



Intervention Response by Genetic Subtype: PRETEND-Preschool Program for Children with Prader-Willi Syndrome via Remote Parent Training

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Abstract

Prader-Willi Syndrome (PWS) is a rare neurodevelopmental disorder associated with social cognitive challenges, and pretend play has been demonstrated as a tool to achieve developmental goals. Following previous report on feasibility and acceptability of a remote, play-based parent-training program (Zyga, Russ, & Dimitropoulos, 2018), we now report on preliminary efficacy of this program to enhance pretend play skills and social cognitive skills in preschoolers with PWS. Results across two studies demonstrated efficacy when live-coaching play sessions incorporated children into the intervention. Increases in play skills were observed for children with the mUPD subtype of PWS who underwent intervention, compared with children with mUPD who were waitlisted. Children with DEL subtype were less likely to respond to intervention. Implications for results are discussed.

Keywords Prader-Willi Syndrome · Pretend play · Social Cognition · Telehealth · Parent-training

Introduction

Individuals with Prader-Willi syndrome (PWS), a rare neurodevelopmental disorder associated with alterations to chromosome 15, experience a number of physical, cognitive, and behavioral challenges (see Butler et al., 2019 for updated review of PWS). Among these challenges, children and adults with PWS are at increased risk of having difficulty with various aspects of social cognition (Whittington & Holland, 2017), including reciprocal communication and interpreting social cues (Dimitropoulos et al., 2013; Rosner et al., 2004), face and emotion discrimination (Key et al.,

2013; Whittington & Holland, 2011; Feldman & Dimitropoulos, 2014; Dykens et al., 2019; Debladis et al., 2019), symbolic play (Zyga et al., 2015; Dimitropoulos et al., 2019), and theory of mind (Lo et al., 2013). For example, Dykens et al., (2019) found that children and adults with PWS had difficulty accurately interpreting the intention of others, with particular difficulty perceiving other people's sincere intentions. In addition, they found many participants had difficulty recognizing emotions, more often showing challenges with negative emotions (Dykens et al., 2019). However, while research has shown that children with PWS are at increased risk of exhibiting social cognitive challenges akin to symptoms of autism spectrum disorder (ASD), the majority do not meet diagnostic criteria for the disorder (Dykens et al., 2017; Bennett et al., 2015). In addition, while these social cognitive difficulties may at times be superseded by other more challenging behaviors, such as the life-threatening hyperphagia (Miller et al., 2011) and temper outbursts (Rice, Woodcock, & Einfeld, 2018), improving social-cognitive abilities may increase other important areas of children's skill development, increase quality of life, and reduce the significant burden experienced by caregivers (Kayadjanian et al., 2018; Wong et al., 2020).

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Much of the literature in PWS has been focused on older children and adults with respect to broader social functioning. We recently reported challenges with social functioning at younger ages within the context of pretend (symbolic) play (Dimitropoulos et al., 2019; Zyga & Dimitropoulos, 2020). Although there are many versions of play, pretend play refers to the ability to play in an “as-if” manner (Fein, 1981). Krasnor and Pepler (1980) defined this type of play using four specific criteria: nonliterality, positive affect, intrinsic motivation, and flexibility. Another hallmark of pretend play is its use of symbolism and make-believe scenarios (Russ, 2014). Pretend play emerges in a child’s development around 18 months and hits its peak between the ages of three to five (Krasnor & Pepler, 1980). Its emergence follows a developmental trajectory, where children’s abilities move from more concrete to abstract actions and themes (Bodrova et al., 2013). Further, pretend play can be seen as a marker of social cognitive development where children are able to practice social skills in a safe environment (Russ, 2014). Specifically, pretend play has been shown to relate and impact areas such as divergent thinking ability, social adjustment, social communication ability surrounding joint attention, theory of mind, understanding other’s emotional states, and even socioemotional development surrounding expressing, managing, and coping with emotions (Barnett, 1990; Dansky, 1999; Fehr & Russ, 2016; Harris, 2000; Russ, 2014; Singer & Singer, 1992). Further, there is also a large research literature that has found that pretend play ability is associated with many areas of adaptive functioning in children, such as creativity, coping, and emotion regulation (Danksy, 1999; Russ, 2014).

While pretend play involves essential skills that are important for all children, children with autism and other neurodevelopmental disorders have been shown to exhibit deficits in the emergence and development of their pretend play skills, including imagination, flexible thinking, and symbolic play (Kasari et al., 2006). Our findings indicate preschoolers with PWS have difficulties with engaging in pretend play and decreased social skills (Dimitropoulos et al., 2019). In addition, parent-child engagement during pretend play was found to be lower among PWS parent-child dyads than typical child-parent dyads (Zyga & Dimitropoulos, 2020). Studies have demonstrated differences related to social cognitive processes between children with the two most commonly recognized subtypes of PWS: maternal uniparental disomy (mUPD) and paternal deletion of chromosome 15 (DEL). Children diagnosed with the mUPD subtype of PWS have been shown to be at greater risk for autistic symptomatology compared with children with the DEL subtype of PWS (Hogart et al., 2010). Additionally, children with mUPD have scored similarly to children with ASD on measures of social cognition, communication, and

