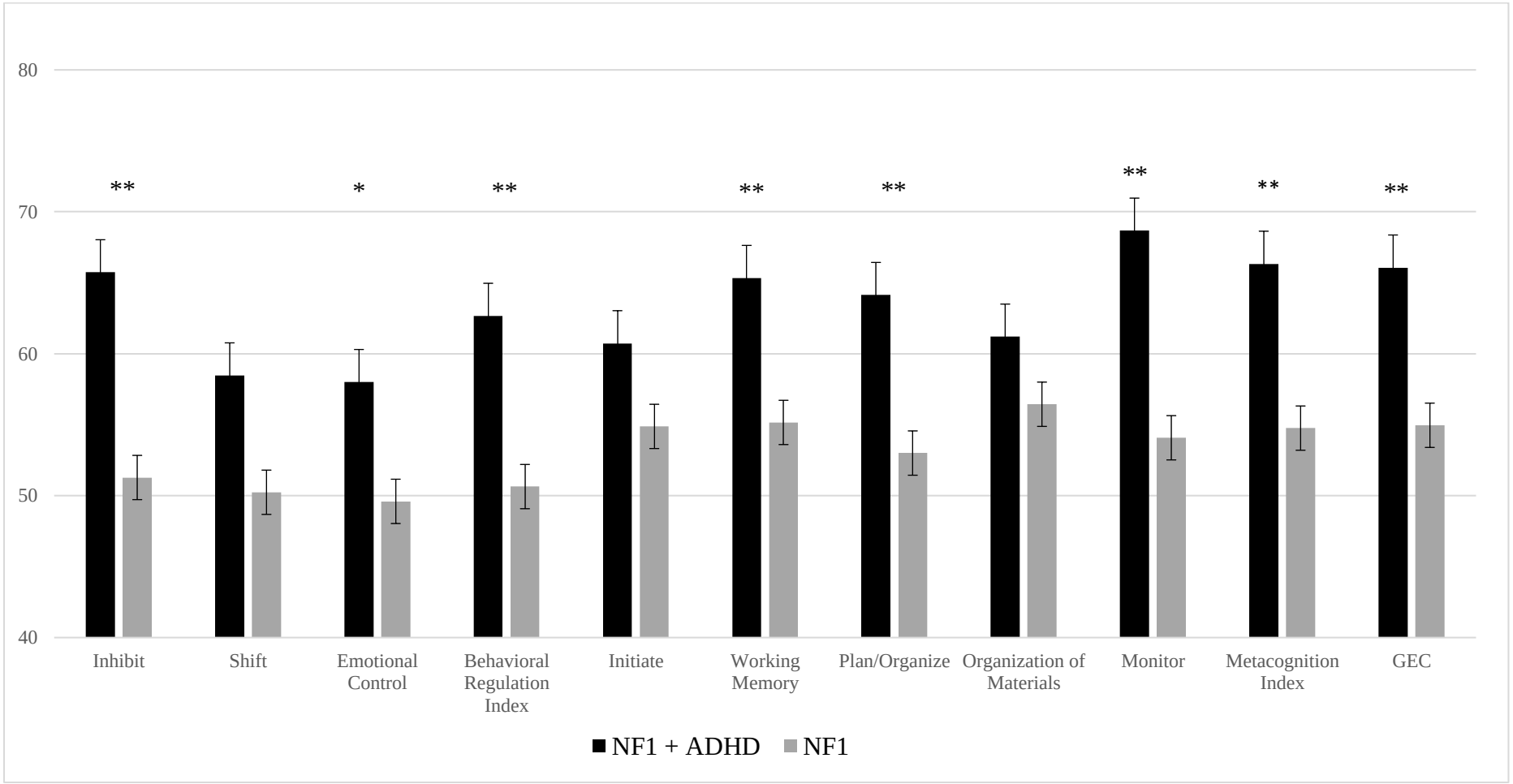
BRI = Behavioral Regulation Index; MI= Metacognition Index; GEC = Global Executive Composite.

Figure 2. *Level of Difficulties on Performance-based EF Measures*



	DAS-II RSO	DAS-II RDB	Toolbox DCCS	Toolbox LSWM	Toolbox Flanker	Cogstate ID	Cogstate OBT Speed	Cogstate OBT Accuracy	NEPSY-II AA	NEPSY-II RS
< -2 SD	1	4	1	2	3	0	0	2	4	6
< -1 SD	8	5	7	5	14	10	2	2	6	3
Within 1 SD	30	30	27	26	18	22	26	29	30	29
> 1 SD	1	1	2	4	1	7	10	6	0	2
> 2 SD	0	0	0	0	0	0	1	0	0	0

