12-2022

Self-regulation in young school-aged children with Williams syndrome.

Holley Pitts Arnold
University of Louisville

SELF-REGULATION IN YOUNG SCHOOL-AGED CHILDREN WITH WILLIAMS SYNDROME

By

Holley Pitts Arnold
B.A. University of South Carolina, 2010
M.S., University of Louisville, 2015

A Dissertation
Submitted to the Faculty of the
College of Arts and Sciences of the University of Louisville
in Partial Fulfillment of the Requirements
for the Degree of

Doctor of Philosophy
In Experimental Psychology

Department of Psychological and Brain Sciences
University of Louisville
Louisville, Kentucky

December 2022

SELF-REGULATION IN YOUNG SCHOOL-AGED CHILDREN WITH WILLIAMS SYNDROME

By

Holley Pitts Arnold
B.A., University of South Carolina, 2010
M.S., University of Louisville, 2015

A Dissertation Approved on

November 16, 2022

by the following Dissertation Committee:

_____
Carolyn B. Mervis, Ph.D.
Dissertation Director

_____
Cara H. Cashon, Ph.D.

_____
Deborah W. Davis, Ph.D.

_____
John R. Pani, Ph.D.

_____
Paul J. Rosen, Ph.D.

DEDICATION

To the wonderful children and their families who I have had the privilege to meet and work with during my time at the University of Louisville.

To the many people who have motivated, supported, and cheered me on throughout my life and graduate school. In particular, to my amazing Mom and Dad, for providing me with so much love, support, and oppurtunity. To my Granny and Papa, for their unconditional love, encouragment, and optimism. To my brother, Alex, for simltaneously being a voice of reason, compassion, and sarcasm. To my niece and pilot participant, Eliza, who brings me so much joy. To my husband, Bob, for being patient, understanding, and bringing me my “too strong” coffee every day.

This dissertation is also dedicated in loving memory of my Grandmama.

ACKNOWLEDGMENTS

I would like to acknowledge my committee members, Dr. Carolyn Mervis, Dr. Cara Cashon, Dr. Deborah Winders Davis, Dr. John Pani, and Dr. Paul Rosen. I am grateful for their time and feedback. A special thank you to Carolyn for her expertise, mentorship, support, and guidance.

I would also like to thank the members of the Neurodevelopmental Sciences Lab—especially Dr. Angela Becerra, Adam Moseley, Dr. Caroline Richter, and Gopika Gopan—for helping conduct this research project. Additionally, I appreciate all the families who participated in this research for giving me the opportunity to work with their children.

I want to acknowledge the following financial support provided to this project through grants awarded to my mentor, Carolyn: Williams Syndrome Association (WSA 0104, WSA 0111), National Institute of Child Health and Human Development (R37 HD29957), and National Institute of Neurological Disorders and Stroke (R01 NS35102).

I want to thank my husband, my family, and my friends who became my family for their unwavering patience, acceptance, and support. Lastly, I want to thank my "dogter," Carmie, who was literally by my side for most of the writing of this dissertation.

ABSTRACT

SELF-REGULATION IN YOUNG SCHOOL-AGED CHILDREN WITH WILLIAMS SYNDROME

Holley Pitts Arnold

November 16, 2022

My dissertation included two manuscripts which broadly focused on the self-regulation abilities of young school-aged children with Williams syndrome (WS), a rare neurodevelopmental disorder. Children with WS often exhibit mild to moderate intellectual disability (Kozel et al., 2021), impairments in behavioral and emotional regulation (Greiner de Magalhães et al., 2022), low effortful control (Leyfer et al., 2012), and deficits in adaptive skills (Brawn & Porter, 2018). In the first manuscript, the performance of children with WS on a gift-wrap delay of gratification task was characterized. In the second manuscript, the concurrent effects of the ability to regulate emotions, the ability to regulate behaviors, and overall intellectual ability on adaptive skills were explored.

Overall, results demonstrated that young school-aged children with WS have difficulty with both the emotional and behavioral aspects of self-regulation. At the same time, the ability to regulate emotions and behaviors was characterized by considerable variability. In line with the literature on typically developing children, the ability to inhibit the urge to access an enticing reward as measured by a gift-delay task was related

to regulation of emotions and inhibitory control for young school-aged children with WS. Furthermore, lower self-regulatory skills and more limited intellectual ability were each associated with limitations in adaptive functioning. In addition to overall intellectual ability, emotion-related self-regulation contributed a substantial amount of unique variance to individual differences in socialization skills and daily living skills.

Results from this dissertation suggest the development and validation of interventions targeting the enhancement of emotion regulation by children with WS and other syndromes associated with intellectual disability is crucial for optimizing the opportunity of these children to reach their full potential.

Keywords: Williams syndrome, self-regulation, inhibitory control, emotion regulation, executive function, effortful control, adaptive function, intellectual disability

TABLE OF CONTENTS

LIST OF TABLES

TABLE PAGE

CHAPTER I

GENERAL INTRODUCTION

When children enter elementary school, demands and expectations increase in both the home and school settings (McClelland & Cameron, 2012). For example, children are expected to persist to learn new academic concepts that are challenging, stay focused on specific tasks rather than doing something that is preferred, and overcome impulses to comply with social norms. Self-regulation, which describes children's ability to purposefully or automatically regulate their actions, attention, emotions, or thoughts (see Eisenberg & Zhou, 2016; Karoly, 1993), is foundational to success in meeting these expectations (Diamond, 2013, 2016; McClelland & Cameron, 2012; Nigg, 2017). There is increasing evidence that children who have genetic syndromes associated with intellectual disability (ID) have impaired self-regulatory skills, as indicated by difficulties with emotion regulation, impulsivity, and other behavioral challenges (see Shaffer et al., in press for review; Cuskelly et al., 2013).

Williams syndrome (WS) is a rare neurodevelopmental disorder associated with mild to moderate ID (Kozel et al., 2021). Children with WS often exhibit impairments in emotional and behavioral aspects of self-regulation as well as difficulties in socialization, communication, and daily living skills (Brawn & Porter, 2018; Mervis & Greiner de Magalhães, 2022). Although individuals with WS are frequently described as indiscriminately friendly and having a strong desire to interact with others, individuals with WS have difficulty maintaining friendships and are at risk for victimization and

bullying (Fisher et al., 2017; Thurman & Fisher, 2015). Impairments in emotion regulation and/or behavior regulation may play a role in these difficulties.

In my dissertation, I present two studies that focus on self-regulatory skills in young school-aged children with WS. My dissertation is divided into four chapters. Chapter 1 consists of a general introduction. First, I provide a brief overview of the prior research on self-regulation as well as literature addressing the relation between self-regulation and functional outcomes for typically developing (TD) children or individuals with ID. Then, I briefly describe WS and provide an overview of prior research on self-regulation skills. Finally, I briefly state the research questions to be addressed in the two empirical studies included in my dissertation. In Chapter 2, I report a study which evaluated the measurement validity of the emotionally salient gift-delay task for children with WS. To my knowledge, no previous study has evaluated emotion related aspects of self-regulation using a performance-based measure for children with WS. In Chapter 3, I report a study which explored the unique contributions of emotional and behavioral aspects of self-regulation and overall intellectual ability to individual differences in adaptive functioning. Chapter 4 consists of a general discussion. I provide an overview of the findings of the two dissertation studies, implications, and future directions.

Self-Regulation

In the research literature, the terms "self-regulation," "executive function," and "effortful control" are often used interchangeably (Diamond, 2013; Nigg, 2000, 2017; Zhou et al., 2012). In the current study, "self-regulation" refers to an umbrella construct to describe the processes by which an individual purposefully or automatically regulates his or her actions, attention, emotions, or thoughts (see Eisenberg & Zhou, 2016; Karoly,

1993). Self-regulation can be divided into emotional (hot) and non-emotional (cool) aspects (Willoughby et al., 2011; Zelazo & Müller, 2010). A key distinction between these interrelated aspects is the relative involvement of the regulation of emotion and motivation: Emotion regulation predominates in situations when motivation and emotions are higher, whereas unemotional (behavior/cognitive) regulation predominates in emotionally neutral circumstances (Zelazo & Carlson, 2012; Zelazo & Müller, 2010).

Overlapping with self-regulation, executive functions are integral in the goal-directed regulation of behaviors, thoughts, and emotions (Diamond, 2013; Miyake et al., 2000; Zelazo & Carlson, 2012). Effortful control is the aspect of temperament which refers to a child's propensity to self-regulate (Diamond, 2013; Nigg, 2017). It includes both the ability to focus attention and the ability to inhibit and replace an automatic response and is considered an innate, stable personality characteristic (Diamond, 2016; Rothbart & Bates, 2006). Inhibitory control—the ability to overcome prepotent responses (Garon et al., 2008; Miyake et al., 2000)—is a core component of executive function and effortful control (see Diamond et al., 2007; Nigg, 2017; Zhou et al., 2012 for reviews), both of which are associated with self-regulatory processes and share both conceptual and methodological overlap (Zhou et al., 2012).

In the executive function and effortful control literatures, hot emotion-related inhibitory control has often been evaluated using a delay paradigm in which an enticing reward (e.g., toy, gift, food) is presented (Allan et al., 2014). The participant's task is to delay gratification by inhibiting a behavior (e.g., eating, playing, peeking) while waiting on the examiner. As the regulation of emotions assists in problem-solving to overcome

the temptation, the more desirable the reward, the "hotter" the task may be (Carlson & Wang, 2007; Zelazo & Carlson, 2012).

Delay tasks can vary in cognitive demands, difficulty, and motivation (Garon et al., 2008; Luerssen & Ayduk, 2013). For example, in the classic marshmallow task (Mischel et al., 1972) the child is given a choice—have one marshmallow now or wait and receive the preferred, larger reward of two marshmallows. The motivator to delay gratification is to get more marshmallows. To succeed at this task, children need to have the capacity to understand the contingency (if they wait, then they will get a larger reward) and to find marshmallows enticing.

In the gift-wrap delay paradigm (Kochanska et al., 1996), the child is told not to peek while a gift is being noisily wrapped by a researcher. Success at this task involves impulse control because it requires the participant to overcome the temptation to peek and instead to wait for the surprise (Kim et al., 2013). The motivator to delay is to follow the rules set by the examiner to receive social approval and so the gift may remain a surprise (Garon et al., 2008). Thus, the demands of the gift delay task are less sophisticated than the marshmallow task (Garon et al., 2008; Luerssen & Ayduk, 2013), making the gift delay task ideal for younger children, children who may not be food motivated, or children who may have cognitive impairments.

Cool, or unemotional, inhibitory control has often been evaluated using Stroop-like tasks which involve interference control, or inhibition of attention to distractors and prepotent responses in an emotionally neutral context (Diamond, 2016; Liu et al., 2015; Montgomery & Koeltzow, 2010). Verbal Stroop-like tasks (similar to verbal opposites tasks) require the participant to overcome a verbal prepotent response and replace it with

the response specified by the researcher (Diamond, 2013). For example, in the Day-Night task, the participant must say "day" when shown a moon and "night" when shown a sun (Gerstadt et al., 1994).

During the preschool and early school years, TD children evidence rapid improvement in inhibitory control as indicated by performance-based measures. For example, during the gift-task, 42% of young 3-year-olds but 74% of 5-year-olds never peeked during the gift-wrapping process (Carlson, 2005). On the classic Day/Night Stroop-like task, mean percentage of correct responses was 69% for young 4-year-olds but 87% for 6-year-olds (Gerstadt et al., 1994; Montgomery & Koeltzow, 2010). Inhibitory control continues to improve at a slightly slower rate during the early school years (ages 6 – 8 years) and more slowly throughout adolescence (Best & Miller, 2010).

Contributions of Emotion Regulation and Effortful Control

Previous research on TD children has indicated that emotion regulation facilitates hot inhibitory control when children are faced with an enticing reward that they are asked to refrain temporarily from accessing (see Holodynski et al., 2013; Zelazo & Cunningham, 2007 for reviews). Although hot inhibitory control is thought to be facilitated primarily by emotion regulation (Carlson & Wang, 2007; Zelazo & Carlson, 2012), cool behavioral and attentional control may also contribute to a lesser extent (Eisenberg & Zhou, 2016; Zelazo & Müller, 2010).

Effortful control is thought to be utilized in delay tasks and has been implicated in emotion regulation (Eisenberg et al., 2014; Eisenberg & Zhou, 2016; Kim et al., 2013). Gusdorf et al. (2011) administered a battery of delay of gratification measures (including a gift delay task) to a large group of TD 3-year-olds and evaluated the components of

effortful control. Parent report of inhibitory control, the ability to plan and suppress inappropriate responses when instructed to do so or in novel situations, was significantly related to the performance-based delay-of gratification factor score but attentional focusing, the ability to maintain attentional focus/stay on task, was not.

Relations between Lab-based Measures and Parental Ratings

Relations between performance-based lab measures and parental ratings of self-regulation are generally low. Performance-based lab measures directly and objectively assess self-regulation in an optimal, controlled environment (Allan et al., 2014; Toplak et al., 2013). The researcher determines the goal of the task and tells the child what is expected from him or her. Then, how well the child performs is measured. Parent report measures capture the parent's perspective on the child's self-regulation abilities during daily life (Toplak et al., 2013). These ratings characterize the child's usual behaviors and emotions across a wide range of situations in which the child does not have support and has his/her own goal in mind (Toplak et al., 2013). Although similar processes may be involved in self-regulation in the real-world settings that are assessed in parent-report measures and the controlled laboratory setting, the constructs assessed in parent-report measures and direct assessments are complementary, not identical (Isquith et al., 2013; Toplak et al., 2013).

Toplak et al. (2013) conducted a review of 13 studies that reported associations between performance-based measures (all of which assessed cool self-regulation) and informant ratings of executive functioning in the context of self-regulation. The resulting correlations ranged from very weak to moderate, and only 19% were statistically significant, with an average effect size of .15. Similar findings were reported by Esbensen

and Hoffman (2018) for children with Down syndrome (DS) for relations between performance-based tasks of inhibitory control and parent report of self-regulation. The low correlations between parent-report measures of everyday skills and performance-based measures administered in a research lab are likely due to differences in the type of skills assessed and in task demands (Isquith et al., 2013).

Adaptive Functioning and Self-Regulation

Adaptive skills include the ability to function in social situations, exchange information with others, and perform tasks of daily living (Sparrow et al., 2016). Conceptually, being able to regulate one's own behaviors and emotions appropriately allows an individual to better navigate the demands and complexities of societal norms in day-to-day life (Baumeister et al., 2007). Empirically, findings from a recent meta-analysis indicated that self-regulation measured when children were in preschool was positively associated with social competence, academic achievement, and school engagement and negatively associated with externalizing problems, internalizing problems, and peer victimization several years later. The relation with early self-regulation extended into adulthood; early difficulty with self-regulatory skills was associated with depression, anxiety, and unemployment in adulthood (Robson et al., 2020).

For TD preschool and young school-aged children, hot and cool aspects of self-regulation have been shown to differentially impact functional outcomes (Allan et al., 2014; Backer-Grøndahl et al., 2019; Bassett et al., 2012; Brock et al., 2009; Di Norcia et al., 2015; Kim et al., 2013; Mulder et al., 2014). For example, teacher-reported social competence (i.e., sensitivity and cooperation with others) was uniquely predicted by

children's hot inhibitory control latent factor score derived from performance on a battery of direct assessments, but not by the cool latent factor. (Bassett et al., 2012). In contrast, for a large sample of kindergarteners, overall intellectual ability was moderately related to performance on the cool inhibitory control factor score but only weakly correlated with the hot inhibitory control factor score (Brock et al., 2009).

Relations between adaptive behavior and self-regulation also have been considered for individuals with DS. For adolescents with DS, when the relations between adaptive behavior and a cool performance-based measure of inhibitory control were considered, a significant and large positive correlation between teacher-reported conceptual (communication, functional academic, and self-direction) skills and performance on a spatial interference control task was found. For parent-reported conceptual skills, the correlation was moderate and positive, although not statistically significant (Sabat et al., 2020). For children and adolescents aged 7 – 16 years, parent-reported socialization, communication, and daily living skills were moderately and significantly correlated with parental ratings of both emotion regulation and behavior regulation (Onnivello et al., 2022).

Williams Syndrome

WS is a neurodevelopmental disorder caused by a hemideletion of 25 – 27 genes on chromosome 7q11.23 (Kozel et al., 2021). WS occurs in 1 in 7,500 live births (Strømme et al., 2002) at the same frequency for males and females (Morris et al., 2020). WS is characterized by specific cognitive, behavioral, and personality profiles (see Mervis & Greiner de Magalhães, 2022 for review). For example, children with WS are described as indiscriminately friendly, socially disinhibited, more inclined to

inappropriately approach strangers than TD children or children with ID of other etiologies, gregarious, having low tolerance for frustration, low persistence, and attention problems (Davies et al., 1998; Dodd et al., 2010; Jones et al., 2000; Klein-Tasman & Mervis, 2003; Mervis & Klein-Tasman, 2000; Pérez-García et al., 2017; Zitzer-Comfort et al., 2007).

On average, individuals with WS have mild to moderate ID with the full range of abilities extending from severe ID to average relative to the general population (Martens et al., 2008; Mervis & Greiner de Magalhães, 2022; Mervis & John, 2010). Within the cognitive profile, a pattern of relative strengths in verbal and/or nonverbal reasoning abilities and a relative weakness in spatial abilities is typically observed (see Mervis & John, 2010 for review). For children and adolescents with WS, higher intellectual ability is associated with that better adaptive functioning (Dimitropoulos et al., 2009; Mervis et al., 2001). On average, the overall level of adaptive skills is in the mild to moderate disability range, with abilities extending from severe disability to average relative to the general population (see Brawn & Porter, 2018 for systematic review). At the group level, children with WS exhibit relative strengths in socialization skills and communication skills and a relative weakness in daily living skills. However, variability in the pattern of relative strengths and weaknesses shown by individual children has been reported (see Brawn & Porter, 2018 for systematic review).

Self-Regulation: Lab-based Direct Assessment

As indicated by both direct assessment and parent ratings, children with WS evidence considerable difficulty with self-regulatory skills. Four studies of children with WS have directly assessed inhibitory control using cool affectively neutral performance-

based measures (Atkinson et al., 2003; Breckenridge et al., 2013; Carney et al., 2013; Tager-Flusberg et al., 1997). Findings consistently indicated deficits in inhibitory control, with performance significantly worse than similarly aged TD peers and either similar to or significantly worse than similarly aged children with other genetic disorders associated with impairments in executive functions such as DS or Prader-Willi syndrome. None of these studies addressed relations between task performance and either overall intellectual ability or parent-report of emotion or behavior regulation. To the best of our knowledge, no studies have evaluated performance on either the gift-wrap task or any other direct assessment of hot inhibitory control in individuals with WS.

Self-Regulation: Parental Ratings

Parental ratings have also indicated that deficits in regulating emotion and actions during everyday conditions are common among children with WS (Camp et al., 2016; Greiner de Magalhães et al., 2022; Phillips, 2008; Woodruff-Borden et al., 2010). When compared to similarly aged individuals with DS matched for nonverbal ability, adolescents and young adults with WS performed worse on parental ratings of aspects of both emotion and behavior regulation (Camp et al., 2016).

Parent ratings of temperament and personality characteristics have indicated children with WS exhibit low effortful control as measured by the CBQ (Leyfer et al., 2012). Relative to the CBQ authors' reference sample of TD 6 – 7-year-olds (Rothbart et al., 2001), 5 – 10-year-olds with WS on average were rated significantly worse for both the inhibitory control and attentional focusing aspects of effortful control (Leyfer et al., 2012).

Self-Regulation and Adaptive Function

A few studies have evaluated the relation between parent report of self-regulation and adaptive function in individuals with WS. Greiner de Magalhães et al. (2022), using a large sample of 6 – 17-year-olds with WS, found that parental ratings of emotion regulation and behavior regulation were each significantly correlated with personal living skills, social interaction and communication skills, and community living skills. Ng-Cordell et al. (2018) reported that deficits in social interaction were significantly correlated with parent reported difficulty with emotion regulation and behavior regulation for a small sample of 5 – 37-year-olds with WS. Similarly, Phillips (2008), based on a relatively small sample of 8 – 15-year-olds with WS, found that the bivariate correlations between emotion regulation skills and personal living skills, community living skills, and social interaction and communication skills were all statistically significant.

Dissertation Studies

My dissertation included two manuscripts to be submitted for publication to peer-reviewed journals. Broadly, my dissertation focused on the self-regulation abilities of young school-aged children with WS. In the first manuscript (Chapter II), I characterized performance on the gift-delay task, a measure commonly used to evaluate self-regulation in TD children. I also investigated the relations of performance on this lab-based measure of hot inhibitory control to chronological age (CA), overall intellectual ability, and parent report of behavioral regulation, emotion regulation, and effortful control. In the second manuscript (Chapter III), I provided the first empirical evaluation of the *concurrent* effects of the ability to regulate emotions, the ability to regulate behaviors, and overall

intellectual ability on adaptive skills in children with WS. Both lab-based measures and parent report measures of emotion regulation and behavior regulation were considered.

CHAPTER II

HOT INHIBITORY CONTROL IN CHILDREN WITH WILLIAMS SYNDROME: RELATIONS BETWEEN DELAY OF GRATIFICATION, INTELLECTUAL ABILITY, AND SELF-REGULATION[1]

When children enter elementary school, demands and expectations increase in both the home and school settings (McClelland & Cameron, 2012). For example, children are expected to persist to learn new academic concepts that are challenging, stay focused on specific tasks rather than doing something that is preferred, and overcome impulses in order to comply with social norms. Inhibitory control – a broad construct which includes aspects of self-control such as the regulation of actions, attention, and emotions to overcome impulses and delay gratification – is integral to success in meeting these expectations (Diamond, 2013, 2016; McClelland & Cameron, 2012; Nigg, 2017). Depending upon the emotional and motivational context of the situation, cool or hot subdomains of inhibitory control may be differentially activated (Zelazo & Müller, 2010). Cool or cognitive aspects of inhibition predominate in emotionally neutral circumstances, whereas hot or affective aspects of inhibition predominate in situations when motivation and emotions are higher (Zelazo & Carlson, 2012). Thus, cool

[1]This chapter was submitted for publication in *Research in Developmental Disabilities* on November 30, 2022 and is under review.

(cognitive) inhibitory control is more strongly associated with cognitive abilities and academic skills, whereas hot (affective) inhibitory control is more strongly implicated in social skills and persistence (Allan et al., 2014; Backer-Grøndahl et al., 2019; Bassett et al., 2012; Brock et al., 2009; Di Norcia et al., 2015; Kim et al., 2013; Mulder et al., 2014).

Despite its importance, hot aspects of inhibitory control have been directly investigated considerably less often than cool aspects for TD children (Zelazo & Carlson, 2012). The dearth of research on hot aspects of inhibitory control is especially striking in the literature on individuals with intellectual disabilities (ID). For example, although there is evidence from direct assessment that children with Williams syndrome (WS), a rare genetic disorder associated with mild to moderate ID, have impairments in cool inhibitory control (see Mervis & Greiner de Magalhães, 2022 for review), to the best of our knowledge, there are no published studies reporting direct assessment of hot inhibitory control. At the same time, studies using parent report measures have shown that children with WS have impairments in both the behavioral and emotional aspects of self-regulation (Greiner de Magalhães et al., 2022; Phillips, 2008; Woodruff-Borden et al., 2010) and low effortful control (Leyfer et al., 2012). Given these parent-report findings, direct assessment of hot inhibitory control in children with WS is important. In the present study, we evaluated hot inhibitory control in 6 – 8-year-old children with WS using the gift-wrap delay of gratification task developed by Kochanska and her colleagues (1996). We also explored the relations of child performance on this task with chronological age (CA), overall intellectual ability, and parent report of behavior regulation, emotion regulation, and effortful control.

Inhibitory Control: Gift-Wrap Delay of Gratification Task

Inhibitory control—the ability to overcome prepotent responses (Garon et al., 2008; Miyake et al., 2000)—is a core component of effortful control and executive function (see Diamond et al., 2007; Nigg, 2017; Zhou et al., 2012 for reviews), both of which are associated with self-regulatory processes and share some conceptual and methodological overlap (Zhou et al., 2012). In the executive function and effortful control literatures, hot processes have often been evaluated using a delay paradigm (Allan et al., 2014). In this paradigm, an enticing reward is presented, and the participant's task is to delay gratification by inhibiting a prepotent behavior while waiting on the examiner. The gift delay task has been commonly used with TD children (e.g., Bassett et al., 2012; Carlson, 2005; Kim et al., 2013; Kochanska et al., 1996; see Mehsen et al., 2022 for review). In the gift-wrap paradigm (Kochanska et al., 1996), the child is prohibited from looking at a gift while it is being noisily wrapped, creating a novel, motivationally significant situation. Success on this task involves impulse control and requires the participant to overcome the temptation to peek (Kim et al., 2013). The immediate gratification comes with breaking the rule to see the gift, whereas waiting will result in a surprise. Garon (2016) suggested that the gift-wrap task also has a social component as the examiner puts the demand on the child to not peek and complying is likely to result in social approval.

During the preschool years, TD children evidence rapid improvement in inhibitory control during the gift-wrap task: 42% of young 3-year-olds but 74% of 5-year-olds never peeked during the gift-wrapping process (Carlson, 2005; for additional studies with similar results for smaller samples, see Anaya, 2016; Bassett et al., 2012;

O'Toole et al., 2018). Inhibitory control continues to improve at a slightly slower rate during the early school years (ages 6 – 8 years) and more slowly throughout adolescence (Best & Miller, 2010).

1.1.1. Contribution of emotion regulation

As a component of self-regulation, executive functions are integral in the goal-directed regulation of behaviors, thoughts, and emotions (Diamond, 2013; Miyake et al., 2000; Zelazo & Carlson, 2012). Previous research on TD children has indicated that emotion regulation facilitates hot inhibitory control when children are faced with an enticing reward that they are asked to refrain temporarily from accessing (see Holodynski et al., 2013; Zelazo & Cunningham, 2007 for reviews). In line with Zelazo's and Cunningham's (2007) bidirectional hypothesis of the relation between inhibitory control and emotion regulation, Carlson and Wang (2007) proposed that, in the case of the gift-delay of gratification task, the regulation of emotions assists in problem-solving in order to overcome the desire to peek at the gift. Thus, hot inhibitory control is thought to be facilitated primarily by emotion regulation (Carlson & Wang, 2007; Zelazo & Carlson, 2012) although cool behavioral and attentional control may also contribute to a lesser extent (Eisenberg & Zhou, 2016; Zelazo & Müller, 2010).

Emotion regulation includes both emotional control (ability to monitor and modulate emotional responses) and flexible emotion regulation (ability to move smoothly from one facet of a problem, task, or situation to another; sometimes referred to as shifting; similar to attentional deployment; Gioia et al., 2002, 2015). For TD children, both components have been suggested to be involved in overcoming temptation during the gift-wrap task. Carlson and Wang (2007) found that for 4 – 6-year-olds, parent report

of emotional control of positive and negative affect was related to overall performance on a battery of inhibitory control measures which included a gift-delay of gratification task. Furthermore, when faced with an enticing reward, children waited longer if their attention was shifted from the hot temptation and focused on cool irrelevant distractions (see Luerssen & Ayduk, 2014 for review).

1.1.2 Contribution of effortful control

Effortful control is the aspect of temperament which refers to a child's propensity to self-regulate (Diamond, 2013; Nigg, 2017). It includes both the ability to focus attention and the ability to inhibit and replace an automatic response and is considered an innate, stable personality characteristic (Diamond, 2016; Rothbart & Bates, 2006). Conceptually, effortful control is thought to be utilized in delay tasks and has been implicated in emotion regulation (Eisenberg et al., 2014; Eisenberg & Zhou, 2016; Kim et al., 2013). To address this hypothesis, Gusdorf et al. (2011) administered a battery of delay of gratification measures (including a gift delay task) to a large group of TD 3-year-olds. A composite delay of gratification factor score was calculated. Effortful control was measured by parent report on the Children's Behavior Questionnaire (CBQ; Rothbart et al., 2001), which is commonly used to evaluate several empirically defined domains of temperament. As theoretically predicted, parent report of the ability to plan and suppress inappropriate responses when instructed to do so or in novel situations (CBQ Inhibitory Control) was significantly related to the performance-based delay-of gratification factor score but the ability to maintain attentional focus/stay on task (CBQ Attentional Focusing) was not.

Children with Williams Syndrome

WS is a neurodevelopmental disorder caused by a hemideletion of 25 – 27 genes on chromosome 7q11.23 (Kozel et al., 2021). WS occurs in 1 in 7,500 live births (Strømme et al., 2002) at the same frequency for males and females (Morris et al., 2020). WS is characterized by specific cognitive, behavioral, and personality profiles (see Mervis & Greiner de Magalhães, 2022 for review). Individuals with WS typically exhibit mild to moderate ID with the full range extending from severe ID to average relative to the general population (Martens et al., 2008; Mervis & Greiner de Magalhães, 2022; Mervis & John, 2010).

Findings from parent-report studies indicate that impairments in both behavior regulation and emotion regulation are common among children with WS (Greiner de Magalhães et al., 2022; Phillips, 2008; Woodruff-Borden et al., 2010). These difficulties are exemplified by Greiner de Magalhaes et al.'s (2022) findings. Of a sample of 306 6 – 17-year-olds with WS whose primary caregiver completed the Behavior Rating Inventory of Executive Function-2 (BRIEF-2; Gioia et al., 2015), the test authors' criteria for potentially clinically elevated or clinically elevated impairment were met by 52% for emotion regulation and 43% for behavior regulation. Relations between parent report of components of emotion regulation and behavior regulation and parent report of practical problem solving also have been reported. Camp et al. (2016) found that for a sample of 10 – 27-year-olds with WS, flexible emotion regulation, emotional control, and behavioral inhibition as measured by the BRIEF were independently associated with practical problem-solving in novel situations. As difficulties in emotion regulation and behavioral inhibition increased, practical problem-solving abilities decreased.

Parent ratings on the CBQ have been used to describe temperament and personality characteristics of children with WS (e.g., Klein-Tasman & Mervis, 2003; Lai et al., 2020; Leyfer et al., 2012). An exploratory factor analysis indicated that the CBQ Inhibitory Control and Attentional Focusing scales load onto the higher-order Effortful Control domain (Leyfer et al., 2012). Relative to the CBQ authors' normative sample of 341 6 – 7-year-olds (Rothbart et al., 2001), Leyfer et al.'s (2012) sample of 192 5 – 10-year-olds with WS on average was rated significantly worse for both inhibitory control and attentional focusing, with similar amounts of variability.

Four studies of children with WS have directly assessed inhibitory control using cool affectively neutral performance-based measures (Atkinson et al., 2003; Breckenridge et al., 2013; Carney et al., 2013; Tager-Flusberg et al., 1997). Findings consistently indicated deficits in inhibitory control, with performance significantly worse than similarly aged TD peers and either similar to or significantly worse than similarly aged children with other genetic disorders associated with impairments in executive functions such as Down syndrome (DS) or Prader-Willi syndrome.

To the best of our knowledge, no studies have evaluated performance on either the gift-wrap task or any other direct assessment of hot inhibitory control in individuals with WS. However, a study using the gift-wrap delay task with participants with DS was recently published (Fontana et al., 2021). The sample of children and adolescents with DS (mean CA = 12.27 years) reportedly waited 26s before peeking, which was significantly shorter than the 47s reported for the considerably younger group of TD 4 – 8 year-olds (mean CA = 6.17 years). Based on these findings, it is anticipated that children with WS will also evidence deficits in delaying gratification.

Current Study

Although there is evidence that children with WS have deficits in inhibitory control during cool emotionally-neutral contexts (Atkinson et al., 2003; Breckenridge et al., 2013; Carney et al., 2013; Tager-Flusberg et al., 1997), impairments in both behavioral and emotional aspects of executive function (Greiner de Magalhães et al., 2022; Phillips, 2008; Woodruff-Borden et al., 2010), and low effortful control (Leyfer et al., 2012), hot inhibitory control has not been directly evaluated. The purpose of the current study is to characterize the performance of young school-aged children with WS on the gift-wrap delay of gratification task (Kochanska et al., 1996), which has previously been used as a measure of hot, affective inhibitory control in TD children and children and adolescents with DS. We also investigated the relations of performance on this lab-based measure of inhibitory control to CA, overall intellectual ability, and parent report of behavioral regulation, emotion regulation, and effortful control. Three research questions were addressed:

1) How do 6 – 8-year-old children who have WS perform on the gift-wrap task?
2) Do parent-reported aspects of behavior regulation and/or emotion regulation, overall intellectual ability, and CA contribute uniquely to individual differences in gift-wrap task performance?
3) Do parent-reported aspects of effortful control, overall intellectual ability, and CA contribute uniquely to individual differences in gift-wrap task performance?

Method

Participants

The final sample included 34 children (18 girls, 16 boys) with genetically confirmed classic-length WS deletions aged 6.01 – 8.05 years (M = 6.88 years, SD = 0.68). Participants lived in 23 different U. S. states (representing all U. S. census regions: 17.6% Northeast, 44.1% South, 23.5% Midwest, 8.8% West) and one Canadian province (5.9%). The native language for all participants was English. The distribution of participants' racial/ethnic background was: 94.1% White non-Hispanic, 2.9% White Hispanic, and 2.9% multiracial non-Hispanic. Three additional children were excluded because they did not have enough language to understand the gift-wrap task instructions. Data collection began in July 2017 and ended in February 2020.

Measures

Performance-based inhibitory control: Gift-wrap delay of gratification

Inhibitory control was directly assessed using a gift-wrap task which taps into response inhibition and delayed gratification (Kochanska et al., 1996). At the beginning of the task, the child was seated in a small chair facing the wall. The researcher knelt beside the child and informed the child she had a present for him/her but had forgotten to wrap it. The researcher then instructed the child to remain seated in the chair and not look or peek while the researcher wrapped the gift, so it remained a "big surprise." The researcher then walked across the room to a small cabinet that was located behind the child and out of the child's sight. The researcher's back was to the child while she loudly wrapped the gift at the cabinet for approximately 60 seconds (see Figure 1). The

researcher did not respond to the child if the child spoke. If the child approached the researcher, she ignored the child and continued wrapping the gift.

Children's performance during the 60-second gift wrap task was characterized by two measures of ability to delay gratification and two measures of inhibitory control. To describe the ability to delay gratification, the latency to first peek/look was determined. In addition, a dichotomous pass (never looked)/fail (looked or peeked) score was assigned.

To describe inhibitory control, the total number of seconds (out of 60 possible) the child looked away from the gift was determined. This measure went beyond the latency to first look measure by coding the entire 60 second interval, thus giving children who had peeked/looked but then reoriented away from the gift credit not only for the time before they first peeked but also any time after they peaked that they had reoriented and resumed looking away from the gift. In addition, a 4-point ordinal level of inhibitory control score (described in Table 1) was assigned to capture the child's sitting and looking behaviors which were related to compliance with the task goal. Similar to previous studies (e.g., Kim et al., 2013; Kochanska et al., 1996), a gift-wrap task composite score was computed by averaging the *z*-scores for the total duration the child looked away from the gift and the child's level of inhibitory control to best capture inhibitory control performance. For all measures, higher scores indicated greater inhibition.

The task was filmed, and behavioral coding based on the recording was conducted using Behavioral Observation Research Interactive Software (BORIS; Friard & Gamba, 2016). Behaviors were coded by a research assistant and 21% (n = 7) of the videotapes

were independently coded by the first author. Interrater agreement was excellent for both the categorical and continuous variables (rs $\geq$ .99).

Figure 1

Gift-Wrap Delay of Gratification Task

A)  B) 

Note. Screenshots from a videorecording of a participant during the gift-wrap delay of gratification task. (A) At the start of the task, the child faced forward while the examiner nosily wrapped the present behind her. (B) As the task progressed, the child turned around to look at the gift while seated (level of inhibitory control = 1).

Table 1

Distribution of Level of Inhibitory Control Scores

Level of inhibitory control	*n*	%
0 = turned around to look at gift and stood up	7	20.6
1 = turned around to look at gift but remained seated	10	29.6
2 = looked over shoulder at gift but remained seated	6	17.6
3 = never attempted to look at gift and remained seated	11	32.4

Note. Higher scores indicate better performance.

Parent report of self-regulation

The BRIEF-2 (Gioia et al., 2015) is a parent-report questionnaire which evaluates aspects of executive functioning in the context of self-regulation. Using a 3-point scale, parents indicate whether a behavior has never (0), sometimes (1), or often (2) been a problem during the past six months. The current study focused on the Behavior Regulation Index (BRI), Emotion Regulation Index (ERI), and the scales included in each index. The BRI is a composite of the Self-Monitor (ability to monitor one's own behaviors) and Inhibit (ability to stop behaviors and act appropriately) scales. The ERI includes the Emotional Control (ability to modulate emotional responses) and Shift (ability to move flexibly from one facet of a problem, task, or situation to another) scales.

For each index and scale, T-scores are provided (M = 50, SD = 10 for the general population, range: 35 – 90). Higher scores are associated with greater difficulty. Internal consistency for the indices ranged from .94 – .96 for the normative and clinical sample.

Parent report of effortful control

The CBQ (Rothbart et al., 2001) is a parent-report questionnaire which evaluates several empirically defined domains of temperament. Using a 7-point scale, parents indicate whether a statement is extremely untrue (1) to extremely true (7) regarding how their child would react in that situation within the past six months. The current study focused on the two scales that previously were identified to load onto the Effortful Control domain for children with WS (Leyfer et al., 2012): Inhibitory Control (ability to plan and to suppress inappropriate responses when instructed or in novel/uncertain situations) and Attentional Focusing (ability to maintain attentional focus/stay on task). Scale scores are calculated by taking the mean of the scores for the items that load onto

the scale with reverse scoring of negatively worded items. Scale scores range from 1.00 – 7.00. Higher scores indicate better effortful control. For children with WS, internal consistency was .82 for Inhibitory Control and .72 for Attentional Focusing (Leyfer et al., 2012).

Intellectual ability

The Differential Ability Scales-II, Early Years Form (DAS-II; Elliott, 2007) is a standardized assessment of cognitive abilities. The General Conceptual Ability (GCA; similar to Full-Scale IQ), which is a composite of verbal, nonverbal reasoning, and spatial abilities, was used to measure intellectual ability. Standard scores are provided (M = 100, SD = 15 for the general population; range: ~30 – 170). For children aged 6 – 8 years in the normative sample, reliability coefficients ranged from .95 – .97.

Procedure

The university's Institutional Review Board reviewed and approved the study protocol. Parents or legal guardians of all participants provided written informed consent and children provided oral assent. Parent questionnaires were mailed approximately two weeks before the child's assessment and collected upon arrival to the laboratory. The child's mother completed the questionnaires.