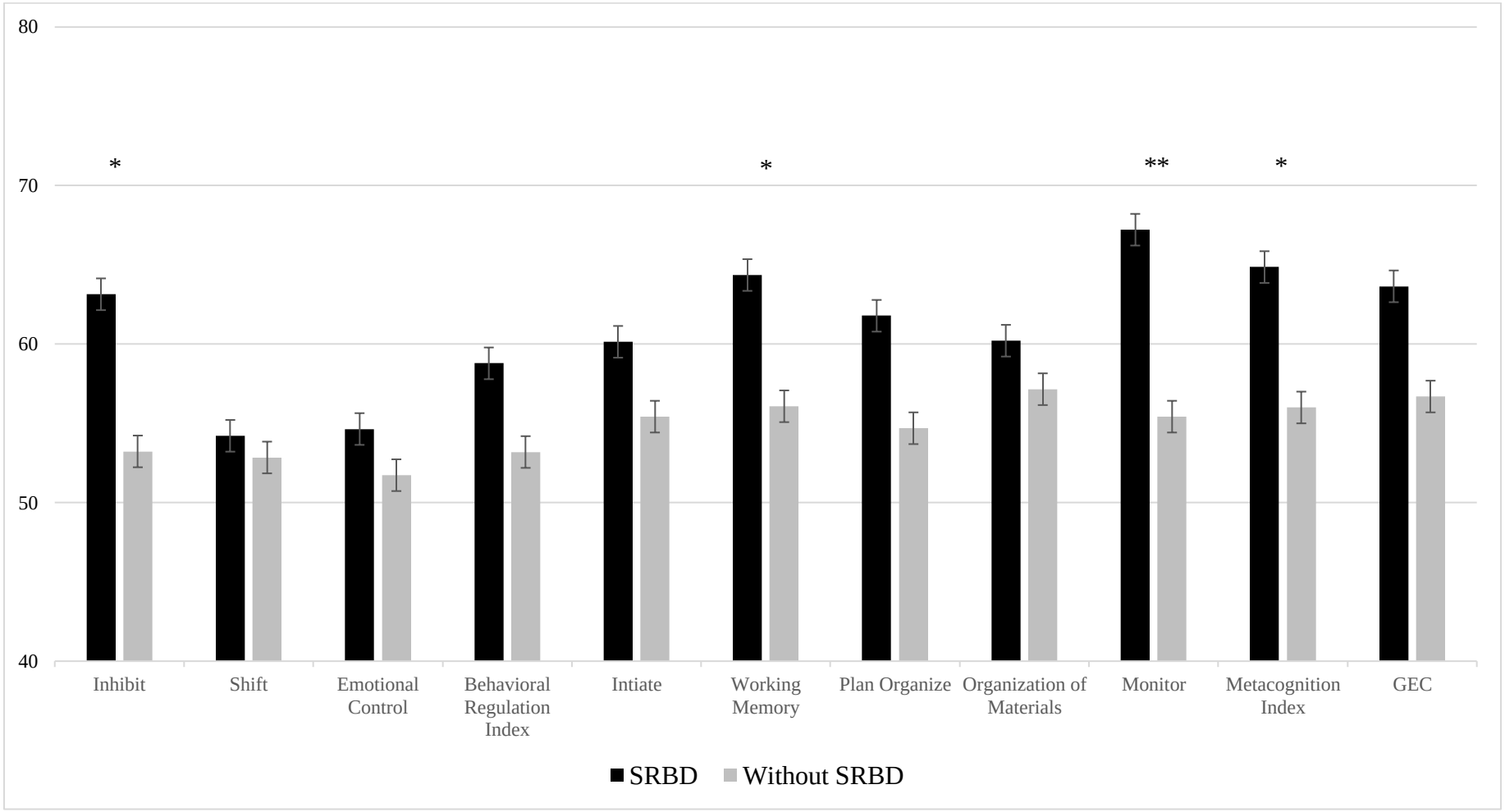
Figure 3. *Group Differences in Parent-Reported EF for children with NF1 with and without ADHD*



Note: Higher BRIEF mean scores reflect more difficulty; * $p < .05$; ** $p < .01$; significant difference determined based on q-value (FDR derived significance threshold)

GEC = Global Executive Composite

Figure 4. *Group Differences in Parent-Reported EF for children with NF1 with and without Sleep-related Breathing Disorder*



Note: Higher BRIEF mean scores reflect more difficulty; * $p < .05$; ** $p < .01$; significant difference determined based on q-value (FDR derived significance threshold)

GEC = Global Executive Composite; SRBD = Sleep-related breathing disorder from the Pediatric Sleep Questionnaire

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Brianna D. Yund, Ph.D.
Curriculum Vitae

EDUCATION

2020 – Present	**Postdoctoral Fellowship**	Pediatric Neuropsychology | Division of Clinical Behavioral Neuroscience University of Minnesota | Minneapolis, MN
2017 – 2020	**Ph.D.**	Clinical Psychology (APA-Accredited) University of Wisconsin – Milwaukee | Milwaukee, WI Dissertation: Factors contributing to executive functioning in children with neurofibromatosis type 1
2019 – 2020	**Internship**	Pediatric Neuropsychology & Pediatric Psychology (APA-Accredited) University of Minnesota Medical School | Minneapolis, MN
2014 – 2017	**M.S.**	Clinical Psychology | University of Wisconsin – Milwaukee | Milwaukee, WI Thesis: Aggression in children with 7q11.23 duplication syndrome
2007 – 2010	**B.A.**	Psychology | University of Minnesota – Twin Cities | Minneapolis, MN

CLINICAL EXPERIENCE

POSTDOCTORAL FELLOWSHIP

2020 – Present **University of Minnesota Medical Center, Department of Pediatrics,** Minneapolis, MN

Pediatric Neuropsychology Postdoctoral Associate
Supervisors: Richard Ziegler, Ph.D., Kelly King, Ph.D., ABPP-CN, Elizabeth (Rene) Pierpont, Ph.D., Julie Eisengart, Ph.D., Alicia Kunin-Batson, Ph.D.

PREDOCTORAL INTERNSHIP

2019 – 2020 **University of Minnesota Medical Center, Department of Pediatrics,** Minneapolis, MN

Pediatric Neuropsychology Rotation
Pediatric Neuropsychology Intern
Supervisors: Margaret Semrud-Clikeman, Ph.D., ABPdN, Richard Ziegler, Ph.D., Kelly King, Ph.D., ABPP-CN, Elizabeth (Rene) Pierpont, Ph.D., Julie Eisengart, Ph.D., Alicia Kunin-Batson, Ph.D.