The DAS-II and the gift-wrap task were administered to the child as part of the protocol for a larger study of language and cognitive development in children with WS. The DAS-II was administered in a quiet testing room following standardized procedures. The gift-wrap task was conducted in a different testing room which had two remote-controlled video cameras concealed in wooden cabinets with clear glass windows in diagonally opposed corners of the room (Figure 1). The task was filmed by a research

assistant who operated the video cameras from a control room located behind a one-way mirror. The furniture in the room used for the gift-wrap task was set up in a standardized configuration.

Results

Data were analyzed using IBM SPSS 28. First, descriptive statistics were computed to describe performance on the gift-wrap delay of gratification task. To identify significant concurrent predictors of children's performance on the gift-wrap task, three multiple regression analyses were performed. For each regression, the dependent variable was performance-based inhibitory control as measured by the gift-wrap task composite score. Independent variables were centered on the sample mean (Osborne, 2017). All assumptions of multiple linear regression analyses were met. Cohen's f^2 was used to evaluate the effect sizes (0.02 = small, 0.15 = medium, 0.35 = large; Cohen, 1988) of the predictors.

Descriptives: Gift-Wrap Delay of Gratification Task Performance

To describe gift-wrap task performance, frequencies for the categorical levels of inhibitory control are reported in Table 1. About one-third of the children (32.4%) successfully inhibited the urge to peek for 60 seconds. As indicated by the level of inhibitory control variable, the behaviors of the other participants ranged from remaining seated but looking over a shoulder at the gift to standing up and turning around to look at the gift.

Descriptive statistics for the continuous coded variables are indicated in Table 2 first for the full sample and then as a function of the dichotomous pass/fail score. As indicated in the table, there was a wide range observed in both total amount of time

children looked away from the gift during gift-wrapping and latency to first peek. Of the 23 children who failed, all but four disengaged from their first look and resumed looking away for at least part of the remaining time. Thus, the total amount of time children looked away provided a better measure of variability in behavioral inhibition than the time to first look. The *z*-scores for the total amount of time children looked away and level of inhibitory control were significantly positively correlated ($r_s = .79$, $p < .001$) and were averaged to form the gift-wrap task composite score. This composite score was included as the dependent variable in the remaining analyses as the measure of inhibitory control performance.

Table 2

Descriptive statistics for continuous variables coded for the gift-wrap task for full sample and as a function of overall performance

	Full sample (N = 34)			Passed (n = 11)			Failed (n = 23)		
Measure	M	SD	Range	M	SD	Range	M	SD	Range
Total amount of time looked away from gift (s)	39.27	21.22	2.25 – 60.00	60.00	0	60.00	29.35	18.92	2.25 – 58.50
Latency to first look (s)	28.76	25.29	2.17 – 60.00	60.00	0	60.00	13.82	15.43	2.17 – 54.36

Note. Passed = looked away from gift for 60s. Higher scores indicate better performance. For both measures, the maximum possible value is 60s.

Multiple Regressions: Concurrent Predictors of Gift-Wrap Performance

Descriptive statistics for child overall intellectual ability, parent-report of self-regulation, and parent-report of effortful control are provided in Table 3. Scores were consistent with larger studies of children with WS (e.g., Greiner de Magalhães et al., 2022; Leyfer et al., 2012; Mervis & John, 2010). Pearson correlations ($\alpha = .01$) among the variables included in the regression analyses are reported in Table 4. The gift-wrap task composite was significantly correlated with DAS-II GCA, BRIEF-2 ERI T, BRIEF-2 Shift T, and CBQ Inhibitory Control.

Self-Regulation

To investigate the relations between parent report of self-regulation and child performance on the gift-wrap task, two multiple regression analyses were performed. First, we evaluated if emotion regulation, behavior regulation, overall intellectual ability, and/or CA uniquely contributed to performance-based inhibitory control as measured by the gift-wrap task composite. BRIEF-2 BRI T, BRIEF-2 ERI T, DAS-II GCA, and CA were entered as independent variables. As indicated in Table 5, the model explained 60% of the variance in gift-wrap performance. ERI T (large effect), GCA (large effect), and CA (medium effect) made significant independent contributions to the variance in the gift-wrap task composite. After controlling for BRI T, as ERI T decreased (indicating less difficulty in emotion regulation) and GCA and CA increased, performance on the gift-wrap task improved.

To determine if one or both components of emotion regulation as measured by the BRIEF-2 were uniquely related to performance-based inhibitory control, even after considering overall intellectual ability and CA, we conducted a second regression.

BRIEF-2 Emotional Control T, BRIEF-2 Shift T, DAS-II GCA, and CA were entered as independent variables. As indicated in Table 6, the model explained 64% of the variance in gift-wrap performance. Shift T (large effect) and GCA (large effect) made significant independent contributions to the variance in the gift-wrap task composite. After controlling for BRIEF-2 Emotional Control T and CA, as BRIEF-2 Shift T decreased (indicating less difficulty with flexibility) and GCA increased, performance on the gift-wrap task improved.

Effortful control

To determine if one or both of the scales of parent-report effortful control and/or overall intellectual ability were uniquely related to performance-based inhibitory control as measured by the gift-wrap task composite, a third multiple regression was performed. CBQ Inhibitory Control, CBQ Attentional Focusing, DAS-II GCA, and CA were entered as independent variables. As indicated in Table 7, the model explained 48% of the variance in gift-wrap performance. CBQ Inhibitory Control (medium effect) and GCA (medium effect) made significant independent contributions to the variance in the gift-wrap task composite. After controlling for CBQ Attentional Focusing and CA, as CBQ Inhibitory Control and GCA increased, performance on the gift-wrap task improved.

Table 3

Descriptive statistics for child intellectual ability, parent report of self-regulation, and parent report of effortful control

Measure	*M*	Mdn	*SD*	Range
DAS-II GCA	64.15	65.50	12.55	38 – 84
BRIEF-2				
Behavior Regulation Index T	65.50	66.50	9.55	41 – 82
Emotion Regulation Index T	63.12	65.00	9.48	47 – 81
Emotional Control T	62.00	62.00	10.19	40 – 82
Shift T	62.24	64.00	9.49	42 – 77
Children's Behavior Questionnaire				
Effortful Control	3.22	3.08	0.64	1.80 – 4.88
Inhibitory Control	3.51	3.46	0.75	1.69 – 5.54
Attentional Focusing	2.84	2.78	0.85	1.22 – 5.33

Note. DAS-II = Differential Ability Scales-II; GCA = General Conceptual Ability; BRIEF-2 = Behavior Rating Inventory of Executive Function-2; T = T-score.

Table 4

Bivariate correlations among gift-wrap task composite, chronological age, intellectual ability, parent report of self-regulation, and parent report of effortful control

	2	3	4	5	6	7	8	9	10
1. Gift-wrap task composite score	.41	.44*	-.32	-.59**	-.45*	-.60**	.15	.50*	-.01
2. Chronological Age		-.04	-.37	-.32	-.23	-.37	.12	.30	-.14
3. DAS-II GCA			-.10	-.03	-.03	.05	.04	.27	.07
4. BRIEF-2 BRI T				.52**	.49*	.45*	-.39	-.39	-.26
5. BRIEF-2 ERI T							-.39	-.58**	-.30
6. BRIEF-2 Emotional Control T						.57**	-.29	-.46*	-.20
7. BRIEF-2 Shift T							-.44*	-.57**	-.34
8. CBQ Effortful Control									
9. CBQ Inhibitory Control									.48*
10. CBQ Attentional Focusing									

*$p < .01$, **$p < .001$

Note. Overall Gift-wrap task composite = mean of z-scores for total duration looking away from gift and level of inhibitory control. DAS-II = Differential Ability Scales-II; GCA = General Conceptual Ability; BRIEF-2 = Behavior Rating Inventory of Executive Function-2; BRI = Behavior Regulation Index; T = T-score; ERI = Emotion Regulation Index; CBQ = Children's Behavior Questionnaire.

Table 5

Multiple regression: Predicting gift-wrap performance based on parent report of behavior regulation and emotion regulation, overall intellectual ability, and chronological age

Predictor	*B*	*t*	*p*-value	95% CI for *B*	*semi-partial r*	Cohen's f^2
Intercept	<0.01	-0.02	.987	[-0.22, 0.21]		
BRIEF-2 BRI T	0.01	0.84	.410	[-0.02, 0.04]	.10	.02
BRIEF-2 ERI T	-0.05	-3.89	.001	[-0.08, -0.02]	-.46	.51
DAS-II GCA	0.03	3.85	.001	[0.02, 0.05]	.45	.50
Chronological Age	0.41	2.35	.026	[0.05, 0.76]	.28	.18
$R^2 = .60$, adjusted $R^2 = .55$, $F(4, 29) = 10.99$, $p < .001$						

Note. BRIEF-2 = Behavior Rating Inventory of Executive Function-2; BRI = Behavior Regulation Index; T = T-score; ERI = Emotion Regulation Index DAS-II = Differential Ability Scales-II; GCA = General Conceptual Ability.

Table 6

Multiple regression: Predicting gift-wrap performance based on parent report of aspects of emotion regulation, overall intellectual ability, and chronological age

Predictor	*B*	*t*	*p*-value	95% CI for *B*	*semi-partial r*	Cohen's f^2
Intercept	<0.01	-0.01	.989	[-0.21, 0.20]		
BRIEF-2 Emotional Control T	-0.01	-0.83	.415	[-0.03, 0.01]	-.09	.02
BRIEF-2 Shift T	-0.05	-3.35	.002	[-0.07, -0.02]	-.37	.39
DAS-II GCA	0.03	4.22	<.001	[0.02, 0.05]	.47	.61
Chronological Age	0.30	1.87	.072	[-0.03, 0.64]	.21	.12
R^2 = .64, adjusted R^2 = .59, $F(4, 33) = 12.83$, $p < .001$						

Note. BRIEF-2 = Behavior Rating Inventory of Executive Function-2; T = T-score; DAS-II = Differential Ability Scales-II; GCA = General Conceptual Ability**.**

Table 7

Multiple regression: Predicting gift-wrap performance based on parent report of aspects of effortful control, overall intellectual ability, and chronological age

Predictor	*B*	*t*	*p*-value	95% CI for *B*	*semi-partial r*	Cohen's f^2
Intercept	<0.01	-0.02	.986	[-0.25, 0.24]		
CBQ Inhibitory Control	0.50	2.34	.027	[0.06, 0.94]	.31	.19
CBQ Attentional Focusing	-0.21	-1.19	.245	[-0.57, 0.15]	-.16	.05
DAS-II GCA	0.03	2.53	.017	[0.01, 0.05]	.34	.22
Chronological Age	0.37	1.80	.083	[-0.05, 0.79]	.24	.11
R^2 = .48, adjusted R^2 = .41, $F(4, 29) = 6.63$, $p < .001$						

Note. CBQ = Children's Behavior Questionnaire; DAS-II = Differential Ability Scales-II; GCA = General Conceptual Ability.

Discussion

The current study is the first to directly evaluate hot inhibitory control for children with WS. Results showed that for 6 – 8-year-olds, deficits in inhibitory control during a gift-wrap task were evident, with two-thirds of the participants unable to delay gratification for even 60 seconds. Parent-reported emotion regulation flexibility as measured by the BRIEF-2 Shift scale and overall intellectual ability contributed uniquely to individual differences in gift-delay performance, after accounting for parent-reported emotional control and CA. Similarly, parent-reported inhibitory control as measured by the CBQ and overall intellectual ability contributed uniquely to individual differences in gift-delay performance, after controlling for parent-reported attentional focusing and CA. In the remainder of the Discussion, we discuss these findings, limitations, and directions for future research.

Gift-Wrap Task Performance

Two-thirds of the participants were unable to exercise inhibitory control for the full duration of the 60-second hot gift-wrap task. At the same time, 83% of the children who peeked were later able to disengage and reorient away from the gift at least temporarily, thus demonstrating some inhibitory control. Across the full sample, a wide range of performance was observed; children followed the instruction not to look at the gift for between 2s and 60s of the 60-second task. The mean latency to first peak (including the children who never peaked) was 29 seconds. This is similar to the previously-reported performance of an older sample of children and adolescents with DS (Fontana et al., 2021). However, the present participants performed considerably worse than the TD participants in that study, even though the TD group was slightly younger,

providing further evidence of delay in the development of inhibitory control by children with WS.

Gift-Wrap Task Performance: Relations with Intellectual Ability and Parent-Reported Self-Regulation

Performance on the gift-wrap task was positively related to overall intellectual ability. This finding is in line with results from a recent study indicating that intellectual ability is positively related to performance on directly-assessed measures of hot inhibitory control for adults with Prader-Willi syndrome and adults with mild ID without a known genetic cause (Chevalère et al., 2022). Similar findings also have been reported for both preschool-aged TD children from low-income families (Allen & Lewis, 2020) and TD adults (Shamosh & Gray, 2008).

Performance on the gift-wrap task also was positively associated with both components of emotion regulation that were measured by parent report (flexible emotion regulation ability and emotional control ability). These results are consistent with prior findings for both individuals with WS and TD children. Camp et al. (2016) found that for children and young adults with WS, both parent-reported flexible emotion regulation and parent-reported emotional control were positively correlated with practical problem-solving in novel situations. Carlson and Wang (2007) found that parent report of emotional control was related to the performance of TD preschoolers and young school age children on a battery of inhibitory control measures that included a gift-wrap task. (Flexible emotion regulation was not evaluated in that study.) We also detected differential relations between gift-wrap performance and ratings on the effortful control subdomains measured by the CBQ: Performance on the gift-wrap task was significantly

positively correlated with parent report of inhibitory control but was not significantly associated with parent-reported attentional focusing. Gudorf et al. (2011) reported similar findings for TD children.

Concurrent Predictors of Gift-Wrap Task Performance

The current study was the first to consider the concurrent effects of parent-reported aspects of self-regulation and overall intellectual ability on performance on the gift-wrap inhibitory control task. We found that parent-reported emotion regulation – but not parent-reported behavior regulation – overall intellectual ability, and, to a lesser extent age, contributed uniquely to individual differences in gift-wrap performance. This pattern of findings, in concordance with prior findings from studies of TD children, provides further support for the theoretical expectation that hot inhibitory control is facilitated primarily by emotion regulation rather than behavior regulation (Carlson & Wang, 2007; see Luerssen & Ayduk, 2014 for review; Zelazo & Cunningham, 2007).

Findings from a further analysis in which we considered the concurrent effects of the two components of emotion regulation measured by the BRIEF-2 on hot inhibitory control indicated that flexible emotion regulation – but not emotional control – in conjunction with overall intellectual ability, contributed a significant amount of unique variance to individual differences in gift-wrap performance, even after controlling for CA. In line with previous explanations of individual differences among TD children in overcoming prepotent responses in emotionally-salient contexts (Luerssen & Ayduk, 2014), our results suggest that children who are better at shifting their focus of attention away from a reward for which they need to wait are better able to implement effective regulation and problem-solving strategies to delay gratification.

A parallel analysis focused on the two components of effortful control as measured by parent-report on the CBQ indicated that inhibitory control – but not attentional focusing – and overall intellectual ability contributed uniquely to individual differences in gift-wrap performance, even after controlling for age. These findings provide empirical evidence supporting the theoretical argument that the inhibitory control component of effortful control is utilized in delaying gratification (e.g., Eisenberg et al., 2014).

Limitations and Future Directions

As this study is the first to use a performance-based measure to directly evaluate hot inhibitory control in individuals with WS, the current sample was intentionally restricted to young school-aged children. The sample size, although relatively small, is substantial given the rarity of WS, the narrow (25-month) age range from 72 – 96 months, and the necessity to end data collection due to the COVID pandemic. However, despite efforts to recruit diverse participants, the final sample was predominantly White non-Hispanic. To identify the onset and development of hot inhibitory control in children with WS, longitudinal studies with more diverse samples in which multiple performance-based measures of both hot and cool inhibitory control are directly assessed throughout childhood would be of value. Investigating if hot and cool inhibitory control differentially impact mastery motivation, academic achievement, and adaptive functioning may provide a better understanding of the underpinnings of the heterogeneity among children with WS that is evident in these domains.

Conclusion

For young school-aged children with WS, deficits in hot (affective) inhibitory control on the gift-wrap task were evident. At the same time, the ability to delay gratification by inhibiting the urge to peek when faced with an enticing reward was characterized by considerable variability. In line with the literature on TD children, the ability to inhibit the urge to access an enticing reward was related to regulation of emotions and inhibitory control. Furthermore, both overall intellectual ability and parental ratings of flexible emotion regulation or the inhibitory control component of effortful control accounted for unique variance in individual differences in performance on the gift-wrap task. These findings indicate that the gift-wrap task is emotionally and motivationally significant for young school-aged children with WS and is an appropriate hot performance-based measure of their delay of gratification and inhibitory control abilities.

CHAPTER III

INDIVIDUAL DIFFERENCES IN ADAPTIVE FUNCTIONING AMONG CHILDREN WITH WILLIAMS SYNDROME: CONTRIBUTIONS OF EMOTION-RELATED SELF-REGULATION AND OVERALL INTELLECTUAL ABILITY[2]

When children enter elementary school, demands and expectations increase both at home and at school (McClelland & Cameron, 2012). For example, children are expected to persist to learn new concepts and skills that they find challenging, stay focused on a task specified by an adult rather than doing a preferred activity, engage in cooperative play with others, and overcome impulses to comply with social norms. Self-regulation – the ability to regulate emotions, actions, attention, or thoughts purposefully or automatically – is foundational for navigating these increasing complexities in their daily lives (Baumeister et al., 2007; Eisenberg & Zhou, 2016; Karoly, 1993).

There is increasing evidence that children who have genetic syndromes associated with intellectual disability (ID) have impaired self-regulatory skills, as indicated by difficulties with emotion regulation, impulsivity, and other behavioral challenges (see Shaffer et al., in press for review; Cuskelly et al., 2013). Difficulties in adaptive functioning, including socialization, communication, and daily living skills, also are common (Brawn & Porter, 2018; Hatton et al., 2003; Schworer et al., 2022). These commonalities across syndromes suggest that both limited intellectual ability and difficulties with self-regulation may negatively impact adaptive functioning.

The goal of the present study was to examine the contributions of emotion regulation, behavior regulation, and overall intellectual ability to individual differences in

[2] This chapter was prepared for submission to the *Journal of Autism and Developmental Disorders*.

adaptive skills in young school-aged children with Williams syndrome (WS), a genetic disorder caused by a deletion of 25 – 27 genes on chromosome 7q11.23 (Kozel et al., 2021).

Self-Regulation

In the research literature, the terms "self-regulation," "executive function," and "effortful control" are often used interchangeably (Diamond, 2013; Nigg, 2000, 2017; Zhou et al., 2012). In the current study, we use "self-regulation" as an umbrella construct to describe the processes by which an individual purposefully or automatically regulates his or her actions, attention, emotions, or thoughts (see Eisenberg & Zhou, 2016; Karoly, 1993). Self-regulation can be divided into emotional (hot) and non-emotional (cool) aspects (Willoughby et al., 2011; Zelazo & Müller, 2010). A key distinction between these interrelated aspects is the relative involvement of the regulation of emotion and motivation: Emotion regulation predominates in situations when motivation and emotions are higher, whereas unemotional (behavior/cognitive) regulation predominates in emotionally neutral circumstances (Zelazo & Carlson, 2012; Zelazo & Müller, 2010).

Self-regulation can be assessed using either performance-based lab measures or parent rating scales (Diamond, 2013). Performance-based lab measures directly and objectively assess self-regulation in an optimal, controlled environment (Isquith et al., 2013; Toplak et al., 2013). The researcher determines the goal of the task and tells the child what is expected from him or her. Then, how well the child performs is measured. Direct assessment of self-regulatory skills often includes performance-based measures of inhibitory control – the ability to overcome prepotent responses [e.g., actions, attention, and/or emotions (Garon et al., 2008; Miyake et al., 2000)]. Within the context of self-

regulation, research findings (including results of factor analyses) indicate that inhibitory control is best represented by hot (emotional) and cool (non-emotional) subdomains (Backer-Grøndahl et al., 2019; Bassett et al., 2012; Brock et al., 2009; Eisenberg & Zhou, 2016; Grazyna Kochanska & Kim, 2013; Mulder et al., 2014; Zelazo & Müller, 2010).

For example, successful performance in a delay paradigm requires impulse control to overcome the temptation for immediate gratification and instead to wait for a reward, such as a gift (Kim et al., 2013). This type of task is commonly used as a performance-based measure of hot inhibitory control, as the regulation of emotions assists in problem-solving to overcome the desire to peek at the gift (Allan et al., 2014; Carlson & Wang, 2007). In contrast, Stroop-like tasks involve interference control, or inhibition of attention to distractors and prepotent responses in an emotionally neutral context and are frequently used as a performance-based measure of cool inhibitory control (Diamond, 2013, 2016; Liu et al., 2015). Verbal Stroop-like tasks (similar to verbal opposites tasks) require the participant to overcome a verbal prepotent response and replace it with the response specified by the researcher (Diamond, 2013).

During the preschool and early school years, typically developing (TD) children evidence rapid improvement in inhibitory control as indicated by performance-based measures. For example, during the gift-task, 42% of young 3-year-olds but 74% of 5-year-olds never peeked during the gift-wrapping process (Carlson, 2005). On the classic Day/Night Stroop-like task, mean percentage of correct responses was 69% for young 4-year-olds but 87% for 6-year-olds (Gerstadt et al., 1994; Montgomery & Koeltzow, 2010).

Parent report measures capture the parent's perspective on the child's self-regulation abilities during daily life (Toplak et al., 2013). These ratings characterize the child's usual behaviors and emotions across a wide range of situations in which the child does not have support and has his/her own goal in mind (Toplak et al., 2013). As indicated by Zelazo et al. (2016), the most commonly used executive function questionnaires are the various forms of the Behavioral Rating Inventory of Executive Function (BRIEF; Gioia et al., 2000, 2002, 2015; Roth et al., 2005). The BRIEF-2 (Gioia et al., 2015) parent questionnaire yields an Emotional Regulation Index (ERI) which assesses a child's ability to modulate emotional responses and to move flexibly from one facet of a problem, task, or situation to another and includes two scales: Emotional Control and Shift. The Behavior Regulation Index (BRI) assesses a child's ability to monitor one's own behaviors, stop behaviors, and act appropriately and includes two scales: Self-Monitor and Inhibit. The distinction between emotion regulation and behavior regulation has been empirically supported by a factor analysis conducted for a large independent neuropediatric sample (Halvorsen et al., 2019).

Toplak et al. (2013) conducted a review of 13 studies that reported associations between performance-based measures (all of which assessed cool self-regulation) and informant ratings on the BRIEF. The resulting correlations ranged from very weak to moderate, and only 19% were statistically significant, with an average effect size of .15. Similar findings were reported by Esbensen and Hoffman (2018) for children with Down syndrome (DS) when the relations between performance-based tasks of inhibitory control and the age-appropriate version of the BRIEF were considered. To the best of our knowledge, no published study has evaluated the relation between a delay performance-

based task and parent ratings on the age-appropriate BRIEF in either school-aged TD children or children with ID. The low correlations between parent report measures of everyday skills and performance-based measures administered in a research lab may be explained by differences in the type of skills assessed and in task demands (Isquith et al., 2013). Although similar processes may be involved in self-regulation in the real-world settings that are assessed in parent-report measures and the controlled laboratory setting, the constructs assessed in parent-report measures and direct assessments are complementary, not identical (Isquith et al., 2013; Toplak et al., 2013).

Adaptive Function and Self-Regulation

Adaptive skills include the ability to function in social situations, exchange information with others, and perform tasks of daily living (Sparrow et al., 2016). Conceptually, being able to regulate one's own behaviors and emotions appropriately allows an individual to better navigate the demands and complexities of societal norms in day-to-day life (Baumeister et al., 2007). Empirically, findings from a recent meta-analysis indicated that self-regulation measured when children were in preschool was positively associated with social competence, academic achievement, and school engagement and negatively associated with externalizing problems, internalizing problems, and peer victimization several years later. The relation with early self-regulation extended into adulthood. Children who had lower self-regulatory skills were more likely to experience depression, anxiety, and unemployment as adults (Robson et al., 2020).

For TD preschool and young school-aged children, hot and cool aspects of self-regulation have been shown to differentially impact functional outcomes (Allan et al.,

2014; Backer-Grøndahl et al., 2019; Bassett et al., 2012; Brock et al., 2009; Di Norcia et al., 2015; Kim et al., 2013; Mulder et al., 2014). For example, teacher-reported social competence (i.e., sensitivity and cooperation with others) was uniquely predicted by children's hot inhibitory control latent factor score derived from performance on a battery of direct assessments, but not by the cool latent factor (Bassett et al., 2012). In contrast, for a large sample of kindergarteners, overall intellectual ability was moderately related to performance on the cool inhibitory control factor score but only weakly correlated with the hot inhibitory control factor score (Brock et al., 2009).

Sabat et al. (2020) considered the relations between adaptive behavior and a cool performance-based measure of inhibitory control for adolescents with DS and found a significant and large positive correlation between teacher-reported conceptual (communication, functional academic, and self-direction) skills and performance on a spatial interference control task. For parent-reported conceptual skills, the correlation was moderate and positive, although not statistically significant. Relations between adaptive skills and parent ratings of emotion regulation and behavior regulation for children and adolescents with DS also have been reported. Both BRIEF-2 ERI T and BRI T were moderately and significantly correlated with parent-reported socialization, communication, and daily living skills (Onnivello et al., 2022).

Williams Syndrome

WS is a rare neurodevelopmental disorder caused by a hemideletion of 25 – 27 genes on chromosome 7q11.23 (Kozel et al., 2021). The prevalence of WS is approximately 1 in 7,500 births (Strømme et al., 2002), occurring at the same frequency for males and females (Morris et al., 2020). WS is characterized by specific personality,

behavioral, and cognitive profiles (see Mervis & Greiner de Magalhães, 2022 for review). For example, children with WS are described as indiscriminately friendly, socially disinhibited, more inclined to inappropriately approach strangers than TD children or children with ID of other etiologies, gregarious, and having low tolerance for frustration and low persistence (Davies et al., 1998; Dodd et al., 2010; Jones et al., 2000; Klein-Tasman & Mervis, 2003; Mervis & Klein-Tasman, 2000; Zitzer-Comfort et al., 2007; Rowe, 2007). On average, individuals with WS have mild to moderate ID with the full range of abilities extending from severe ID to average relative to the general population (Martens et al., 2008; Mervis & Greiner de Magalhães, 2022).

As indicated by both direct assessment and parent ratings, children with WS evidence considerable difficulty with self-regulatory skills. The results of all four published studies that used a structured lab-based measure to assess verbal inhibitory control during emotionally neutral, cool tasks in children with WS indicated that performance was significantly worse than that of same-aged TD children (Atkinson & Braddick, 2011; Breckenridge et al., 2013; Carney et al., 2013; Tager-Flusberg et al., 1997). In the one study that controlled for both chronological and mental age, the children with WS performed significantly worse than considerably younger TD children (Carney et al., 2013). None of these studies addressed relations between task performance and either overall intellectual ability or parent-report of emotion or behavior regulation.

To the best of our knowledge, only one study has used a structured lab-based measure to assess inhibitory control during an emotionally salient task in children with WS (Pitts & Mervis, under review). In this study, which used a gift-wrap delay of gratification task with 6 – 8-year-olds, deficits in inhibitory control were evident. Only

32% of the participants were able to delay gratification (refrain from peeking) for the entire 60-second task. The level of inhibitory control shown by children with WS on the task was similar to that previously reported for an older sample of children and adolescents with DS (Fontana et al., 2021) but considerably more limited than that previously reported for TD 6-year-olds (Carlson, 2005; Fontana et al., 2021).

Parental ratings have also indicated that deficits in regulating emotion and actions during everyday conditions are common among children with WS (Camp et al., 2016; Greiner de Magalhães et al., 2022; Phillips, 2008; Woodruff-Borden et al., 2010). For example, based on parent report on the BRIEF-2, 52% of a large sample of 6 – 17-year-olds with WS met the test authors' criteria for either potentially clinically elevated or clinically elevated impairment for emotion regulation and 43% met these criteria for behavior regulation (Greiner de Magalhães et al., 2022). Higher intellectual ability was associated with better performance on both the Shift scale and the Self-monitor scale. When compared to similarly aged individuals with DS matched for nonverbal ability, adolescents and young adults with WS performed worse on all of the BRIEF scales (Camp et al., 2016).

To our knowledge, only two studies have been reported that evaluated the relation between lab measures of inhibitory control and parent ratings of emotion regulation or behavior regulation for individuals with WS. For young-school aged children, individual differences in performance on the gift-task were significantly related to both overall intellectual ability and BRIEF-2 ERI but not to BRIEF-2 BRI (Pitts & Mervis, under review), suggesting that the ability to inhibit the urge to access an enticing reward may be facilitated by everyday emotion regulation (cf. Holodynski et al., 2013; Zelazo &

Cunningham, 2007). For a small sample of adults with WS, parent ratings on the BRIEF-Adult BRI (which includes both behavior regulation and emotion regulation scales) were moderately associated with performance on the inhibit condition of the Shape School test, although the correlation was not statistically significant (Hocking et al., 2015).

In addition to deficits in self-regulation, the adaptive skills of children with WS are below age-expectations. On average, the overall level of adaptive skills is in the mild to moderate disability range, with abilities extending from severe disability to average relative to the general population (see Brawn & Porter, 2018 for systematic review). At the group level, children with WS exhibit relative strengths in socialization and communication skills and a relative weakness in daily living skills. However, variability in the pattern of relative strengths and weaknesses shown by individual children has been reported.

Two studies reported correlations between overall adaptive functioning and intellectual ability for children and adolescents with WS. Results indicated that better adaptive functioning was significantly associated with higher intellectual ability (Dimitropoulos et al., 2009; Mervis et al., 2001). Within adaptive behavior domains, intellectual ability was significantly correlated with Communication and Daily Living Skills but not with Socialization skills. Two studies which focused on adults with WS and included larger sample sizes reported significant correlations between overall intellectual ability and each of the three adaptive behavior domains (Davies et al., 1998; Howlin et al., 2010). The mixed findings for socialization skills may be due to differences in sample sizes, measures, and/or age.

A few studies have evaluated the relation between parent report of self-regulation and adaptive function in individuals with WS. Phillips (2008), based on a relatively small sample of 8 – 15-year-olds with WS, found that the bivariate correlations between emotion regulation skills and personal living skills, community living skills, and social interaction and communication skills were all statistically significant. However, after controlling for overall intellectual ability, only the relation between emotion regulation skills and personal living skills remained significant. Greiner de Magalhães et al. (2022), using a considerably larger sample of 6 – 17-year-olds with WS, found that parental ratings on the BRIEF-2 ERI and BRI were each significantly correlated with personal living skills, social interaction and communication skills, and community living skills. Relations controlling for overall intellectual ability were not reported. For a small sample of 5 – 37-year-olds with WS, deficits in social interactions were found to be significantly and independently correlated with parent reported difficulty with emotion regulation and behavior regulation (Ng-Cordell et al., 2018). Even after controlling for structural language ability, chronological age, sex, and nonverbal reasoning ability, a composite measure of the BRIEF-2 ERI and BRI accounted for a significant amount of unique variance in the pragmatic communication abilities of 6 – 15-year-olds with WS (Harmon, 2020).

Current Study

Children with WS have deficits in self-regulation as indicated by both parent report on the regulation of emotions and behaviors (Greiner de Magalhães et al., 2022; Phillips, 2008; Woodruff-Borden et al., 2010) and direct assessment of inhibitory control during emotional or unemotional contexts (Atkinson et al., 2003; Breckenridge et al.,

2013; Carney et al., 2013, Pitts & Mervis, under review; Tager-Flusberg et al., 1997). Difficulties in adaptive functioning are also common, with children performing worse than same-aged TD peers on standardized assessments of socialization, communication, and daily living skills (Brawn & Porter, 2018). Although higher intellectual ability has been shown to be associated with better adaptive functioning (Brawn & Porter, 2018), intellectual ability alone is unlikely to fully account for the within-syndrome variability evidenced in adaptive skills. It is possible that emotion regulation ability and/or behavior regulation ability also contribute to individual differences in adaptive behavior. The present study provides the first empirical evaluation of the *concurrent* effects of the ability to regulate emotions, the ability to regulate behaviors, and overall intellectual ability on adaptive skills in children with WS. We addressed two research questions:

1) Does performance on lab-based measures of hot or cool inhibitory control and/or directly assessed overall intellectual ability contribute uniquely to individual differences in the socialization, communication, and/or daily living skills domains of adaptive functioning?
2) Do parental ratings of everyday emotion regulation or behavior regulation and/or directly assessed overall intellectual ability contribute uniquely to individual differences in the socialization, communication, and/or daily living skills domains of adaptive functioning?

Method

Participants

The final sample included 34 children (18 girls, 16 boys) with genetically confirmed classic-length WS deletions aged 6.01 – 8.05 years (M = 6.88

years, *SD* = 0.68). The participants lived in 23 different U. S. states (representing all U. S. census regions: 17.6% Northeast, 44.1% South, 23.5% Midwest, 8.8% West) and one Canadian province (5.9%). The native language for all participants was English. The distribution of participants' racial/ethnic background was: 94.1% White non-Hispanic, 2.9% White Hispanic, and 2.9% multiracial non-Hispanic. Three additional children with genetically confirmed classic WS deletions were excluded because they did not have enough language to understand the instructions for the lab-based measures of inhibitory control. Data collection began in July 2017 and ended in February 2020. The participants in the current sample were also included in Pitts and Mervis (under review).

Measures

Hot Gift-wrap Delay of Gratification Task

Hot inhibitory control was directly assessed using a gift-wrap task which taps into response inhibition and delay of gratification (Kochanska et al., 1996). The researcher instructed the child to remain seated and not look or peek while the researcher wrapped the gift, so it remained a "big surprise." Then, the researcher loudly wrapped the gift behind the child and out of the child's sight for approximately 60 seconds.

The task was filmed, and behavioral coding based on the recording was conducted using Behavioral Observation Research Interactive Software (BORIS; Friard & Gamba, 2016). Behaviors were coded from the videotapes by a research assistant, and 21% (n = 7) of the videotapes were independently coded by the first author. Interrater agreement was excellent for both the categorical and continuous measures (rs $\geq$ .99).

To characterize inhibitory control, the total number of seconds (out of 60 possible) the child looked away from the gift was determined. In addition, a rating on a 4-

point categorical inhibitory control rating was assigned (0 = turned around to look at gift and stood up; 1 = turned around to look at gift but remained seated; 2 = looked over shoulder at gift but remained seated; 3 = never attempted to look at gift and remained seated). For both measures, higher scores indicated higher inhibition and better performance. A gift-task composite score was computed by averaging the *z*-scores for the total duration a child looked away from the gift and the level of inhibitory control (for more detail see Pitts & Mervis, under review).

Cool Car/Duck Stroop-like Interference Control Task

Cool interference control was directly assessed using a modified version of the Day/Night Stroop-like task (Gerstadt et al., 1994). The goal of the Car/Duck task was the same as the original Day/Night task—to inhibit an automatic verbal response and replace it with an alternate verbal response. The child was shown cards with a picture of a duck or a car and was instructed to say "quack" when shown the car and "vroom" when shown the duck. These stimuli were selected because pilot testing confirmed that participants would be familiar with the standard association of the stimuli and spontaneously produce the sounds of a car and a duck without needing teaching or prompting.[3] The automatic stimulus response was the sound associated with the stimulus (duck-quack; car-vroom); thus, the automatic response would need to be suppressed in order to respond correctly on the interference task.

[3] The Grass/Snow Stroop-like task (Carlson & Moses, 2001) was piloted. In this task, the child was instructed to point to a green square when the examiner said "snow" and to point to a white square when the examiner said "grass." However, many children did not readily provide the color association and had difficulty understanding the task directions even after teaching and readministering the practice trials. Thus, the Grass/Snow data were not useable as a measure of inhibition for the current participants.

Before beginning the experimental trials, the researcher showed the child a card with a black and white drawing of a duck and asked the child to tell her what a duck says. The researcher then picked up the card with a black and white drawing of a car and asked the child to tell her what a car says. This was repeated once more for each card with feedback and reinforcement. To continue, the child needed to correctly produce the sound of a car (“vroom”) and a duck ("quack”). Then the interference control paradigm was introduced, and four practice trials (two ducks, two cars) were administered. The child was instructed to say “quack” when shown a picture of a car and “vroom” when shown a picture of a duck. To ensure the child understood the task, feedback and reinforcement were provided after each trial.

Next, 16 experimental trials (eight ducks, eight cars) were administered. To reduce the demand on working memory, before each trial the examiner repeated the task rule, “When you see the duck, say ‘vroom.’ When you see the car, say ‘quack.’” No feedback was given for the experimental trials. Trials were presented in the same pseudo-random order for each participant. The proportion of correct responses was determined.

Parent Report of Emotion Regulation and Behavior Regulation

The Behavior Rating Inventory of Executive Function-2 (BRIEF-2; Gioia et al., 2015) is a parent-report questionnaire which evaluates aspects of executive functioning. Using a 3-point scale, parents indicate whether each of 63 behaviors has never (0), sometimes (1), or often (2) been a problem during the past six months. The current study focused on the Emotion Regulation Index (ERI) and the Behavior Regulation Index (BRI). The ERI assesses a child’s ability to modulate emotional responses and to move flexibly from one facet of a problem, task, or situation to another. The BRI assesses a

child's ability to monitor one's own behaviors, stop behaviors, and act appropriately. For each index and scale, T-scores are provided (M = 50, SD = 10 for the general population, range: 35 – 90). Higher scores are associated with greater difficulty. For the ERI and BRI, internal consistency ranged from .94 – .96 for the normative and clinical sample.

Intellectual Ability

The Differential Ability Scales-II, Early Years (DAS-II; Elliott, 2007) is a standardized assessment of intellectual abilities. The General Conceptual Ability (GCA; similar to Full-Scale IQ) is a composite of verbal, nonverbal reasoning, and spatial abilities. Standard scores (SSs) are provided (M = 100, SD = 15 for the general population, range: ~30 – 170). For children aged 6 – 8 years in the normative sample, reliability coefficients ranged from .95 – .97.

Adaptive Functioning

The Vineland Adaptive Behavior Scales-3, Comprehensive Interview Form (VABS-3; Sparrow et al., 2016) is a standardized, semi-structured parent interview measuring adaptive skills. The VABS-3 uses a 3-point scale to indicate whether the child performs a behavior never (0), sometimes (1), or usually (2) without needing help or prompting. This study focused on three domains of adaptive skills. The Socialization domain measures functioning in social situations (social appropriateness, engaging in play with others, expressing and controlling emotions in situations involving others). The Communication domain assesses how well a person exchanges information with others (listening and understanding; expressing self through spoken or signed language; reading and writing). The Daily Living Skills domain measures performance of practical, everyday tasks of daily living (e.g., eating, personal hygiene, basic safety, and household

chores). SSs are provided (M = 100, SD = 15 for the general population, range: 20 – 160). For children aged 6 – 8 years in the normative sample, internal consistency coefficients ranged from .96 – .97 for the three domains.