Pediatric Psychology Rotation
Pediatric Psychology Intern
Supervisors: Christopher Boys, Ph.D.; Amy Gross, Ph.D., BCBA-D

2017 – 2019 **Neuropsychology Advanced Practicum Student**
Medical College of Wisconsin, Milwaukee, WI
Department of Neurology, Division of Pediatric Neuropsychology
Supervisors: Amy Heffelfinger, Ph.D., ABPP-CN; Jennifer Koop, Ph.D., ABPP-CN.;

Michelle Loman, Ph.D.; Joseph Amaral, Ph.D.

2016 – 2019 **Group Therapy Facilitator**
Marquette University, Milwaukee, WI
Interdisciplinary Autism Clinic: PEERS®
Supervisor: Amy Van Hecke, Ph.D.

2016 – 2019 **Graduate Student Therapist**
University of Wisconsin – Milwaukee, Milwaukee, WI
UWM Psychology Clinic
Supervisor: Robyn Ridley, Ph.D.

2014 – 2019 **Neuropsychology Graduate Trainee**
University of Wisconsin – Milwaukee
Child Neuropsychology Clinic
Supervisor: Bonnie Klein-Tasman, Ph.D.**,** Kristin D. Smith, Ph.D.

2016 – 2017 **Graduate Student Trainee**
Children's Hospital of Wisconsin, Milwaukee, WI
Pediatric Gastroenterology, Outpatient Toileting Clinic
Supervisors: Alan Silverman, Ph.D.; W. Hobart Davies, Ph.D.

2015 – 2016 **Assessment Practicum Student**
University of Wisconsin – Milwaukee
UWM Psychology Clinic
Supervisors: Kristin D. Smith, Ph.D., Han Joo Lee, Ph.D.

2014 – 2015 **Practicum Student**
University of Wisconsin – Milwaukee
UWM Psychology Clinic
Supervisors: Kristin D. Smith, Ph.D.; Han Joo Lee, Ph.D.

2014 – 2016 **Graduate Student Trainee**
University of Wisconsin – Milwaukee
UWM Psychology Clinic
Child Anxiety Therapy Team, Supervisor: Bonnie Klein-Tasman, Ph.D.
Integrative Behavioral Couples Therapy Team, Supervisor: Christopher Martell, Ph.D.
Behavioral Activation Therapy Team, Supervisor: Christopher Martell, Ph.D.

RESEARCH EXPERIENCE

2017 – 2019 **Project Coordinator/Graduate Research Assistant**
University of Wisconsin – Milwaukee
Child Neurodevelopmental Research Lab
Treatment Development Study of Williams Syndrome Behavioral Play Therapy: WSA funded study for the development of a manual based on the Replays approach by Dr. Karen Levine for the treatment of strong emotional reactions in response to fears or stimuli in children with Williams syndrome ages 4-10.
Advisor: Bonnie Klein-Tasman, Ph.D.

2014 – 2019 **Graduate Research Assistant**
University of Wisconsin – Milwaukee
Child Neurodevelopment Research Lab
Advisor: Bonnie Klein-Tasman, Ph.D.

- **Children with 7q11.23 Duplication Syndrome: Shared Characteristics with ASD:** Examination of behavioral phenotype of children with 7q11.23 duplication syndrome, with focus on characterizing social functioning, areas of psychopathology risk, and autism spectrum symptomatology.
- **School-Aged Outcomes in NF1: Attention, Social, and Academic Functioning**: Follow-up study examining attention, social, and academic functioning in 9-13-year-olds with neurofibromatosis type 1, as well as characterize children and adolescents with NF1 regarding core diagnostic features, associated features, and predictors of ASD symptomatology.
- **Early Indicators of Emotional, Cognitive, and Learning Difficulties in Neurofibromatosis Type 1**: Longitudinal study aimed at characterizing the cognitive and behavioral phenotype of young children with neurofibromatosis type 1. Examination of risk factors for later learning and/or emotional problems that allow for early intervention.
- **Development of Children with Developmental Disabilities**: Examination of the social, emotional, and cognitive development of children with developmental delays and disabilities, including children with Williams syndrome.

2010 – 2014 **Psychometric Assistant/Data Coordinator**
University of Minnesota- Twin Cities
Department of Pediatrics, Division of Behavioral Neuroscience

- **Longitudinal Studies of Brain Structure and Function in MPS Disorders**: Five-year longitudinal study investigating the brain basis for cognitive difficulties in mucopolysaccharidosis (MPS) types I, II, and VI, and VII.
  Supervisor: Elsa Shapiro, Ph.D.

- **Brain Structure and Function in Developmentally Normal Children Ages 4-7 Years**: Longitudinal study examining brain structure and function in developmentally normal children utilizing neuropsychology battery and MRI.
  Supervisors: Margaret Semrud-Clikeman, Ph.D.; Elsa Shapiro, Ph.D.; Igor Nestrasil, M.D.

2009 – 2014 **Research Assistant**
University of Minnesota- Twin Cities
Department of Pediatrics, Division of Behavioral Neuroscience

- **Social-Emotional, Cognitive, and Neurologic Phenotype(s) of Children with Sanfilippo Syndrome**: Examination of the behavioral and learning difficulties in children with MPS IIIA (Sanfilippo syndrome). Comparison of behavioral profiles of children with MPS IIIA to those with MPS I to understand the disease processes underlying behavioral problems affecting children with MPS IIIA.
  Supervisors: Elsa Shapiro, Ph.D.; Michael Potegal, Ph.D.