Procedure

The university's Institutional Review Board reviewed and approved the study protocol. Parents or legal guardians of all participants provided written informed consent and children provided oral assent. Parent questionnaires were mailed approximately two weeks before the child's assessment and collected upon arrival to the laboratory. The child's mother completed the BRIEF-2 parent questionnaire. The mother (or both parents together) completed the VABS-3 interview with the senior author while the child was participating in the study.

The DAS-II and inhibitory control tasks were administered to the child as part of the protocol for a larger study of language and cognitive development in children with WS. The DAS-II was administered in a quiet testing room following standardized procedures. After completing the standardized assessments, the inhibitory control tasks were conducted in a different testing room which had two remote-controlled video cameras concealed in wooden cabinets with clear glass windows in diagonally opposed corners of the room. The tasks were filmed by a research assistant who operated the video cameras from a control room located behind a one-way mirror.

Statistical Analyses

Data were analyzed using IBM SPSS 28. Descriptive statistics and bivariate correlations (α = .01) were computed for performance-based inhibitory control, parent-report of emotion and behavior regulation, child overall intellectual ability, and parent-

report of adaptive skills. To identify significant concurrent predictors of components of children's adaptive skills, six multiple regression analyses were performed, two for each of the three primary adaptive behavior domains included in the Vineland-3: Socialization, Communication, and Daily Living Skills. The three performance-based regressions included gift-task, Stroop-like task, and overall intellectual ability as predictors. The three parental rating-based regressions included ERI T, BRI T, and overall intellectual ability as predictors. Chronological age was not significantly correlated with any of these variables and therefore was not included in the regression analyses.

Independent variables were converted to a standard normal distribution (i.e., transformed to *z*-scores) and thus were centered on the sample mean, as recommended by Osborne (2017). All assumptions of multiple linear regression analyses were met. Cohen's f^2 was used to characterize the effect sizes of the predictors (0.02 = small, 0.15 = medium, 0.35 = large; Cohen, 1988).

Results

Descriptives and Correlations

Descriptive statistics for performance-based hot and cool inhibitory control, parent-report of behavior regulation and emotion regulation, overall intellectual ability, and adaptive skills are provided in Table 8. Scores were consistent with those from larger studies of children with WS (Brawn & Porter, 2018; Greiner de Magalhães et al., 2022; Mervis & John, 2010).

Pearson correlations ($\alpha = .01$) among the variables included in the regression analyses are reported in Table 9. Of particular interest were the relations between child performance on hot and cool inhibitory control tasks, parental ratings of everyday

emotion regulation and behavior regulation, and overall intellectual ability. Child performance on the gift-task and the Stroop-like task was moderately correlated although the correlation value was not statistically significant. GCA was significantly related to both performance-based measures of inhibitory control but not to parent ratings of emotion regulation or behavior regulation. As indicated in Pitts and Mervis (under review), the two measures of hot emotion regulation (gift-task and ERI T) were significantly related. However, the two measures of cool behavior regulation (Stroop-like task and BRI T) were not.

Table 8

Descriptive statistics for performance-based inhibitory control, parent-report of emotion and behavior regulation, overall intellectual ability, and adaptive functioning measures

Measure	*M*	Mdn	*SD*	Range
Gift-wrap task composite score	0	-0.31	0.91	-1.57 – 1.09
Stroop-like task proportion correct	0.69	0.75	0.29	0 – 1.00
BRIEF-2 Emotion Regulation Index T	63.12	65.00	9.48	47 – 81
BRIEF-2 Behavior Regulation Index T	65.50	66.50	9.55	41 – 82
DAS-II GCA	64.15	65.50	12.55	38 – 84
VABS-3 Socialization SS	70.29	70.50	8.59	46 – 90
VABS-3 Communication SS	69.00	70.00	9.05	40 – 85
VABS-3 Daily Living Skills SS	64.53	65.00	6.90	47 – 82

Note. BRIEF-2 = Behavior Rating Inventory of Executive Function-2; T = T-score. DAS-II = Differential Ability Scales-II; GCA = General Conceptual Ability; VABS-3 = Vineland Adaptive Behavior Scales-3; SS = standard score.

Table 9

Bivariate correlations among performance-based measures of inhibitory control, parent report of emotion and behavior regulation, overall intellectual ability, and adaptive functioning

	2	3	4	5	6	7	8
1. Gift-wrap task composite score	.40^{+}	-.59**	-.32	.44*	.53*	.29	.46*
2. Stroop-like task proportion correct		-.22	-.22	.56**	.50*	.60**	.43
3. BRIEF-2 Emotion Regulation Index T			.52*	-.03	-.49*	-.24	-.46*
4. BRIEF-2 Behavior Regulation Index T				-.10	-.33	-.28	-.32
5. DAS-II GCA					.51*	.64**	.50*
6. VABS-3 Socialization SS						.73**	.90**
7. VABS-3 Communication SS							.75**
8. VABS-3 Daily Living Skills SS							

*p < .01, **p <.001

Note. BRIEF-2 = Behavior Rating Inventory of Executive Function-2; T = T-score; DAS-II = Differential Ability Scales-II; GCA = General Conceptual Ability; SS = standard score; VABS-3 = Vineland Adaptive Behavior Scales-3.

Concurrent Predictors of Adaptive Functioning

To investigate the relations of performance-based structured inhibitory control tasks and overall intellectual ability to socialization, communication, and daily living skills, three multiple regression analyses were conducted (Table 10). The model for Socialization SS explained 42% of the variance among children. Gift-task performance (medium effect) was the only independent variable that made a significant unique contribution. After controlling for Stroop-like task performance and GCA, as performance on the gift-task improved, Socialization SS increased. The model for Communication SS explained 50% of the variance among children. Stroop-like task performance (medium effect) and GCA (medium effect) made significant unique contributions. After controlling for gift-task performance, as performance on the Stroop-like task improved and GCA increased, communication skills increased. Although the model for Daily Living Skills SS explained a significant amount of the variance (34%) among participants, none of the independent variables made a significant independent contribution to this variance.

To investigate the relations of parent-reported everyday emotion regulation and behavior regulation and directly measured overall intellectual ability with socialization, communication, and daily living skills, three additional multiple regression analyses were conducted (Table 11). The model for Socialization SS explained 49% of the variance among children. ERI T (medium effect) and GCA (large effect) made significant unique contributions. After controlling for BRI T, as ERI T decreased (indicating less impairment) and as GCA increased, socialization skills increased. The model for Communication SS explained 48% of the variance among children. GCA (large effect)

was the only independent variable that made a significant unique contribution to the variance among children.[4] After controlling for ERI T and BRI T, as GCA increased communication skills increased. The model for Daily Living Skills SS explained 45% of the variance among children. ERI T (medium effect) and GCA (large effect) made significant unique contributions to the variance. After controlling for BRI T, as ERI T decreased (indicating less impairment) and as GCA increased, daily living skills increased.

[4] The pattern of findings is the same if the DAS-II Special Nonverbal Composite (SNC), which is based on performance on the nonverbal reasoning and spatial subtests but not the verbal subtests, is included as the measure of intellectual ability instead of GCA.

Table 10

Multiple regressions: Predicting adaptive functioning based on performance-based measures of inhibitory control and overall intellectual ability

Predictor	*B*	*t*	*p*-value	95% CI for *B*	*semi-partial r*	Cohen's f^2
Predicting VABS-3 Socialization SS						
Intercept	70.29	59.48	<.001	[67.88, 72.71]		
Gift-wrap task composite score	3.19	2.13	.042	[0.13, 6.24]	.30	0.15
Stroop-like task proportion correct	1.98	1.34	.190	[-1.04, 4.99]	.19	0.06
DAS-II GCA	2.01	1.33	.193	[-1.07, 5.09]	.19	0.06
R^2 = .42, adjusted, R^2 = .36, $F(3, 30) = 7.09$, $p < .001$						
Predicting VABS-3 Communication SS						
Intercept	69.00	60.04	<.001	[66.65, 71.35]		
Gift-wrap task composite score	-0.58	-0.40	.695	[-3.55, 2.40]	-.05	0.01
Stroop-like task proportion correct	3.29	2.29	.029	[0.36, 6.22]	.30	0.17
DAS-II GCA	4.23	2.88	.007	[1.23, 7.22]	.37	0.28
R^2 = .50, adjusted, R^2 = .45, $F(3, 30) = 10.08$, $p < .001$						
Predicting VABS-3 Daily Living Skills SS						
Intercept	64.53	63.88	<.001	[62.47, 66.59]		
Gift-wrap task composite score	2.04	1.59	.122	[-0.58, 4.65]	.24	0.08
Stroop-like task proportion correct	1.10	0.87	.391	[-1.48, 3.68]	.13	0.03
DAS-II GCA	2.01	1.56	.130	[-0.62, 4.64]	.23	0.08
R^2 = .34, adjusted, R^2 = .27, $F(3, 30) = 5.11$, $p = .006$						

Note. VABS-3 = Vineland Adaptive Behavior Scales-3; SS = standard score; DAS-II = Differential Ability Scales-II; GCA = General Conceptual Ability.

Table 11

Multiple regressions: Predicting adaptive functioning based on parent report of behavior regulation and emotion regulation and overall intellectual ability

Predictor	*B*	*t*	*p*-value	95% CI for *B*	*semi-partial r*	Cohen's f^2
Predicting VABS-3 Socialization SS						
Intercept	70.29	63.69	<.001	[68.04, 72.55]		
BRIEF-2 ERI T	-3.91	-2.97	.006	[-6.60, -1.22]	-.39	0.29
BRIEF-2 BRI T	-0.34	-0.25	.802	[-3.04, 2.37]	-.03	<0.01
DAS-II GCA	4.26	3.79	.001	[1.96, 6.57]	.49	0.48
R^2 = .49, adjusted, R^2 = .44, $F(3, 30) = 9.60$, $p < .001$						
Predicting VABS-3 Communication SS						
Intercept	69.00	58.69	<.001	[66.60, 71.40]		
BRIEF-2 ERI T	-1.44	-1.03	.311	[-4.32, 1.42]	-.14	0.04
BRIEF-2 BRI T	-1.19	-0.84	.407	[-4.06, 1.69]	-.11	0.02
DAS-II GCA	5.67	4.73	<.001	[3.22, 8.12]	.62	0.75
R^2 = .48, adjusted, R^2 = .43, $F(3, 30) = 9.18$, $p < .001$						
Predicting VABS-3 Daily Living Skills SS						
Intercept	64.53	69.95	<.001	[62.65, 66.41]		
BRIEF-2 ERI T	-2.87	-2.61	.014	[-5.12, -0.62]	-.35	0.23
BRIEF-2 BRI T	-0.37	-0.34	.738	[-2.63, 1.88]	-.05	<0.01
DAS-II GCA	3.33	3.54	.001	[1.41, 5.26]	.48	0.42
R^2 = .45, adjusted, R^2 = .39, $F(3, 30) = 8.12$, $p < .001$						

Note. VABS-3 = Vineland Adaptive Behavior Scales-3; SS = standard score; DAS-II = Differential Ability Scales-II; GCA = General Conceptual Ability; BRIEF-2 = Behavior Rating Inventory of Executive Function-2; T = T-score.

Discussion

The present study is the first to evaluate both emotional and behavioral aspects of self-regulation for young school-aged children with WS using both direct assessment of behavior and parental ratings of everyday ability. As expected, deficits in self-regulation were evident. We also investigated the unique effects of emotion regulation ability, behavior regulation ability, and overall intellectual ability on individual differences in adaptive skills. In general, findings indicated that overall intellectual ability and/or emotion-related self-regulation contributed uniquely to individual differences in socialization and daily living skills, but behavior regulation did not. In contrast, overall intellectual ability or verbal (cool) inhibitory control but not emotion regulation contributed significant unique variance to individual differences in communication skills. In the remainder of the Discussion, we discuss these findings, limitations, and future directions for research.

Self-Regulation

As a group, the present sample of children with WS displayed considerably more difficulty exercising self-regulation during the performance-based measures than did similarly-aged TD children who participated in previous studies using the same or very similar measures (for the gift task, see Carlson, 2005; Fontana et al., 2021; for the Stroop-like interference control task, see Gerstadt et al., 1994; Montgomery & Koeltzow, 2010). Our findings for the Stroop-like task are consistent with previous reports (Atkinson et al., 2003; Breckenridge et al., 2013; Carney et al., 2013; Tager-Flusberg et al., 1997) that children and adolescents with WS performed significantly worse on verbal inhibitory control tasks than age-matched TD peers and comparably or worse than similarly aged

individuals who had other genetic syndromes. Our findings for the gift-task were consistent with those reported for an older sample of children and adolescents with DS (Fontana et al., 2021). As indicated by the elevated parent ratings on the BRIEF-2, impairments in both everyday emotion regulation and everyday behavior regulation also were evident for the participants in the present study, coinciding with previous reports including larger samples of children and adolescents with WS (e.g., Greiner de Magalhães et al., 2022). In sum, the present findings for both lab-based measures and parent-ratings of emotion regulation and behavior regulation provide further evidence of delay in the development of inhibitory control by children with WS.

Relations among Adaptive Functioning, Intellectual Ability, and Self-Regulation

Our finding that socialization, communication, and daily living skills were significantly positively associated with overall intellectual ability is consistent with results of prior studies of larger samples of adults with WS (Davies et al., 1998; Howlin et al., 2010). This finding also provides the first evidence of a significant positive relation between socialization skills and overall intellectual ability for children with WS; prior studies (Dimitropoulos et al., 2009; Mervis et al., 2001) had identified significant relations only for communication skills and daily living skills.

Differential patterns of relations emerged between the behavioral and emotional components of self-regulation and the three domains of adaptive functioning. For both performance-based and parent report measures of self-regulation, socialization skills and daily living skills were significantly related to hot emotional-related self-regulation, but not cool behavior-related self-regulation. This pattern of relations for emotion regulation is consistent with previous studies of TD children that used lab-based measures of self-

regulation (Bassett et al., 2012) and with previous studies of children and adolescents with WS (Greiner de Magalhães et al., 2022) or DS (Onnivello et al., 2022) that used parent report measures of self-regulation.

For communication skills, a different pattern emerged. These skills were not significantly related to either emotion regulation (whether measured by parent report or direct assessment) or to parent report of behavior regulation but were significantly related to performance on the lab-based measure of cool inhibitory control. The latter finding is consistent with Sabat et al. (2020), who reported a significant positive relation between teacher-reported conceptual skills (which include communication skills) and performance on a cool lab-based measure of inhibitory control for adolescents with DS.

On first glance, our findings that parent-report of behavior regulation was not significantly correlated with adaptive skills in any of the three domains and that parent-report of emotion regulation was not significantly correlated with communication skills appear not to be consistent with prior findings of significant bivariate relations between all three types of adaptive skills and parent report of both behavior regulation and emotion regulation for children and adolescents with WS (Greiner de Magalhães et al., 2022) or DS (Onnivello et al., 2022). However, these findings are most likely due to the large differences in sample size. For example, the values of the nonsignificant correlations in the present study were in most cases slightly higher than the values of the significant correlations in Greiner de Magalhães et al. (2022).

Concurrent Predictors of Adaptive Functioning

The current study is the first to evaluate the concurrent effects of emotional and behavioral aspects of self-regulation and overall intellectual ability on the adaptive skills

of children with a genetic syndrome using both lab-based direct assessment of self-regulation and parental ratings of everyday self-regulation. These relations were examined separately for socialization skills, daily living skills, and communication skills.

Emotion regulation – whether assessed by lab-based or parent report measures – and overall intellectual ability significantly explained individual differences in socialization skills, after controlling for behavior regulation. Monitoring and modulating emotional responses, flexibly shifting, overcoming impulses, and intellectual ability facilitate the ability to appropriately navigate social situations. Our finding empirically supports the theoretical expectation that better emotion regulation is associated with better interpersonal and social skills (Zelazo et al., 2016).

For daily living skills, parent report of emotion regulation and overall intellectual ability made unique concurrent contributions, after controlling for parent report of behavior regulation, which suggests everyday emotion regulation skills are important for the successful performance of daily living skills. Considering the low tolerance for frustration and the challenging nature of everyday tasks of daily living for children with WS (Brawn & Porter, 2018; Mervis & Klein-Tasman, 2000), it is not surprising that monitoring and modulating emotional responses, flexibly shifting, and overall intellectual ability contribute to daily living skills. In contrast, no unique effects were detected for the performance-based measure of emotion regulation. This finding may be due to low power in combination with the moderate positive bivariate relation (r = .44) between performance-based emotion regulation and overall intellectual ability.

For communication skills, performance on the cool inhibitory control task and overall intellectual ability made unique concurrent contributions, after controlling for

performance on the hot inhibitory control task. Given that the Stroop-like task measures verbal inhibitory control and verbal ability is a component of overall intellectual ability, it is not surprising that these two independent variables contributed significant unique variance to individual differences in communication skills. In contrast, when parent-report measures were considered, only overall intellectual ability made a unique contribution to individual differences in communication skills, after controlling for emotion regulation and behavior regulation. The lack of effect for parent-report measures is likely due to the framework in which the BRIEF-2 assesses emotion and behavior self-regulation – in the context of everyday actions rather than in the context of verbal communication skills.

Although results across performance-based and parent report measures were consistent for socialization skills, different patterns of results were identified for daily living skills and communication skills. Differential patterns are not surprising, given that direct assessments occur only once in an optimal, controlled laboratory setting, whereas parental ratings measure children's skills over time, across multiple real-world settings (Isquith et al., 2013; Toplak et al., 2013). In the current study, parent report of emotion regulation and performance on the gift-task were significantly correlated, suggesting the gift task has ecological validity. In contrast, the Stroop-like task was not related to parent report of everyday behavior regulation, suggesting they are not assessing the same construct.

As previous research has indicated, individuals with WS are socially disinhibited, have difficulty maintaining friendships, and are at risk for victimization and bullying (e.g., Fisher et al., 2017; Thurman & Fisher, 2015). For most individuals with WS, the

most limited area of adaptive behavior is daily living skills (Brawn & Porter, 2018), which are important for community inclusion and for maximizing independence (Mervis & Morris, 2007). In combination with prior meta-analytic findings for TD children (Robson et al., 2020), our findings support the hypothesis that difficulties in emotion-related regulatory skills are an important contributor to both the deficits and the individual differences in socialization skills and daily living skills that are evident among individuals with WS.

Given that self-regulation is foundational to functional outcomes, it is likely that children with WS would benefit from intervention programs focused on strengthening emotion regulation skills. As suggested by Mervis and Greiner de Magalhães (2022), strategic play-based therapies such as Replays (Levine & Chedd, 2007) which take children's current social, emotional, and communication abilities into account while helping them acquire the skills needed to manage their emotional and behavioral responses at a developmentally appropriate level are likely to promote self-regulatory skills in young children with WS. Additionally, the Alert Program® (Williams & Shellenberger, 1996; see Gill et al., 2018 for critical review), which is a self-regulation intervention program that has been shown to improve aspects of emotion regulation, behavior regulation, and social skills in school-aged children with fetal alcohol syndrome (Nash et al., 2015; Wells et al., 2012), has promise for school-aged children with WS. Caregiver training designed to teach parents evidence-based strategies for helping children regulate their emotions and increase their adaptive skills, as described in the parent component of GoFAR® (Coles et al., 2018; Kable et al., 2016), also is likely to be important. These types of interventions have the potential to positively impact the

adaptive functioning and later life outcomes (e.g., community inclusion, independence, interpersonal relationships, psychological well-being) of individuals with WS.

Limitations

Although this study was the first to evaluate the concurrent effects of emotion and behavior self-regulatory skills and overall intellectual ability on domains of adaptive functioning for individuals with WS, there are some limitations to consider. Given that the BRIEF-2 instructs the parent to indicate to what degree the behavior in question has been a problem over the past six months, the ratings may be influenced by parent expectations (Gioia et al., 2015). In the context of this potential concern, it is especially noteworthy that performance on the gift-task and ERI T were significantly and strongly correlated. Additionally, because data collection had to be terminated due to the COVID-19 pandemic, the size of the sample was relatively small. Despite efforts to enroll a racially and ethnically diverse sample, most of the participants were White non-Hispanic, although the current sample did include participants who resided across a wide geographical area. Future studies including a larger number of participants from more diverse backgrounds and addressing not only adaptive competence but also other domains that previously have been found to be affected by self-regulation for TD children, such as academic achievement, internalizing problems, and externalizing problems are needed.

Conclusion

Young school-aged children with WS have difficulty with both the emotional and behavioral aspects of self-regulation, whether measured by lab-based direct assessment or parental ratings of everyday ability. Lower self-regulatory skills and more limited

intellectual ability are associated with limitations in adaptive functioning. In addition to overall intellectual ability, emotion-related self-regulation contributes a substantial amount of unique variance to individual differences in socialization skills and daily living skills in young school-aged children with WS. Implementation of validated interventions targeting the enhancement of emotion regulation by children with WS and other syndromes associated with ID is crucial for optimizing the opportunity of these children to reach their full potential.

CHAPTER IV

GENERAL DISCUSSION

My dissertation included two manuscripts which focused on the self-regulation abilities of 6 – 8-year-olds with WS. Findings from both manuscripts provided further evidence that children with WS exhibit deficits in self-regulatory skills. In the first manuscript, the performance of young school-aged children with WS on the gift-wrap delay of gratification task was characterized. The relations of performance on this lab-based measure of hot inhibitory control to parent report of self-regulation, overall intellectual ability, and chronological age were investigated. Overall intellectual ability and parental ratings of flexible emotion regulation or the inhibitory control component of effortful control accounted for unique variance in individual differences in performance on the gift-wrap task. In the second manuscript, the concurrent effects of the ability to regulate emotions, the ability to regulate behaviors, and overall intellectual ability on adaptive skills were explored. Overall intellectual ability and emotion-related self-regulation each contributed a substantial amount of unique variance to individual differences in socialization skills and daily living skills in young school-aged children with WS. In the remainder of the General Discussion, I provide an overview of the findings and implications of the two dissertation studies and briefly discuss future directions.

Deficits in Self-regulation

Findings from both manuscripts provided further evidence that children with WS exhibit deficits in self-regulatory skills. As a group, the present sample of children with WS displayed considerably more difficulty exercising self-regulation during the performance-based measures than did similarly aged TD children who participated in previous studies using the gift task (Carlson, 2005; Fontana et al., 2021) or verbal tasks similar to the Stroop-like interference control task (Gerstadt et al., 1994; Montgomery & Koeltzow, 2010). Our findings for the Stroop-like task are consistent with previous findings for children and adolescents with WS (Atkinson et al., 2003; Breckenridge et al., 2013; Carney et al., 2013; Tager-Flusberg et al., 1997). Performance on the gift-task were comparable to those reported for an older sample of children and adolescents with DS (Fontana et al., 2021). Coinciding with previous reports including larger samples of children and adolescents with WS (e.g., Greiner de Magalhães et al., 2022), impairments in both everyday emotion regulation and everyday behavior regulation also were evident for the participants in the present study as indicated by the elevated parent ratings on the BRIEF-2. For performance-based and parent report measures, a wide range of performance was observed. Thus, performance on lab-based measures and parent ratings of emotion regulation and behavior regulation highlight the heterogeneity observed among children with WS and provide evidence of delay in the development of inhibitory control by children with WS.

Gift-Delay Task: Appropriate Measure of Emotion-Related Regulation

To my knowledge, the first manuscript describes the only study that has characterized the performance of young school-aged children with WS on the gift-wrap

delay of gratification task (Kochanska et al., 1996), which has previously been used as a measure of hot, affective inhibitory control in TD children and children and adolescents with DS. Two-thirds of the present participants were unable to exercise inhibitory control for the full duration of the 60-second task. At the same time, 83% of the children who peeked were later able to disengage and reorient away from the gift at least temporarily, thus demonstrating some inhibitory control. Across the full sample, there was a wide range of performance. Children followed the instruction not to look at the gift for between 2 seconds and 60 seconds of the 60-second task.

In addition to overall intellectual ability, parent ratings of flexible emotion regulation or the inhibitory control component of effortful control accounted for unique variance in individual differences in performance on the gift-wrap task. In concordance with previous explanations of individual differences among TD children in overcoming prepotent responses in emotionally-salient contexts (Luerssen & Ayduk, 2014), our results suggest that children who are better at shifting their focus of attention away from a reward for which they need to wait are better able to implement effective regulation and problem-solving strategies to delay gratification. These findings support the conceptual overlap between these self-regulatory processes (Diamond et al., 2007; Eisenberg et al., 2014; Nigg, 2017; Zhou et al., 2012) and provide further support for the theoretical expectation that hot inhibitory control is facilitated primarily by emotion regulation rather than behavior regulation, as has been indicated in TD children (Carlson & Wang, 2007; Luerssen & Ayduk, 2014; Zelazo & Cunningham, 2007). The findings reported in the first manuscript indicated that the gift-wrap task is emotionally and motivationally significant for young school-aged children with WS and is an appropriate hot

performance-based measure of their delay of gratification and inhibitory control abilities. Furthermore, the gift-delay task is likely an appropriate measure of self-regulation for children who have syndromes associated with cognitive impairments who may not be food motivated.

Performance-Based and Parent Report Measures

The second manuscript is the first report that evaluates both emotional and behavioral aspects of self-regulation for young school-aged children with WS using both direct assessment of behavior and parent ratings of everyday ability. Findings from both manuscripts support that although similar processes may be involved, the constructs assessed in parent-report measures and direct assessments are complementary, not identical (Isquith et al., 2013; Toplak et al., 2013). In the second manuscript, the weak correlation between the Stroop-like task and parent report of behavior regulation is consistent with previous findings for both TD and clinical samples (Esbensen & Hoffman, 2018; Isquith et al., 2013; Toplak et al., 2013). To the best of my knowledge, the first manuscript it the only study to report the relation between a delay performance-based task and parent ratings on the age-appropriate BRIEF in either school-aged TD children or children with ID. It is important to note that performance on the gift-delay task was significantly correlated with parent report of emotion regulation. This suggests that the gift-delay task taps into everyday emotion regulation, thereby demonstrating ecological validity. Differences in the type of skills assessed and in task demands may partially explain the weak to moderate correlations reported between lab-based and parent-report measures (Isquith et al., 2013; Toplak et al., 2013), with emotion-related measures being more ecologically valid.

Self-Regulation and Adaptive Skills

The second manuscript described the first study to evaluate the concurrent effects of emotional and behavioral aspects of self-regulation and overall intellectual ability on the adaptive skills of children with a genetic syndrome using both lab-based direct assessment of self-regulation and parental ratings of everyday self-regulation. Conceptually, being able to regulate one's own behaviors and emotions appropriately allows an individual to better navigate the demands and complexities of societal norms in day-to-day life (Baumeister et al., 2007; Eisenberg & Zhou, 2016; Karoly, 1993). Our findings, in combination with prior studies of TD preschool and young school-aged children (Allan et al., 2014; Backer-Grøndahl et al., 2019; Bassett et al., 2012; Brock et al., 2009; Di Norcia et al., 2015; Kim et al., 2013; Mulder et al., 2014)., provide empirical evidence that hot emotion-related and cool behavior-related aspects of self-regulation differentially impact functional outcomes

Consistent with results of prior studies of larger samples of adults with WS (Davies et al., 1998; Howlin et al., 2010), socialization, communication, and daily living skills were each significantly positively associated with overall intellectual ability. The findings from the second manuscript also are in line with the pattern of relations for emotion regulation reported in previous studies of TD children that used lab-based measures of self-regulation (Bassett et al., 2012) and previous studies of children and adolescents with WS (Greiner de Magalhães et al., 2022) or DS (Onnivello et al., 2022) that used parent report measures of self-regulation. Emotion-related self-regulation contributed uniquely to individual differences in socialization and daily living skills, but behavior regulation did not.

In contrast, a different pattern emerged for communication skills. These skills were not significantly related to either emotion regulation (whether measured by parent report or direct assessment) or to parent report of behavior regulation but were significantly related to performance on the lab-based measure of cool inhibitory control. This is consistent with prior findings for adolescents with DS in which a significant positive relation between teacher-reported conceptual skills (which include communication skills) and performance on a cool lab-based measure of inhibitory control was reported (Sabat et al., 2020). Given that the Stroop-like task measures verbal inhibitory control and verbal ability is a component of overall intellectual ability, it is not surprising that these two independent variables contributed significant unique variance to everyday communication skills.

Emotion regulation – whether assessed by lab-based or parent report measures – and overall intellectual ability significantly explained individual differences in socialization skills, after controlling for behavior regulation. This finding empirically supports the theoretical expectation that better emotion regulation is associated with better interpersonal and social skills (Zelazo et al., 2016). Monitoring and modulating emotional responses, flexibly shifting, overcoming impulses, and intellectual ability facilitate the ability to appropriately navigate social situations.

For daily living skills, parent report of emotion regulation and overall intellectual ability made unique concurrent contributions, after controlling for parent report of behavior regulation, which suggests everyday emotion regulation skills are important for the successful performance of daily living skills. Considering the low tolerance for frustration and the challenging nature of everyday tasks of daily living for children with

WS (Brawn & Porter, 2018; Mervis & Klein-Tasman, 2000), it is not surprising that monitoring and modulating emotional responses, flexibly shifting, and overall intellectual ability contribute to daily living skills.

Future Directions

Both manuscripts provided evidence that self-regulatory skills in children with WS are impaired and highlighted the particular importance of emotion regulation for adaptive behaviors. Future longitudinal studies which assess self-regulation throughout childhood to identify the onset and development of self-regulatory skills in children with WS would be of value. Future studies addressing not only adaptive competence but also other domains that previously have been found to be affected by self-regulation for TD children, such as academic achievement, persistence, internalizing problems, and externalizing problems (Allan et al., 2014; Backer-Grøndahl et al., 2019; Bassett et al., 2012; Brock et al., 2009; Di Norcia et al., 2015; Kim et al., 2013; Mulder et al., 2014; see Robson et al., 2020 for meta-analysis) may provide a better understanding of the underpinnings of the heterogeneity among children with WS that is evident in these domains.

Although the findings from the second manuscript indicated that a substantial amount of unique variance in individual differences in socialization skills and daily living skills among young school-aged children with WS was accounted for by overall intellectual ability and emotion-related self-regulation, additional factors likely also play a role in self-regulatory skills and adaptive functioning and are worth investigating. Previous researchers have suggested that parental expectations influence which abilities and responsibilities they choose to emphasize (Mervis & John, 2010; Mervis & Morris,

2007). For example, parents who demand more of their child at a developmentally appropriate level and provide the initial support needed for the child to meet these expectations are likely to positively impact both their children's adaptive skills and their emotion regulation (Kozel et al., 2021; Mervis & Morris, 2007; Mervis & Greiner de Magalhães, 2022; Phillips & Klein-Tasman, 2009).

Children with WS exhibit low mastery motivation (Rowe, 2007), which includes both the drive to persist on a moderately challenging task as well as expressing pleasure when a task is mastered (Morgan et al., 1990). In line with prior research with TD children indicating that persistence is strongly associated with hot inhibitory control (Bassett et al., 2012; Sulik et al., 2010), mastery motivation in combination with emotion regulation likely facilitates children's progress toward goals and learning new skills in both the home and school settings. Future research empirically evaluating the relations between mastery motivation, emotion regulation, and adaptive skills in children with WS would be of value.

Given that self-regulation is foundational to functional outcomes, it is likely that children with WS would benefit from intervention programs focused on strengthening emotion regulation skills. As suggested by Mervis and Greiner de Magalhães (2022), play-based therapies such as Replays (Levine & Chedd, 2007) which take children's current social, emotional, and communication abilities into account while helping them acquire the skills needed to manage their emotional and behavioral responses at a developmentally appropriate level are likely to promote self-regulatory skills in young children with WS. Additionally, the Alert Program® is a self-regulation intervention which incorporates sensory integration and focuses on processing and regulating

emotions, problem-solving, planning, self-awareness, and coping strategies (Williams & Shellenberger, 1996; see Gill et al., 2018 for critical review). The Alert Program® has been shown to improve aspects of emotion regulation, behavior regulation, and social skills in school-aged children with fetal alcohol syndrome (Nash et al., 2015; Wells et al., 2012) and is promising for school-aged children with WS. Caregiver training aimed at improving children's self-regulatory and adaptive skills is also likely to be helpful (Coles et al., 2018; Kable et al., 2016). Thus, the implementation and further development of evidenced-based practices to improve emotion regulation skills that also take into account the strong desire of children with WS to interact with others has the potential to positively impact the adaptive functioning and later life outcomes (e.g., community inclusion, independence, interpersonal relationships, psychological well-being) of individuals with WS.

Conclusion

The results presented in this dissertation demonstrate that young school-aged children with WS have difficulty with both the emotional and behavioral aspects of self-regulation, whether measured by lab-based direct assessment or parent ratings of everyday ability. At the same time, the ability to regulate emotions and behaviors was characterized by considerable variability. In line with the literature on TD children, the ability of young school-aged children with WS to inhibit the urge to access an enticing reward was related to regulation of emotions and inhibitory control. Furthermore, lower self-regulatory skills and more limited intellectual ability were each associated with limitations in adaptive functioning. In addition to overall intellectual ability, emotion-related self-regulation contributed a substantial amount of unique variance to individual

differences in socialization skills and daily living skills in young school-aged children with WS. The development and validation of interventions targeting the enhancement of emotion regulation by children with WS and other syndromes associated with ID is crucial for optimizing the opportunity of these children to reach their full potential.

REFERENCES

Allan, N. P., Hume, L. E., Allan, D. M., Farrington, A. L., & Lonigan, C. J. (2014). Relations between inhibitory control and the development of academic skills in preschool and kindergarten: A meta-analysis. *Developmental Psychology, 50*(10), 2368–2379. https://doi.org/10.1037/a0037493

Allen, J. W. P., & Lewis, M. (2020). Who peeks: Cognitive, emotional, behavioral, socialization, and child correlates of preschoolers' resistance to temptation. *European Journal of Developmental Psychology, 17*(4), 481–503. https://doi.org/10.1080/17405629.2019.1665014

Anaya, B. (2016). *Self-regulation in preschoolers: Validity of hot and cool tasks as predictive measures of academic and socio-emotional aspects of school readiness* [Masters Theses & Specialist Projects, Western Kentucky University]. http://digitalcommons.wku.edu/theses/1644

Atkinson, J., & Braddick, O. (2011). From genes to brain development to phenotypic behavior. "Dorsal-stream vulnerability" in relation to spatial cognition, attention, and planning of actions in Williams syndrome (WS) and other developmental disorders. *Progress in Brain Research, 189*(2003), 261–283. https://doi.org/10.1016/B978-0-444-53884-0.00029-4

Atkinson, J., Braddick, O., Anker, S., Curran, W., Andrew, R., Wattam-Bell, J., & Braddick, F. (2003). Neurobiological models of visuospatial cognition in children with Williams syndrome: Measures of dorsal-stream and frontal function. *Developmental Neuropsychology, 23*(1–2), 139–172. https://doi.org/10.4324/9780203764800-7

Backer-Grøndahl, A., Nærde, A., & Idsoe, T. (2019). Hot and cool self-regulation, academic competence, and maladjustment: Mediating and differential relations. *Child Development, 90*(6), 2171–2188. https://doi.org/10.1111/cdev.13104

Bassett, H. H., Denham, S., Wyatt, T. M., & Warren-Khot, H. K. (2012). Refining the Preschool Self-Regulation Assessment for use in preschool classrooms. *Infant and Child Development, 21*(6), 596–616. https://doi.org/10.1002/icd.1763

Baumeister, R. F., Schmeichel, B. J., & Vohs, K. D. (2007). Self-regulation and the executive function: The self as controlling agent. In A. W. Kruglanski & E. T. Higgins (Eds.), *Handbook of basic principles* (2nd ed., pp. 516–539). Guilford Press.

Best, J. R., & Miller, P. H. (2010). A developmental perspective on executive function. *Child Development, 81*(6), 1641–1660. https://doi.org/10.1111/j.1467-8624.2010.01499.x

Brawn, G., & Porter, M. (2018). Adaptive functioning in Williams syndrome: A systematic review. *International Journal of Disability, Development and Education, 65*(2), 123–147. https://doi.org/10.1080/1034912X.2017.1353680

Breckenridge, K., Braddick, O., Anker, S., Woodhouse, M., & Atkinson, J. (2013). Attention in Williams syndrome and Down syndrome: Performance on the new early childhood attention battery. *British Journal of Developmental Psychology, 31*(2), 257–269. https://doi.org/10.1111/bjdp.12003

Brock, L. L., Rimm-Kaufman, S. E., Nathanson, L., & Grimm, K. J. (2009). The contributions of "hot" and "cool" executive function to children's academic achievement, learning-related behaviors, and engagement in kindergarten. *Early Childhood Research Quarterly*, *24*(3), 337–349. https://doi.org/10.1016/j.ecresq.2009.06.001

Camp, J. S., Karmiloff-Smith, A., Thomas, M. S. C., & Farran, E. K. (2016). Cross-syndrome comparison of real-world executive functioning and problem solving using a new problem-solving questionnaire. *Research in Developmental Disabilities, 59*, 80–92. https://doi.org/10.1016/j.ridd.2016.07.006

Carlson, S. M. (2005). Developmentally sensitive measures of executive function in preschool children. *Developmental Neuropsychology*, *28*(2), 595–616. https://doi.org/10.1207/s15326942dn2802_3

Carlson, S. M., & Moses, L. J. (2001). Individual differences in inhibitory control and children's theory of mind. *Child Development, 72*(4), 1032–1053. https://doi.org/10.1111/1467-8624.00333

Carlson, S. M., & Wang, T. S. (2007). Inhibitory control and emotion regulation in preschool children. *Cognitive Development, 22*(4), 489–510. https://doi.org/10.1016/j.cogdev.2007.08.002

Carney, D. P., Brown, J. H., & Henry, L. A. (2013). Executive function in Williams and Down syndromes. *Research in Developmental Disabilities, 34*(1), 46–55. https://doi.org/10.1016/j.ridd.2012.07.013

Chevalère, J., Camblats, A.-M., Laurier, V., Tauber, M., Thuilleaux, D., & Postal, V. (2022). Study of decision-making capacity in Prader-Willi syndrome with a gambling task. *Journal of Intellectual & Developmental Disability, 47*(1), 18–26. https://doi.org/10.3109/13668250.2020.1824613

Cohen, J. (1988). *Statistical power analysis for the behavioral sciences* (2nd ed.). Erlbaum.