- **Longitudinal Studies of Early Childhood Vigilance Task-Measured Attention in High Risk Uganda:** Examination of attention abilities in Ugandan children diagnosed with HIV and cerebral malaria. Evaluation of correlative and construct validity of the Early Childhood Vigilance Test (ECVT).
  Supervisors: Regilda Anne Romero, Ph.D., Elsa Shapiro, Ph.D.

2009 – 2010 **Research Assistant**
University of Minnesota- Twin Cities, Department of Pediatrics
Primary Care of Children with Autism: Physician Survey
Supervisors: Regilda Anne Romero, Ph.D., Allison Golnik, M.D., MPH

2009 – 2011 **Data Operator**
University of Minnesota- Twin Cities
Department of Public Health, Division of Environmental Health Sciences

Prostate, Lung, Colorectal and Ovarian Cancer Screening Trial
Supervisor: Timothy Church, Ph.D.

2008 – 2009 **Undergraduate Research Assistant**
University of Minnesota- Twin Cities
Department of Psychology
A Dyadic Analysis of Trust and Communication in Relationships
Supervisors: Jeffry Simpson, Ph.D. & Sandra Shallcross, Ph.D.

PEER-REVIEWED PUBLICATIONS

Glad, D.M., Casnar, C.L., **Yund, B.**, Enderle, M., Siegel, D., Basel, D., & Klein-Tasman, B.P. (2020). Adaptive behavior and executive functioning in children with neurofibromatosis type 1 using a mixed design. *Journal of Developmental & Behavioral Pediatrics*, DOI: 10.1097/DBP.0000000000000833.

Schiltz, H., McVey, A., Dolan, B., Willar, K., Pleiss, S., Carson, A., Caiozzo, C., Vogt, E., **Yund, B.**, & Vaughan Van Hecke, A. (2019). A psychometric analysis of the Social Anxiety Scale for adolescents among youth with autism spectrum disorder: caregiver-adolescent agreement, factor structure, and validity. *Assessment*, DOI: 10.1177/1073191119851563.

McVey, A. J., Schiltz, H. K., Haendel, A. D., Dolan, B. K., Willar, K. S., Pleiss, S., Karst, J. S., Carlson, M., Krueger, W., Murphy, C., Casnar, C. L., **Yund, B**., Van Hecke, A. (2018). Social difficulties in youth with autism with and without anxiety and ADHD symptoms. *Autism Research, 11*(12)*,* 1679-1689.

Schiltz, H.K., McVey, A.J., Dolan, B.K., Willar, K.S., Pleiss, S., Karst, J.S., Carlson, M., Krueger, W., Murphy, C., Casnar, C. L., **Yund, B**., Van Hecke, A. (2018). Changes in depressive symptoms among adolescents with ASD completing the PEERS® social skills intervention. *Journal of Autism and Developmental Disorders*, *48*(3), 834-843.

Shapiro, E.G., Rudser, K., Ahmed, A., Steiner, R.D., Delaney, K.A., **Yund, B**., King, K., Kunin Batson, A., Eisengart, J., & Whitley, C.B. (2016). A longitudinal study of emotional adjustment, quality of life and adaptive function in attenuated MPS II. *Molecular Genetics and Metabolism Reports*, *7*, 32-39.

Shapiro, E., King, K., Ahmed, A., Rudser, K., Rumsey, R., **Yund, B**., ... & Potegal, M. (2016). The neurobehavioral phenotype in mucopolysaccharidosis type IIIB: an exploratory study. *Molecular Genetics and Metabolism Reports*, *6*, 41-47.

Shapiro, E.G., Nestrasil, I., Rudser, K., Delaney, K., Kovac, V., Ahmed, A., **Yund, B**., Orchard, P., Eisengart, J ... & Raiman, J. (2015). Neurocognition across the spectrum of mucopolysaccharidosis type I: age, severity, and treatment. *Molecular Genetics and Metabolism*, *116*(1-2), 61-68.

Yund, B., Rudser, K., Ahmed, A., Kovac, V., Nestrasil, I., Raiman, J., ... & Shapiro, E.G. (2015). Cognitive, medical, and neuroimaging characteristics of attenuated mucopolysaccharidosis type II. *Molecular Genetics and Metabolism*, *114*(2), 170-177.

Ahmed, A., Whitley, C. B., Cooksley, R., Rudser, K., Cagle, S., Ali, N., Delaney, K., **Yund, B**., Shapiro, E. (2014). Neurocognitive and neuropsychiatric phenotypes associated with the mutation L238Q of the α-L-iduronidase gene in Hurler–Scheie syndrome. *Molecular Genetics and Metabolism*, *111*(2), 123-127.

Potegal, M., **Yund, B**., Rudser, K., Ahmed, A., Delaney, K., Nestrasil, I., ... & Shapiro, E.G. (2013). Mucopolysaccharidosis Type IIIA presents as a variant of Klüver–Bucy syndrome. *Journal of Clinical and Experimental Neuropsychology, 35(6),* 608-616.

Delaney, K. A., Rudser, K. R., **Yund, B.**., Whitley, C. B., Haslett, P. A., & Shapiro, E.G. (2013). Methods of neurodevelopmental assessment in children with neurodegenerative disease: Sanfilippo syndrome. In *Journal of Inherited Metabolic Disease Reports-Case and Research Reports, Volume 13* (pp. 129-137). Springer, Berlin, Heidelberg.

PRESENTATIONS AT INTERNATIONAL MEETINGS & CONFERENCES

Pardej, S.K., Lee, K., **Yund, B**., Casnar, C., & Klein-Tasman, B.P. (2020, April). *An investigation of the use of computerized attention/executive functioning measures with school age children with neurofibromatosis type 1.* Gatlinburg Conference (Conference cancelled).

Yund, B., Lee, K., Casnar, C., & Klein-Tasman, B.P. (2020, February). *Longitudinal Examination of Temperament and Executive Functioning in Children with Neurofibromatosis type 1.* Poster presented at the International Neuropsychological Society, Denver, CO.

Pardej, S. K., Glad, D. M., Casnar, C. L., **Yund, B**., & Klein-Tasman, B. P. (2020, February). *Parent report vs. lab based measures: Motor and attention functioning as predictors of working memory in children with neurofibromatosis type 1*. Poster presented at the International Neuropsychological Society, Denver, CO.

Lee, K., **Yund, B**., & Klein-Tasman, B.P. (2019, September). *Longitudinal Examination of Attention Problems and Anxiety in Children with Neurofibromatosis Type 1*. Platform presentation given at NF Conference; San Francisco, CA.

Glad, D.M., **Yund, B**., Lee, K., Casnar, C.L., & Klein-Tasman, B.P. (2019, September). *Longitudinal Examination of Social Skills of Children with Neurofibromatosis Type 1 Beginning in Early Childhood*. Poster presented at the NF Conference; San Francisco, CA.

Pardej, S. K., Glad, D. M., Casnar, C. L., **Yund, B.**, & Klein-Tasman, B. P. (2019, September). *Motor and attention functioning in early childhood as predictors of working memory in the school age years in children with NF1.* Poster presented at the NF Conference, San Francisco, CA.

A., Arias, A., Barrington, A. W., McVey, A. J., Schiltz, H. K. Haendel, A., **Yund, B**., Van Hecke, A. V., Kreugar, W., Carlson, M. (2019, May). *Changes in amygdala and hippocampus volumes among adolescents with autism spectrum disorder over the course of the PEERS® social skills intervention*. Oral presentation at the International Society for Autism Research (INSAR), Montreal, Québec.

Schwarz, G.N., Casnar, C., **Yund, B**., Lee, K., Brei, N., & Klein-Tasman, B. (2019, April). *Stability and Predictive Value of Intellectual Functioning in Neurofibromatosis type 1 beginning in the Preschool Years*. Poster presented at the Gatlinburg Conference, San Antonio, TX.

Yund, B., Lee, K., Casnar, C., & Klein-Tasman, B. (2019, February). *The Impact of Sleep Problems on Executive Functioning in School-Aged Children with NF1*. Poster presented at the International Neuropsychological Society, New York, NY.

Glad, D., **Yund, B**., Lee, K., Casnar, C., Ray, K., & Klein-Tasman, B. (2019, February). *Social Functioning in Children with Neurofibromatosis Type 1: A Longitudinal Investigation*. Poster presented at the International Neuropsychological Society, New York, NY.

Lee, K., **Yund, B**., Schwarz, G.N., Glad, D., & Klein-Tasman, B. (2019, February). *Longitudinal Examination of Problem Behaviors in Children with Neurofibromatosis type 1*. Poster presented at the International Neuropsychological Society, New York, NY.

Glad, D., **Yund, B**., Lee, K., Casnar, C., & Klein-Tasman, B. (2018, November). *Adaptive behavior in children with neurofibromatosis type 1: A longitudinal investigation*. Poster presented at the Joint Global Neurofibromatosis Conference, Paris, France.

Yund, B., Lee, K., Casnar, C., & Klein-Tasman, B. (2018, November). *Executive functioning in children with NF1 with and without ADHD using lab-based and parent and teacher reported functional measures*. Poster presented at the Joint Global Neurofibromatosis Conference, Paris, France.

Glad, D., **Yund, B**., Lee, K., Casnar, C., Enderle, M., & Klein-Tasman, B. (2018, October). *A longitudinal investigation of adaptive behavior in children with neurofibromatosis type 1*. Poster presented at the

National Conference in Clinical Child Psychology, Kansas City, KS.

McVey, A. J., Schiltz, H., Haendel, A., Pleiss, S., Carson, A., Rivera, K., **Yund, B**., Vogt, E., Van Hecke, A. V. (2018, May). *Examining the links between restricted interests/repetitive behaviors, respiratory sinus arrhythmia, and anxiety: Do repetitive behaviors mediate the relation between RSA and anxiety?* Poster presented at the 17th annual International Society for Autism Research (INSAR), Rotterdam, Netherlands.

Haendel, A. D., Barrington, A., Schiltz, H., McVey, A. J., Rivera K., Pleiss, S., Carson, A., **Yund, B**., Van Hecke, A.V. (2018, May). *Inter and Intra Hemispheric EEG Coherence in Adolescents with ASD Related to Social Function*. Poster presented at the 17th annual International Society for Autism Research (INSAR), Rotterdam, Netherlands.

Arias, A., McVey, A. J., Schiltz, H., Haendel, A., Rivera K., Pleiss, S., Carson, A., **Yund, B**., Van Hecke, A. V. (2018, May). *Adolescent autism traits: Associations with parental age and physiological functioning.* Poster presented at the 17th annual International Society for Autism Research (INSAR), Rotterdam, Netherlands, May 2018.