Coles, C. D., Kable, J. A., Taddeo, E., & Strickland, D. (2018). GoFAR: improving attention, behavior and adaptive functioning in children with fetal alcohol spectrum disorders: Brief report. *Developmental Neurorehabilitation, 21*(5), 345–349. https://doi.org/10.1080/17518423.2018.1424263

Cuskelly, M., Gilmore, L., & Carroll, A. (2013). Self-regulation and mastery motivation in individuals with developmental disabilities: Barriers, supports, and strategies. In K. C. Barrett, N. A. Fox, G. A. Morgan, & D. J. Fidler (Eds.), *Handbook of self-regulatory processes in development: New directions and international perspectives* (pp. 381–402). Psychology Press. https://doi.org/10.4324/9780203080719-27

Davies, M., Udwin, O., & Howlin, P. (1998). Adults with Williams syndrome: Preliminary study of social, emotional and behavioural difficulties. *British Journal of Psychiatry, 172*(3), 273–276. https://doi.org/10.1192/bjp.172.3.273

Di Norcia, A., Pecora, G., Bombi, A. S., Baumgartner, E., & Laghi, F. (2015). Hot and cool inhibitory control in Italian toddlers: Associations with social competence and behavioral problems. *Journal of Child and Family Studies, 24*(4), 909–914. https://doi.org/10.1007/s10826-014-9901-z

Diamond, A. (2013). Executive functions. *Annual Review of Psychology, 64*, 135–168. https://doi.org/10.1146/annurev-psych-113011-143750

Diamond, A. (2016). Why improving and assessing executive functions early in life is critical. In J. A. Griffin, P. McCardle, & L. S. Freund (Eds.*), Executive function in preschool-age children: Integrating measurement, neurodevelopment, and translational research* (pp. 11–43). American Psychological Association. https://doi.org/10.1037/14797-002

Diamond, A., Barnett, W. S., Thomas, J., & Munro, S. (2007). Preschool program improves cognitive control. *Science, 318*(5855), 1387–1388. https://doi.org/10.1126/science.1151148

Dimitropoulos, A., Ho, A. Y., Klaiman, C., Koenig, K., & Schultz, R. T. (2009). A comparison of behavioral and emotional characteristics in children with autism, Prader-Willi syndrome, and Williams syndrome. *Journal of Mental Health Research in Intellectual Disabilities, 2*(3), 220–243. https://doi.org/10.1080/19315860903052204

Dodd, H. F., Porter, M. A., Peters, G. L., & Rapee, R. M. (2010). Social approach in pre-school children with Williams syndrome: The role of the face. *Journal of Intellectual Disability Research, 54*(3), 194–203. https://doi.org/10.1111/j.1365-2788.2009.01241.x

Eisenberg, N., Hofer, C., Sulik, M. J., & Spinrad, T. L. (2014). Self-regulation, effortful control, and their socioemotional correlates. In J. J. Gross (Ed.), *Handbook of emotion regulation* (2nd ed., pp. 157–172). Guilford Press.

Eisenberg, N., & Zhou, Q. (2016). Conceptions of executive function and regulation: When and to what degree do they overlap? In J. A. Griffin, P. McCardle, & L. S. Freund (Eds.), *Executive function in preschool-age children: Integrating measurement, neurodevelopment, and translational research* (pp. 115–136). American Psychological Association. https://doi.org/10.1037/14797-006

Elliott, C. D. (2007). *Differential Ability Scales-II.* Psychological Corporation.

Esbensen, A. J., & Hoffman, E. K. (2018). Impact of sleep on executive functioning in school-age children with Down syndrome. *Journal of Intellectual Disability Research, 62*(6), 569–580. https://doi.org/10.1111/jir.12496

Fisher, M. H., Lough, E., Griffin, M. M., & Lane, L. A. (2017). Experiences of bullying for individuals with Williams syndrome. *Journal of Mental Health Research in*

Intellectual Disabilities, 10(2), 108–125. https://doi.org/10.1080/19315864.2016.1278289

Fontana, M., Usai, M. C., Pellizzoni, S., & Passolunghi, M. C. (2021). Inhibitory dimensions and delay of gratification: A comparative study on individuals with Down syndrome and typically developing children. *Brain Sciences, 11*(5), 636. https://doi.org/10.3390/brainsci11050636

Friard, O., & Gamba, M. (2016). BORIS: A free, versatile open-source event-logging software for video/audio coding and live observations. *Methods in Ecology and Evolution, 7*(11), 1325–1330. https://doi.org/10.1111/2041-210X.12584

Garon, N. (2016). A review of hot executive functions in preschoolers. *Journal of Self-Regulation and Regulation, 2,* 56–81. https://doi.org/10.11588/josar.2016.2.34354

Garon, N., Bryson, S. E., & Smith, I. M. (2008). Executive function in preschoolers: A review using an integrative framework. *Psychological Bulletin, 134*(1), 31–60. https://doi.org/10.1037/0033-2909.134.1.31

Gerstadt, C. L., Hong, Y. J., & Diamond, A. (1994). The relationship between cognition and action: Performance of children 3 1/2 – 7 years old on a Stroop-like day-night test. *Cognition, 53*(2), 129–153. https://doi.org/10.1016/0010-0277(94)90068-X

Gill, K., Thompson-Hodgetts, S., & Rasmussen, C. (2018). A critical review of research on the Alert Program®. Journal of Occupational Therapy, Schools, and Early Intervention, 11(2), 212–228. https://doi.org/10.1080/19411243.2018.1432445

Gioia, G. A., Espy, K. A., & Isquith, P. K. (2002). *Behavior Rating Inventory of Executive Function–Preschool Version (BRIEF-P)*. Psychological Assessment Resources.

Gioia, G. A., Isquith, P. K., Guy, S. C., & Kenworthy, L. (2000). *Behavior Rating Inventory of Executive Function (BRIEF).* Psychological Assessment Resources.

Gioia, G. A., Isquith, P. K., Guy, S. C., & Kenworthy, L. (2015). *BRIEF-2: Behavior Rating Inventory of Executive Function: Professional Manual.* Psychological Assessment Resources.

Gioia, G. A., Isquith, P. K., Retzlaff, P. D., & Espy, K. A. (2002). Confirmatory factor analysis of the Behavior Rating Inventory of Executive Function (BRIEF) in a

clinical sample. In *Child Neuropsychology* (Vol. 8, Issue 4, pp. 249–257). https://doi.org/10.1076/chin.8.4.249.13513

Greiner de Magalhães, C., Pitts, C. H., & Mervis, C. B. (2022). Executive function as measured by the Behavior Rating Inventory of Executive Function-2: Children and adolescents with Williams syndrome. *Journal of Intellectual Disability Research, 66*(1-2), 94–107. https://doi.org/10.1111/jir.12858

Gusdorf, L. M., Karreman, A., van Aken, M. A., Deković, M., & van Tuijl, C. (2011). The structure of effortful control in preschoolers and its relation to externalizing problems. *British Journal of Developmental Psychology, 29*(3), 612–634. https://doi.org/10.1348/026151010X526542

Halvorsen, M., Mathiassen, B., Amundsen, T., Ellingsen, J., Brøndbo, P. H., Sundby, J., Steinsvik, O. O., & Martinussen, M. (2019). Confirmatory factor analysis of the Behavior Rating Inventory of Executive Function in a neuro-pediatric sample and its application to mental disorders. *Child Neuropsychology, 25*(5), 599–616. https://doi.org/10.1080/09297049.2018.1508564

Harmon, A. G. (2020). *Language ability and concurrent predictors of pragmatic communication in children with Williams syndrome or 7q11.23 duplication syndrome* [Doctoral Dissertation, University of Louisville]. ProQuest Dissertations and Theses Global.

Hatton, D. D., Wheeler, A. C., Skinner, M. L., Bailey, D. B., Sullivan, K. M., Roberts, J. E., Mirrett, P., & Clark, R. D. (2003). Adaptive behavior in children with fragile X syndrome. *American Journal on Mental Retardation, 108*(6), 373. https://doi.org/10.1352/0895-8017(2003)108<373:ABICWF>2.0.CO;2

Hocking, D. R., Reeve, J., & Porter, M. A. (2015). Characterising the profile of everyday executive functioning and relation to IQ in adults with Williams syndrome: Is the BRIEF adult version a valid rating scale? *PLoS ONE, 10*(9), 1–19. https://doi.org/10.1371/journal.pone.0137628

Holodynski, M., Seeger, D., Kortas-Hartmann, P., & Wörmann, V. (2013). Placing emotion regulation in a developmental framework of self-regulation. In K. C. Barrett, N. A. Fox, G. A. Morgan, D. J. Fidler, & L. A. Daunhauer (Eds.), *Handbook of self-regulatory processes in development: New directions and*

international perspectives (pp. 27–59). Psychology Press. https://doi.org/10.4324/9780203080719.ch3

Howlin, P., Elison, S., Udwin, O., & Stinton, C. (2010). Cognitive, linguistic and adaptive functioning in Williams syndrome: Trajectories from early to middle adulthood. *Journal of Applied Research in Intellectual Disabilities, 23*(4), 322–336. https://doi.org/10.1111/j.1468-3148.2009.00536.x

Isquith, P. K., Roth, R. M., & Gioia, G. A. (2013). Contribution of rating scales to the assessment of executive functions. *Applied Neuropsychology: Child, 2*(2), 125–132. https://doi.org/10.1080/21622965.2013.748389

Jones, W., Bellugi, U., Lai, Z., Chiles, M., Reilly, J., Lincoln, A., & Adolphs, R. (2000). II. Hypersociability in Williams syndrome. *Journal of Cognitive Neuroscience, 12*, 30–46. https://doi.org/10.1162/089892900561968

Kable, J. A., Taddeo, E., Strickland, D., & Coles, C. D. (2016). Improving FASD children's self-regulation: Piloting Phase 1 of the GoFAR intervention. *Child and Family Behavior Therapy, 38*(2), 124–141. https://doi.org/10.1080/07317107.2016.1172880

Karoly, P. (1993). Mechanisms of self-regulation: A systems view. *Annual Review of Psychology, 44*(1), 23–52. https://doi.org/10.1146/annurev.psych.44.1.23

Kim, S., Nordling, J. K., Yoon, J. E., Boldt, L. J., & Kochanska, G. (2013). Effortful control in "hot" and "cool" tasks differentially predicts children's behavior problems and academic performance. *Journal of Abnormal Child Psychology, 41*(1), 43–56. https://doi.org/10.1007/s10802-012-9661-4

Klein-Tasman, B. P., & Mervis, C. B. (2003). Distinctive personality characteristics of 8-, 9-, and 10-year-olds with Williams syndrome. *Developmental Neuropsychology, 23*(1–2), 269–290. https://doi.org/10.1080/87565641.2003.9651895

Kochanska, G., Murray, K., Jacques, T. Y., Koenig, A. L., & Vandegeest, K. A. (1996). *Inhibitory control in young children and its role in emerging internalization. Child Development, 67*(2), 490–507. https://doi.org/10.1111/j.1467-8624.1996.tb01747.x

Kochanska, Grazyna, & Kim, S. (2013). Difficult temperament moderates links between maternal responsiveness and children's compliance and behavior problems in low-income families. *Journal of Child Psychology and Psychiatry and Allied Disciplines, 54*(3), 323–332. https://doi.org/10.1111/jcpp.12002

Kozel, B. A., Barak, B., Kim, C. A., Mervis, C. B., Osborne, L. R., Porter, M., & Pober, B. R. (2021). Williams syndrome. *Nature Reviews Disease Primers, 7*(1). https://doi.org/10.1038/s41572-021-00276-z

Lai, P. T., Ng, R., & Bellugi, U. (2022). Parental report of cognitive and social-emotionality traits in school-age children with autism and Williams syndrome. *International Journal of Developmental Disabilities, 68*(3), 309–316. https://doi.org/10.1080/20473869.2020.1765296

Levine, K., & Chedd, N. (2007). *Replays: Using play to enhance emotional and behavioral development for children with autism spectrum disorders*. Jessica Kingsley Publishers.

Leyfer, O. T., John, A. E., Woodruff-Borden, J., & Mervis, C. B. (2012). Factor structure of the children's behavior questionnaire in children with Williams syndrome. *Journal of Autism and Developmental Disorders, 42*(11), 2346–2353. https://doi.org/10.1007/s10803-012-1482-3

Liu, Q., Zhu, X., Ziegler, A., & Shi, J. (2015). The effects of inhibitory control training for preschoolers on reasoning ability and neural activity. *Scientific Reports, 5*, 1–11. https://doi.org/10.1038/srep14200

Luerssen, A., & Ayduk, O. (2014). The role of emotion and emotion regulation in the ability to delay gratification. In J. J. Gross (Ed.), *Handbook of emotion regulation* (2nd ed., pp. 111–125). Guilford Press.

Martens, M. A., Wilson, S. J., & Reutens, D. C. (2008). Research review: Williams syndrome: A critical review of the cognitive, behavioral, and neuroanatomical phenotype. *Journal of Child Psychology and Psychiatry and Allied Disciplines, 49*(6), 576–608. https://doi.org/10.1111/j.1469-7610.2008.01887.x

McClelland, M. M., & Cameron, C. E. (2012). Self-Regulation in early childhood: Improving conceptual clarity and developing ecologically valid measures. *Child*

Development Perspectives, 6(2), 136–142. https://doi.org/10.1111/j.1750-8606.2011.00191.x

Mehsen, V., Morag, L., Chesta, S., Cleaton, K., & Burgos, H. (2022). Hot executive function assessment instruments in preschool children: A systematic review. *International Journal of Environmental Research and Public Health, 19*(1), 95. https://doi.org/10.3390/ijerph19010095

Mervis, C. B., & Greiner de Magalhães, C. (2022). Williams syndrome. In M. Beauchamp, R. Peterson, M. D. Ris, H. G. Taylor, & K. O. Yeates (Eds.), *Pediatric neuropsychology: Research, theory, and practice* (3rd ed., pp. 377–405). Guilford Press.

Mervis, C. B., & John, A. E. (2010). Cognitive and behavioral characteristics of children with Williams syndrome: Implications for intervention approaches. *American Journal of Medical Genetics, Part C: Seminars in Medical Genetics, 154*(2), 229–248. https://doi.org/10.1002/ajmg.c.30263

Mervis, C. B., Klein-Tasman, B. P., & Mastin, M. E. (2001). Adaptive behavior of 4- through 8-year-old children with Williams syndrome. *American Journal on Mental Retardation, 106*(1), 82–93. https://doi.org/10.1352/0895-8017(2001)106<0082:ABOTYO>2.0.CO;2

Mervis, C. B., & Morris, C. A. (2007). Williams syndrome. In J. L. Ross & M. M. M. Mazzocco (Eds.), *Neurogenetic developmental disorders: Variation of manifestation in childhood* (pp. 199–262). MIT Press.

Mervis, Carolyn B., & Klein-Tasman, B. P. (2000). Williams syndrome: Cognition, personality, and adaptive behavior. *Mental Retardation and Developmental Disabilities Research Reviews, 6*(2), 148–158. https://doi.org/10.1002/1098-2779(2000)6:2<148::AID-MRDD10>3.0.CO;2-T

Mischel, W., Ebbesen, E. B., & Raskoff Zeiss, A. (1972). Cognitive and attentional mechanisms in delay of gratification. *Journal of Personality and Social Psychology, 21*(2), 204–218. https://doi.org/10.1037/h0032198

Miyake, A., Friedman, N. P., Emerson, M. J., Witzki, A. H., Howerter, A., & Wager, T. D. (2000). The unity and diversity of executive functions and their contributions

to complex “frontal lobe” tasks: A latent variable analysis. *Cognitive Psychology, 41*(1), 49–100. https://doi.org/10.1006/cogp.1999.0734

Montgomery, D. E., & Koeltzow, T. E. (2010). A review of the day-night task: The Stroop paradigm and interference control in young children. *Developmental Review, 30*(3), 308–330. https://doi.org/10.1016/j.dr.2010.07.001

Morgan, G. A., Harmon, R. J., & Maslin-Cole, C. A. (1990). Mastery motivation: Definition and measurement. *Early Education & Development, 1*(5), 318–339. https://doi.org/10.1207/s15566935eed0105_1

Morris, C. A., Braddock, S. R., Chen, E., Trotter, T. L., Berry, S. A., Burke, L. W., Geleske, T. A., Hamid, R., Hopkin, R. J., Introne, W. J., Lyons, M. J., Scheuerle, A. E., Stoler, J. M., Freedenberg, D. L., Jones, M. C., Saul, R. A., & Tarini, B. A. (2020). Health care supervision for children with Williams syndrome. *Pediatrics, 145*(2). https://doi.org/10.1542/peds.2019-3761

Mulder, H., Hoofs, H., Verhagen, J., van der Veen, I., & Leseman, P. P. M. (2014). Psychometric properties and convergent and predictive validity of an executive function test battery for two-year-olds. *Frontiers in Psychology, 5*, 1–17. https://doi.org/10.3389/fpsyg.2014.00733

Nash, K., Stevens, S., Greenbaum, R., Weiner, J., Koren, G., & Rovet, J. (2015). Improving executive functioning in children with fetal alcohol spectrum disorders. *Child Neuropsychology, 21*(2), 191–209. https://doi.org/10.1080/09297049.2014.889110

Ng-Cordell, E., Hanley, M., Kelly, A., & Riby, D. M. (2018). Anxiety in Williams syndrome: The role of social behaviour, executive functions and change over time. *Journal of Autism and Developmental Disorders, 48*(3), 796–808. https://doi.org/10.1007/s10803-017-3357-0

Nigg, J. T. (2017). Annual research review: On the relations among self-regulation, self-control, executive functioning, effortful control, cognitive control, impulsivity, risk-taking, and inhibition for developmental psychopathology. *Journal of Child Psychology and Psychiatry and Allied Disciplines, 58*(4), 361–383. https://doi.org/10.1111/jcpp.12675

O'Toole, S., Monks, C. P., & Tsermentseli, S. (2018). Associations between and development of cool and hot executive functions across early childhood. British Journal of Developmental Psychology, 36(1), 142–148. https://doi.org/10.1111/bjdp.12226

Onnivello, S., Colaianni, S., Pulina, F., Locatelli, C., Marcolin, C., Ramacieri, G., Antonaros, F., Vione, B., Piovesan, A., & Lanfranchi, S. (2022). Executive functions and adaptive behaviour in individuals with Down syndrome. *Journal of Intellectual Disability Research, 66*(1–2), 32–49. https://doi.org/10.1111/jir.12897

Osborne, J. W. (2017). *Regression & linear modeling: Best practices and modern methods*. SAGE Publications.

Pérez-García, D., Brun-Gasca, C., Pérez-Jurado, L. A., & Mervis, C. B. (2017). Behavioral profiles of children with Williams syndrome from Spain and the United States: Cross-cultural similarities and differences. American Journal on *Intellectual and Developmental Disabilities, 122*(2), 156–172. https://doi.org/10.1352/1944-7558-122.2.156

Phillips, K. D. (2008). *Emotion regulation and dysregulation in children and adolescents with Williams syndrome* [Doctoral Dissertation, University of Wisconsin-Milwaukee]. ProQuest Dissertations and Theses Global.

Phillips, K. D., & Klein-Tasman, B. P. (2009). Mental health concerns in Williams syndrome: Intervention considerations and illustrations from case examples. *Journal of Mental Health Research in Intellectual Disabilities, 2*(2), 110–133. https://doi.org/10.1080/19315860802627601

Pitts, C. H., & Mervis, C. B. (under review). Hot inhibitory control in children with Williams syndrome: Relations between delay of gratification, intellectual ability, and self-regulation. *Research in Developmental Disabilities*.