Yund, B., Lee, K., Casnar, C., & Klein-Tasman, B.P. (2018, February). *Executive Function Profiles in Children with NF1 Using Lab-Based and Functional Measures: Influence of ADHD?* Poster presented at the International Neuropsychology Society, Washington, DC.

Casnar, C., **Yund, B**., Lee, K., & Klein-Tasman, B.P. (2018, February). *Autism Spectrum Disorder Symptomatology in Children with Neurofibromatosis type 1.* Poster presented at the Children's Tumor Foundation NF Conference, Washington, DC.

Yund, B., Casnar, C., Lee, K., Klein-Tasman, B.P. (2017, February). *Relations between Parent Report of Attention and Sleep in Children with Neurofibromatosis type 1*. Poster presented at the International Neuropsychological Society, New Orleans, LA.

Casnar, C., **Yund, B.**, Lee, K., Klein-Tasman, B.P. (2017, February). *ASD Symptomatology and Related Variables in Children with Neurofibromatosis type 1*. Poster presented at the International Neuropsychological Society, New Orleans, LA.

Yund, B., Mervis, C.B., Klein-Tasman, B.P. (2016, March). *Aggression in Children with 7q11.23 Duplication Syndrome: A Preliminary Examination*. Poster presented at the Gatlinburg Conference, San Diego, CA.

Yund, B., Haberman, D., Casnar, C., & Klein-Tasman, B.P. (2016, February). *Parent Reported Temperament in Young Children with Neurofibromatosis type 1.* Poster presented at the International Neuropsychological Society, Boston, MA.

Nestrasil, I., Shapiro, E., Kovac, V., Wakumoto, A., Ahmed, A., Delaney, K., **Yund, B.**, Rudser, K., Barbier, A., Haslett, P., & Whitley, C. (2015, May). *Brain MRI Patterns of Disease Progression in Sanfilippo Syndrome type A (MPS IIIA).* Poster presented at the European Paediatric Neurology Society Congress, Vienna, Austria.

King, K., Ahmed, A., Rudser, K., Rumsey, R., **Yund, B.**, Nestrasil, I., Potegal, M., Whitley, C., Shapiro, E. (2015, February). *Neurobehavioral Outcomes in Sanfilippo Syndrome type B Compared to type A.* Poster presented at the Lysosomal Disease Network's WORLD Symposium, Orlando, FL.

Yund, B., Rudser, K., Kovac, V., Ahmed, A., Nestrasil, I., Delaney, K., Whitley, C., Shapiro, E. (2014, February). *White Matter Structure and Function in Attenuated MPS II.* Poster presented at the Lysosomal Disease Network's WORLD Symposium, San Diego, CA.

Kovac, V., **Yund, B.**, Ahmed, A., Rudser, K., Shapiro, E., Nestrasil, I. (2014, February). *The Development of Brain and Neurocognitive Function in Typically Developing Children Ages 4-7 Years*. Poster presented at the Lysosomal Disease Network's WORLD Symposium, San Diego, CA.

Shapiro, E., King, K., Delaney, K., Ahmed, A., **Yund, B.**, Rudser, K., Whitley, C. (2014, February). *Factors Affecting Psychological Adjustment in MPS I Patients: An Exploratory Study and Clinical Observations.*

Poster presented at the Lysosomal Disease Network's WORLD Symposium, San Diego, CA.

Nestrasil, I., Bednarik, P., **Yund, B**., Delaney, K., Kovac, V., Ahmed, A., Shapiro, E. (2014, February). *Brain MRI Abnormalities in Mucopolysaccharidosis type I: Cross-Sectional Study.* Poster presented at the Lysosomal Disease Network's WORLD Symposium, San Diego, CA.

Woodworth, K., **Yund, B.**, Rudser, K., Ahmed, A., Shapiro, E. (2014, February). *Age and Developmental Change in Adaptive Behavior in Severe and Attenuated Mucopolysaccharidosis Type 1*. Poster presented at the Lysosomal Disease Network's WORLD Symposium, San Diego, CA.

Kunin-Batson, A., Ahmed, A., **Yund, B.**, Whitley, C., Shapiro, E. (2014, February). *The MPS Health Assessment Questionnaire: Preliminary Normative and Validity Data.* Poster presented at the Lysosomal Disease Network's WORLD Symposium, San Diego, CA.

Ahmed, A., Nestrasil, I., Kovac, V., Delaney, K., **Yund, B.**, Harmatz, P., Shankar, S., Whitley, C., Shapiro, E. (2014, February). *Brain Volumes and Cognition in Mucopolysaccharidosis type VI.* Poster presented at the Lysosomal Disease Network's WORLD Symposium, San Diego, CA.

King, K., Rudser, K., Kovac, V., Nestrasil, I., **Yund, B.**, Delaney, K., De Bellis, MD, Whitley, C., Shapiro, E. (2014, February). *Attention and Corpus Callosum Volumes in Individuals with Hurler and Hurler-Scheie Syndromes and Controls.* Poster presented at the Lysosomal Disease Network's WORLD Symposium, San Diego, CA.

Yund, B., Kovac, V., Nestrasil, I., Delaney, K., Rudser, K., Nguyen-Driver, M., Steiner, R., Shapiro, E. (2013, February). *Neuropsychological Function and Brain Abnormalities in Children with Attenuated Mucopolysaccharidosis type II*. Poster presented at the Lysosomal Disease Network's WORLD Symposium, Orlando, FL.

Kunin-Batson, A., **Yund, B**., Rudser, K., Shapiro, E. (2013, February). *Quality-of-life in Children with Hurler Syndrome Who Have Not Yet Been Transplanted and Those Who Are One-Year Post Transplant*. Poster presented at the Lysosomal Disease Network's WORLD Symposium, Orlando, FL.

Delaney, K., King, K., **Yund, B**., Rudser, K., Whitley, C., Shapiro, E. (2013, February). *Memory in Preschool Children with MPS I and II.* Poster presented at the Lysosomal Disease Network's WORLD Symposium, Orlando, FL.

Shapiro, E., Delaney, K., Nestrasil, I., Ahmed, A., King, K., **Yund, B**, Clarke, L, Raiman, J., Harmatz, P., Steiner, R., Pastores, G., Shankar, S., Whitley, C. (2013, February). *Longitudinal Studies of Brain Structure and Function in MPS disorders: A Study of the Lysosomal Disease Network.* Poster resented at the Lysosomal Disease Network's WORLD Symposium, Orlando, FL.

Shapiro, E., Nestrasil, I., Rudser, K., Delaney, K., **Yund, B**., Ahmed, A., King, K., Kovac, V., Raiman, J., Harmatz, P., Steiner, R., Shankar, S., Whitley, C. (2013, February). *Cognition and Neuroimaging in MPS I: Effects of Age, Severity, and Treatment*. Poster presented at the Lysosomal Disease Network's WORLD Symposium, Orlando, FL.

Nestrasil, I., Delaney, K., **Yund, B**., Ahmed, A., Rudser, K., Kovac, V., Haslett, P., Richard, C., Whitley, C., Shapiro, E. (2013, February). *Longitudinal Change in Brain Volumes and Cognition Function in MPS IIIA.* Poster resented at the Lysosomal Disease Network's WORLD Symposium, Orlando, FL.

Potegal, M., **Yund, B**., Rudser, K., Delaney, K., Nestrasil, I., Whitley, C., Shapiro, E. (2013, February). *Klüver–Bucy-like Symptoms in Sanfilippo syndrome.* Poster presented at the International Neuropsychological Society meeting, Waikoloa, HI.

Romero, R.A., **Yund, B**., Erickson, N., Woodworth, K., Bangirana, P., Boivin, M., Wong, J., Shapiro, E. (2013, February). *Neurocognitive effects of HIV on preschool-aged children in Uganda*. Poster presented at the International Neuropsychological Society meeting, Waikoloa, HI.

Yund, B., Rudser, K., Delaney, K., Kovac, V., Nguyen-Driver, M., Steiner, R., Shapiro, E. (2013, February).

Brain Abnormalities and Neuropsychological Function in Children with Attenuated Mucopolysaccharidosis type II. Poster presented at the International Neuropsychological Society meeting, Waikoloa, HI.

Nestrasil, I., Delaney, K**., Yund, B**., Ahmed, A., Rudser, K., Kovac, V., Haslett, P., Richard, C., Whitley, C., Shapiro, E. (2012, July). *Brain Volume and Cognitive Function in MPS IIIA: A Longitudinal Study*. Presented at the International Symposium on MPS and Related Diseases, Noordwijkerhout, Netherlands.

Yund, B., Delaney, K., Whitley, C., Potegal, M., Shapiro, E. (2012, February). *Visual Attention in Sanfilippo Type A.* Poster presented at the Lysosomal Disease Network's WORLD Symposium, San Diego, CA.

Yund, B., Ahmed, A., Delaney, K., Nestrasil, I., Rudser, K., Whitley, C., Shapiro, E. (2012, February). *Brain Structure and Function in MPS II.* Poster presented at the Lysosomal Disease Network's WORLD Symposium, San Diego, CA.

Potegal, M., **Yund, B**. Rudser, K., Delaney, K., Nestrasil, I., Whitley, C., Shapiro, E. (2012, February). *Mucopolysaccharidosis Type IIIA as a Variant of Klüver–Bucy Syndrome: A Comparison of Social/Emotional Characteristics of Children with MPS IIIA to those with MPS IH*. Poster presented at the Lysosomal Disease Network's WORLD Symposium, San Diego, CA.

King, K., Ahmed, A., **Yund, B**., Rudser, K., Shapiro, E. (2012, February). *Verbal Memory and Hippocampal Volume in Individuals with MPSI.* Poster presented at the Lysosomal Disease Network's WORLD Symposium, San Diego, CA.

Shapiro, E., Delaney, K., Nestrasil, I., Ahmed, A., **Yund, B.**, King, K., Whitley, C., et al. (2012, February). *Longitudinal Studies of Brain Structure and Function in MPS Disorders: A Study of the Lysosomal Disease Network.* Poster presented at the Lysosomal Disease Network's WORLD Symposium, San Diego, CA.

Ahmed, A., Zuck, T., Delaney, K., **Yund, B.**, Whitley, C., Shapiro, E. (2012, February). *Medical Outcomes and Adaptive Functions in Severe and Attenuated MPS I*. Poster presented at the Lysosomal Disease Network's WORLD Symposium, San Diego, CA.

Kunin-Batson, A., Erickson, N., Ahmed, A., **Yund, B.**, Shapiro, E. (2012, February). *Quality of Life after Treatment for Mucopolysaccharidoses*. Poster presented at the Lysosomal Disease Network's WORLD Symposium, San Diego, CA.

Potegal, M., **Yund, B.**, Shapiro, E. (2011, February). *Comparison of Social/Emotional Function in Children with MPS I and MPS III: Interim Report*. Poster presented at the Lysosomal Disease Network's WORLD Symposium, Las Vegas, NV.