Robson, D. A., Allen, M. S., & Howard, S. J. (2020). Self-regulation in childhood as a predictor of future outcomes: A meta-analytic review. *Psychological Bulletin, 146*(4), 324–354. https://doi.org/10.1037/bul0000227

Roth, R. M., Isquith, P. K., & Gioia, G. A. (2005). *Behavior Rating Inventory of Executive Function-Adult Version*. Psychological Assessment Resources.

Rothbart, M. K., Ahadi, S. A., Hershey, K. L., & Fisher, P. (2001). Investigations of temperament at three to seven years: The children's behavior questionnaire. *Child Development, 72*(5), 1394–1408. https://doi.org/10.1111/1467-8624.00355

Rothbart, M. K., & Bates, J. E. (2006). Temperament. In N. Eisenberg, W. Damon, & R. M. Lerne (Eds.), *Handbook of child psychology: Social, emotional, and personality development* (pp. 99–166). John Wiley & Sons, Inc.

Rowe, M. L. (2007). *Mastery motivation in young children with Williams syndrome or Down syndrome.* (Publication No. 3267110) [Doctoral Dissertation, University of Louisville]. ProQuest Dissertations and Theses Global.

Sabat, C., Arango, P., Tassé, M. J., & Tenorio, M. (2020). Different abilities needed at home and school: The relation between executive function and adaptive behaviour in adolescents with Down syndrome. *Scientific Reports, 10*(1), 1–10. https://doi.org/10.1038/s41598-020-58409-5

Schworer, E. K., Esbensen, A. J., Nguyen, V., Bullard, L., Fidler, D. J., Daunhauer, L. A., Mervis, C. B., Becerra, A. M., Abbeduto, L., & Thurman, A. J. (2022). Patterns and predictors of adaptive skills in 2- to 7-year-old children with Down syndrome. *Journal of Neurodevelopmental Disorders, 14*(1), 18. https://doi.org/10.1186/s11689-022-09430-4

Shaffer, R. C., Reisinger, D. L., Schmitt, L. M., Lamy, M., Dominick, K. C., Smith, E. G., Coffman, M. C., & Esbensen, A. J. (in press). Systematic Review: Emotion Dysregulation in Syndromic Causes of Intellectual and Developmental Disabilities. *Journal of the American Academy of Child and Adolescent Psychiatry*. https://doi.org/10.1016/j.jaac.2022.06.020

Shamosh, N. A., & Gray, J. R. (2008). Delay discounting and intelligence: A meta-analysis. *Intelligence, 36*(4), 289–305. https://doi.org/10.1016/j.intell.2007.09.004

Sparrow, S. S., Cicchetti, D. V., & Saulnier, C. A. (2016). *Vineland Adaptive Behavior Scales, Third Edition* (Vineland-3). Pearson.

Strømme, P., Bjømstad, P. G., & Ramstad, K. (2002). Prevalence estimation of Williams syndrome. *Journal of Child Neurology, 17*(4), 269–271. https://doi.org/10.1177/088307380201700406

Sulik, M. J., Huerta, S., Zerr, A. A., Eisenberg, N., Spinrad, T. L., Valiente, C., Di Giunta, L., Pina, A. A., Eggum, N. D., Sallquist, J., Edwards, A., Kupfer, A., Lonigan, C. J., Phillips, B. M., Wilson, S. B., Clancy-Menchetti, J., Landry, S. H., Swank, P. R., Assel, M. A., & Taylor, H. B. (2010). The factor structure of effortful control and measurement invariance across ethnicity and sex in a high-risk sample. *Journal of Psychopathology and Behavioral Assessment, 32*(1), 8–22. https://doi.org/10.1007/s10862-009-9164-y

Tager-Flusberg, H., Sullivan, K., & Boshart, J. (1997). Executive functions and performance on false belief tasks. *Developmental Neuropsychology, 13*(4), 487–493. https://doi.org/10.1080/87565649709540689

Thurman, A. J., & Fisher, M. H. (2015). The Williams syndrome social phenotype: Disentangling the contributions of social interest and social difficulties. *International Review of Research in Developmental Disabilities, 49*, 191–227. https://doi.org/10.1016/bs.irrdd.2015.06.002

Toplak, M. E., West, R. F., & Stanovich, K. E. (2013). Practitioner Review: Do performance-based measures and ratings of executive function assess the same construct? *Journal of Child Psychology and Psychiatry and Allied Disciplines, 54*(2), 131–143. https://doi.org/10.1111/jcpp.12001

Wells, A. M., Chasnoff, I. J., Schmidt, C. A., Telford, E., & Schwartz, L. D. (2012). Neurocognitive habilitation therapy for children with fetal alcohol spectrum disorders: An adaptation of the Alert Program®. *American Journal of Occupational Therapy, 66*(1), 24–34. https://doi.org/10.5014/ajot.2012.002691

Williams, M. S., & Shellenberger, S. (1996). "How does your engine run?"®: A leader's guide to the Alert Program® for self-regulation. TherapyWorks, Inc.

Willoughby, M., Kupersmidt, J., Voegler-Lee, M., & Bryant, D. (2011). Contributions of hot and cool self-regulation to preschool disruptive behavior and academic achievement. *Developmental Neuropsychology, 36*(2), 162–180. https://doi.org/10.1080/87565641.2010.549980

Woodruff-Borden, J., Kistler, D. J., Henderson, D. R., Crawford, N. A., & Mervis, C. B. (2010). Longitudinal course of anxiety in children and adolescents with Williams

syndrome. *American Journal of Medical Genetics, Part C: Seminars in Medical Genetics*, *154*(2), 277–290. https://doi.org/10.1002/ajmg.c.30259

Zelazo, P. D., Blair, C., & Willoughby, M. T. (2016). Executive function: Implications for education. *National Center for Education Research, Institute of Education Sciences, U.S. Department of Education*. https://eric.ed.gov/?&id=ED570880

Zelazo, P. D., & Carlson, S. M. (2012). Hot and cool executive function in childhood and adolescence: Development and plasticity. *Child Development Perspectives*, *6*(4), 354–360. https://doi.org/10.1111/j.1750-8606.2012.00246.x

Zelazo, P. D., & Cunningham, W. A. (2007). Executive function: Mechanisms underlying emotion regulation. In J. J. Gross (Ed.), *Handbook of emotion regulation* (Vol. 12, Issue 11, pp. 135–158). Guilford Press.

Zelazo, P. D., & Müller, U. (2010). Executive function in typical and atypical development. In U. Goswami (Ed.), *Blackwell handbook of childhood cognitive development* (2nd ed., pp. 574–603). Wiley-Blackwell. https://doi.org/10.1002/9781444325485.ch22

Zhou, Q., Chen, S. H., & Main, A. (2012). Commonalities and differences in the research on children's effortful control and executive function: A call for an integrated model of self-regulation. *Child Development Perspectives*, *6*(2), 112–121. https://doi.org/10.1111/j.1750-8606.2011.00176.x

Zitzer-Comfort, C., Doyle, T., Masataka, N., Korenberg, J., & Bellugi, U. (2007). Nature and nurture: Williams syndrome across cultures. *Developmental Science, 10*(6), 755–762. https://doi.org/10.1111/j.1467-7687.2007.00626.x

CURRICULUM VITAE

Holley Pitts Arnold
(C. Holley Pitts)

PERSONAL INFORMATION

Office Address:	1800 Gervais Street Department of Psychology University of South Carolina Columbia, SC 29201
Phone Number:	(803) 292-7913
E-Mail Address:	pittsc@mailbox.sc.edu

EDUCATION

University of Louisville, Louisville, KY
Experimental Psychology
Ph.D., December 2022

University of Louisville, Louisville, KY
Experimental Psychology
M.S., August 2015

University of South Carolina, Columbia, SC
Major: Experimental Psychology
Minor: Statistics
B.A., *magna cum laude*, May 2010

RESEARCH

Peer-reviewed Publications

Thurman, A. J., Bullard, L., Kelly, L. Wong, C., Nguyen, V., Esbensen, A. J., Bekins, J., Schworer, E. K., Fidler, D. J., Daunhauer, L., Mervis, C. B., **Pitts, C. H.**, Becerra, A., & Abbeduto, L. (2022). Defining expressive language benchmarks for young children with Down syndrome. *Brain Sciences.* https://doi.org/10.3390/brainsci12060743

Greiner de Magalhães, C.G., **Pitts, C. H.**, & Mervis, C. B. (2022). Executive function as measured by the Behavior Rating Inventory of Executive Function-2: Children and adolescents with Williams syndrome. *Journal of Intellectual Disability Research*, *66*, 94-107. https://doi.org/10.1111/jir.12858

Pitts, C. H., Klein-Tasman, B. P., Osborne, J. W., & Mervis, C. B. (2016). Predictors of specific phobia in children with Williams syndrome. *Journal of Intellectual Disability Research, 60,* 1031-1042. https://doi.org/10.1111/jir.12327

Pitts, C. H., & Mervis, C. B. (2016). Performance on the Kaufman Brief Intelligence Test-2 by children with Williams syndrome. *American Journal of Intellectual and Developmental Disabilities, 121*, 33-47. https://doi.org/10.1352/1944-7558-121.1.33

Mervis, C. B., & **Pitts, C. H.** (2015). Children with Williams syndrome: Developmental trajectories for intellectual abilities, vocabulary abilities, and adaptive behavior. *American Journal of Medical Genetics Part C: Seminars in Medical Genetics, 169*, 158-171. https://doi.org/10.1002/ajmg.c.31436

Mervis, C. B, Klein-Tasman, B. P., Huffman, M., Velleman, S. L., **Pitts, C. H.**, Henderson, D. R., Woodruff-Borden, J., Morris, C. A., & Osborne, L. R. (2015). Children with 7q11.23 duplication syndrome: Psychological characteristics. *American Journal of Medical Genetics: Part A, 167*, 1436-1450. https://doi.org/10.1002/ajmg.a.37071

Palomares, M. C., Smith, P. R., & **Pitts, C. H.** (2012). Enumeration of small and large numerosities: The effect of element visibility. *The Quarterly Journal of Experimental Psychology, 65*, 1185-1194. https://doi.org/10.1080/17470218.2011.648647

Palomares, M. C., Smith, P. R., **Pitts, C. H.**, & Carter, B. M. (2011). The effect of viewing eccentricity on enumeration. *PLoS ONE 6*(6): e20779. https://doi.org/10.1371/journal.pone

Conference Presentations (peer-reviewed)

Pitts, C. H., Gopan, G., & Mervis, C. B. (2021, April). *Delay of gratification in 6- to 8-year-olds with Williams syndrome: Relations with emotion regulation and IQ.* Talk presented at the Gatlinburg Conference on Research and Theory in Intellectual and Developmental Disabilities, Kansas City, MO.

Kelly, L. A., Wong, C. H., Esbensen, A., Schworer, E., Fidler, D., Daunhauer, L., Mervis, C. B., **Pitts, C. H.**, Becerra, A., Abbeduto, L., & Thurman, A. J.

(2021, April). *Defining expressive language benchmarks for young children with Down syndrome.* Poster presented at the Gatlinburg Conference on Research and Theory in Intellectual and Developmental Disabilities, Kansas City, MO.

Greiner de Magalhães, C. G., **Pitts, C. H.**, & Mervis, C. B (2021, April). *Executive function of children and adolescents with Williams syndrome.* Poster presented at the Gatlinburg Conference on Research and Theory in Intellectual and Developmental Disabilities, Kansas City, MO.

Mervis, C. B., Becerra, A. M., **Pitts, C. H.**, & Marchman, V. (2019, June). *MacArthur-Bates Communicative Development Inventory expressive vocabulary and sentence complexity norms for children with Williams syndrome aged 16 – 48 months.* Poster presented at the Symposium of Research in Child Language Disorders, Madison, WI.

Greiner de Magalhães, C. G., **Pitts, C. H.**, & Mervis, C. B. (2019, June). *Phonological processing profiles of 9-year-old children with Williams syndrome and relations to word reading ability.* Talk presented at the Symposium of Research in Child Language Disorders, Madison, WI.

Pitts, C. H., & Mervis, C. B. (2019, April). *Inhibitory control in 6- to 8-year-olds with Williams syndrome: Relations with adaptive skills, school readiness, and IQ.* Poster presented at the Gatlinburg Conference on Research and Theory in Intellectual and Developmental Disabilities, San Antonio, TX.

Pitts, C. H., Teixeira, M. C. T., Osório, A. A. C., Rossi, N. F., Giacheti, C. M., Sampaio, A., & Mervis, C. B. (2018, April). *Cross-cultural comparison of behavioral profiles of children with Williams syndrome: Brazil and the United States.* Poster presented at the Gatlinburg Conference on Research and Theory in Intellectual and Developmental Disabilities, San Diego, CA.

Greiner de Magalhães, C. G., Becerra, A. M., Eovino, J., Speak, J. M., Thurman, A. J., **Pitts, C. H.**, & Mervis, C. B. (2018, April). *Lexical and grammatical abilities of 3- and 4-year-old children with Down syndrome or Williams syndrome.* Poster presented at the Gatlinburg Conference on Research and Theory in Intellectual and Developmental Disabilities, San Diego, CA.

Joseph, M. P., **Pitts, C. H.**, & Mervis, C. B. (2018, April). *Cognitive profile of 5- to 8-year-olds with Williams syndrome as measured by the Differential Ability Scales-II Early Years.* Poster presented at the Gatlinburg Conference on Research and Theory in Intellectual and Developmental Disabilities, San Diego, CA.

Pitts, C. H., & Mervis, C.B. (2017, March). *Cognitive profile of Children with Williams syndrome as measured by the Differential Ability Scales-II School-Aged Battery.* Poster presented at Gatlinburg Conference on Research and Theory in Intellectual and Developmental Disabilities, San Antonio, TX.

Pitts, C. H., Klein-Tasman, B. P., Osborne, J. W., & Mervis, C. B. (2016, March). *Predictors of specific phobia in children with Williams syndrome: Behavioral regulation and IQ*. Poster presented at Gatlinburg Conference on Research and Theory in Intellectual and Developmental Disabilities, San Diego, CA.

Pitts, C. H., Adelson, J. A., & Mervis, C. B. (2015, April). *Longitudinal stability of expressive vocabulary of children with Williams syndrome: Multilevel modeling of performance on the Expressive Vocabulary Test-2nd edition.* Poster presented at Gatlinburg Conference on Research and Theory in Intellectual and Developmental Disabilities, New Orleans, LA.

Pitts, C. H., Adelson, J. A., & Mervis, C. B. (2014, July). *Longitudinal stability of receptive vocabulary of children with Williams syndrome: Multilevel modeling of performance on the Peabody Picture Vocabulary Test-4th edition*. Talk presented at International Williams Syndrome Scientific and Professional Conference, Garden Grove, CA.

Pitts, C. H., & Mervis, C. B. (2014, July). *Appropriateness of the Kaufman Brief Intelligence Test-2 for capturing the full range of abilities for children with Williams syndrome*. Poster presented at International Williams Syndrome Scientific and Professional Conference, Garden Grove, CA.

Pitts, C. H., Adelson, J. A., & Mervis, C. B. (2014, March). *Early language milestones and later intellectual abilities of children with Williams syndrome*. Poster presented at Gatlinburg Conference on Research and Theory in Intellectual and Developmental Disabilities, Chicago, IL.

Mervis, C. B., Adelson, J. A., & **Pitts, C. H.** (2014, March). *Young children with Down syndrome: Variability in language and cognitive abilities*. Invited talk presented at Alzheimer's Disease in Down Syndrome: From Molecules to Cognition. Wellcome Trust Genome Campus, Hinxton, Cambridge, UK.

DeLapp, C. L., **Pitts, C. H.**, & Mervis, C. B. (2013, June). *Expressive vocabularies of 18 – 48-month-olds with Williams syndrome*. Poster presented at Symposium on Research in Child Language Disorders, Madison, WI.

Pitts, C. H., & Mervis, C. B. (2013, March). *Performance on the Kaufman Brief Intelligence Test-2 by children with Williams syndrome*. Poster presented at Gatlinburg Conference on Research and Theory in Intellectual and Developmental Disabilities, San Antonio, TX.

Pitts, C. H., & Palomares, M. C. (2012, May). *In the averaged crowd, children are better than adults in size discrimination.* Poster presented at Vision Science Society, Naples, FL.

Moyer, J., Payne, A., **Pitts, C. H.**, & Palomares, M. C. (2012, May). *Visual memory: Statistics, masking, configuration.* Poster presented at Vision Science Society, Naples, FL.

Carter, B.C., **Pitts, C. H.**, & Palomares, M. C. (2012, May). *Visual apprehension of small and large numerosities.* Poster presented at Vision Science Society, Naples, FL.

Palomares, M. C., Smith, P. R., & **Pitts, C. H.** (2011, November). *Enumeration of small and large numerosities: The effect of element visibility.* Poster presented at Psychonomic Society, Seattle, WA.

Pitts, C. H., Bourda, K. N., & Palomares, M. C. (2011, May). *Mean vs. range in statistical summary representation.* Poster presented at Vision Science Society, Naples, FL.

Palomares, M. C., & **Pitts, C. H.** (2011, May). *Size discrimination: On the relationship of statistical averaging and "crowding."* Poster presented at Vision Science Society, Naples, FL.

Palomares, M. C., **Pitts, C. H.**, & Morris, W. Z. (2010, November). *The meaning of sets: Information at a single glance.* Poster presented at Psychonomic Society, St. Louis, MO.

TEACHING EXPERIENCE AND CERTIFICATES

Graduate Teaching Assistant Academy (Fall 2016 – Spring 2017). Certificate received from School of Interdisciplinary and Graduate Studies, University of Louisville.

STEM Graduate Teaching Assistant Academy (Summer 2015). Certificate received from School of Interdisciplinary and Graduate Studies, University of Louisville.

Teaching Assistant (Spring 2015). Applied Multiple Regression. Graduate level course in the College of Education and Human Development, University of Louisville.

AWARDS AND HONORS

University Fellowship, University of Louisville (2016 – 2018)
David Zeaman Graduate Travel Award, Gatlinburg Conference on Research and Theory in Intellectual and Developmental Disabilities (2018)
Award for Excellence in Research, Department of Psychological and Brain Sciences (2017)
Graduate Network in Arts and Sciences Research Funding (Fall 2016, 2017, 2018; Spring 2019)
Graduate Student Council Travel Award (Fall 2016, 2017, 2018; Spring 2019)
Graduate Dean's Citation (2015)
South Carolina Life Scholarship (2006 – 2010)
University Scholars Award (2006 – 2010)
Phi Beta Kappa (2009)

METHODOLOGICAL EXPERTISE

Knowledge of CardioPeak and Segmenter, CardioEdit, and CardioBatch electrocardiogram (ECG) processing software

Experience with SPSS, AMOS, and HLM statistical software

Completed courses in structural equation modeling and hierarchical linear modeling

Proficient with Access for data storage and management

Expertise in training and administering standardized assessments to children with neurodevelopmental disorders, intellectual disability, and typically developing children, including:

Sensory Processing Assessment (SPA), Communication and Symbolic Behavior Scales Developmental Profile (CSBS), Mullen Scales of Early Leaning; Differential Ability Scales-II; Clinical Evaluation of Language Fundamentals (CELF-5 and CELF Preschool-2); Kaufman Brief Intelligence Test-2; Peabody Picture Vocabulary Test-4; Expressive Vocabulary Test-2; Preschool Language Scales-5; Wechsler Individual Achievement Test-III; Wechsler Abbreviated Scale of Intelligence-II