PRESENTATIONS AT LOCAL/REGIONAL MEETINGS & CONFERENCES

French, S., Rivera, K., **Yund, B.**, Klein-Tasman, B. *Behavioral Play Therapy with Children with Williams Syndrome: Examination of Parent Ratings of Anxiety*. (2019, April). Poster presented at UWM Undergraduate Research Symposium, Milwaukee, WI.

Corrigan, E., **Yund, B.**, Lee, K., & Klein-Tasman, B.P. (2017, April). *Longitudinal Examination of Anxiety Levels in Children with Neurofibromatosis type 1*. Poster presented at UWM Undergraduate Research Symposium, Milwaukee, WI.

Corrigan, E., Bielunski, G., Stemper, B., Lee, K., **Yund, B**., Casnar, C., & Klein-Tasman, B.P. (2016, April). *Examination of Anxiety Levels in Children with Neurofibromatosis type 1.* Poster presented at UWM Undergraduate Research Symposium, Milwaukee, WI.

TEACHING EXPERIENCE

Fall 2018 **Guest Lecturer: Psych 912 Developmental Psychopathology (Graduate Level)**

University of Wisconsin – Milwaukee, Department of Psychology
Topic: *Autism Spectrum Disorder and Intellectual Disability*

Fall 2018 **Guest Lecturer: Psych 802 First Year Clinical Practicum (Graduate Level)**
University of Wisconsin – Milwaukee, Department of Psychology
Topic: *Clinical Use of the Wechsler Individual Achievement Test – Third Edition*

Fall 2018 **Teaching Assistant: Psych 821: Second Year Clinical Practicum**
University of Wisconsin – Milwaukee, Department of Psychology
Supervisors: Kristin Smith, Ph.D., Han-Joo Lee, Ph.D.; Bonnie Klein-Tasman, Ph.D.

2016 – 2017 **Teaching Assistant: Psych 802/821 First and Second Year Clinical Practicum**
University of Wisconsin – Milwaukee, Department of Psychology
Supervisors: Kristin Smith, Ph.D.; Han-Joo Lee, Ph.D.

2015 – 2016 **Teaching Assistant: Psych 802 First Year Clinical Practicum**
University of Wisconsin – Milwaukee, Department of Psychology
Supervisors: Kristin Smith, Ph.D.; Han-Joo Lee, Ph.D.

Fall 2014 **Teaching Assistant: Psych 260 Child Psychology**
University of Wisconsin – Milwaukee, Department of Psychology
Instructor: Kristin Smith, Ph.D.

PROFESSIONAL MEMBERSHIPS

2018 – present Association for Psychological Science, Student Affiliate
2018 – present APA, Division 40 (Clinical Neuropsychology), Student Affiliate
2016 – present American Psychological Association, Student Affiliate
2016 – present International Neuropsychological Society
2014 – present UWM Association of Graduate Students in Neuropsychology

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SEMINARS, SPECIALIZED TRAINING, and CERTIFICATIONS

2017 – 2019 **Neuropsychology Journal Club**
Medical College of Wisconsin, Milwaukee, WI
Weekly journal club involving discussion of clinical neuropsychology research.

2017 – 2019 **Neuropsychology Seminar**
Medical College of Wisconsin, Milwaukee, WI
Weekly didactic seminar involving broad topics related to clinical neuropsychology.

2017 – 2018 **Cases in Clinical Neuropsychology**
University of Wisconsin-Milwaukee
Faculty Supervisor: Krista Lisdahl, Ph.D.
Graduate seminar involving fact finding sessions and broad topics related to clinical neuropsychology.

Fall 2016 **Evidence-Based Practice for Treatment of Eating Disorders**
University of Wisconsin – Milwaukee, Milwaukee, WI
*Lecturer**:** Stacey Nye, Ph.D., FAED*
Graduate seminar in etiology, assessment, and diagnosis of DSM-5 feeding and eating disorders, the physiological, metabolic, and medical complications related to malnutrition, issues and controversies involved with weight stigma and dieting, ethical issues clinicians face in context of treatment, and basic principles of an integrative treatment approach with an eating disordered population.

Spring 2016	**Behavioral Activation for Depression** University of Wisconsin – Milwaukee, Milwaukee, WI *Lecturer: Christopher Martell, Ph.D.* Graduate seminar in basic theory, principles, and practice of utilizing behavioral activation in the treatment of depressive disorder. Instruction in the development of activity scheduling, functional analysis, behavioral experiments, and combating rumination with cognitive strategies.
Fall 2016	**ADOS-2 Research Reliability** (Modules 3 & 4)
Fall 2015	**ADOS-2 Clinical Training** University of Wisconsin – Madison, Madison, WI *Wisconsin LEND Training Program* Seminar training in the development, psychometric properties, administration, scoring, and operationalizing diagnostic criteria for ASD using the ADOS-2.

HONORS and AWARDS

2019	UWM Association of Graduate Students in Neuropsychology Travel Award
2018	UWM Graduate Student Travel Award
2018	UWM Association of Graduate Students in Neuropsychology Travel Award
2017	UWM Graduate Student Travel Award
2016	David Zeaman Graduate Student Award, Gatlinburg Conference
2016	UWM Graduate Student Travel Award
2016	UWM Association of Graduate Students in Neuropsychology Travel Award
2015	UWM Department of Psychology Summer Fellowship
2009 – 2010	Dean's List, University of Minnesota-Twin Cities