motivation (Dimitropoulos et al., 2013). Whittington and Holland (2010) demonstrated that socialization and age have a greater predictive impact on emotional recognition for children with mUPD, whereas this predictive nature was negligible for children with DEL. Consistent with this accumulating literature on social cognition in PWS, play behavior may also vary by genetic subtype, with decreased play ability among children with mUPD in contrast to children with DEL. For example, we recently reported on the play skills of 50 preschoolers with PWS and found that on average, children with DEL subtype scored higher than both the mUPD and ASD groups on the organization of their play, how often they incorporated affect into their play, and the amount of time they spent engaged in symbolic play (Dimitropoulos et al., 2019).

Pretend play can serve as both a domain of development and an activity platform with which children can develop their play skills and improve upon these skills to reach important developmental goals (Lifter, Mason, & Barton 2011; Lifter and Bloom, 1989; Barton & Wolery 2008). There is an extensive literature on use of play as a platform for intervention with children with autism and other neurodevelopmental disabilities, as well as efforts to understand the challenges associated with pretend play for this population (Charman et al., 2003; Strain et al., 1985; Barton & Wolery 2010; Doernberg et al., 2021). Thus, increasing pretend play skills may not only allow for play optimization but also build a stronger platform for intervention on other challenges the child may be experiencing. Recently, our lab reported on the efficacy of a remote pretend play intervention administered directly to school-aged children with PWS, and findings demonstrated that not only did children increase in their cognitive and affective play skills following the intervention, but these improvements generalized to their divergent thinking skills as well (Dimitropoulos et al., 2021). In addition, while direct intervention on play has been used across children with neurodevelopmental disabilities in structured clinician-led sessions, parent training is often used as a mode of intervention or for ensuring reinforcement of learning outside of the intervention session (Kasari et al., 2015; Greenspan and Wieder, 1997; Mercer, 2017; Solomon et al., 2014). Incorporating parents in these pursuits has several advantages. First, teaching parents best practices for boosting pretend play will help to ensure parents have the knowledge and tools they need to engage with their child. Play can be hard work for children that struggle with the essential foundational behaviors required to play socially with another person (i.e. joint attention, joint engagement, and reciprocity), in contrast to typically developing children for whom play is often easy, naturally reinforcing, and therefore enjoyable for both the child and parent. This natural reinforcement seen in typical

development usually leads to a cycle of continued engagement in parent-child play. This cycle is often disrupted or absent in parent-child dyads of children with developmental disorders. These parents can feel ineffective or unmotivated to initiate play because of lack of engagement from their child or being uncertain as to how to gain their child's interest or attention in the task. Providing coaching to the parent along with psychoeducation can help induce this important parent-child play cycle. Second, parents offer the opportunity to extend learning outside the dedicated intervention session in their day-to-day structured and unstructured interactions with their child to allow for reinforcement and generalization. Third, the use of early intervention via parent training provides an opportunity to target fundamental play skills early in life for children, providing parents a skillset that can be used successfully at home. Research in neurodevelopmental disorders has demonstrated the success of implementing play-based parent-training as a form of early intervention (Kasari, et al., 2015). Finally, providing opportunities for structured play time between parent and child may allow for increased parent-child engagement. Recent research has indicated that play-training for parents and children with ADHD can lead to increases in the quality of parent-child relationships (Wallace, 2018). Specifically, in autism, parent training has been shown to result in significant improvement among preschoolers' behavior (Scahill et al., 2016), and research in children with PWS indicates that the addition of an adult play-partner increases pretend play skills (Zyga et al., 2015).

Given the prevalence of PWS (approximately 1 in 10,000 to 30,000 births; Angulo et al., 2015) and other rare neurodevelopmental disorders, developing efficacious behavioral interventions can be challenging as in-person access to participants is limited and research is often subject to small sample sizes collected at one or two timepoints and/or at limited timepoints (i.e., syndrome-specific conferences). The parameters of behavioral intervention (i.e., need for consistent, weekly or biweekly appointments) are often not feasible and likely associated with travel costs and increased time burden on the family. However, telehealth methodology (videoconferencing) helps to reduce these barriers (Vismara et al., 2013). This modality is becoming increasingly prevalent in the rare disorder community (Cox et al., 2012; McGeary et al., 2012) and in light of the recent COVID-19 pandemic, is garnering even more attention with broader typical and atypical populations (Jeffrey et al., 2020; Fairchild et al., 2020).

We recently reported on the feasibility of a remotely delivered parent training for parents of preschoolers with PWS and found good acceptability and satisfaction with the intervention (Zyga, Russ, & Dimitropoulos, 2018). The purpose of this paper is to now report on the preliminary efficacy

of this program to enhance pretend play skills in preschoolers with Prader-Willi syndrome. We report on two studies. The first describes our initial pilot of the PRETEND parent training program (Study 1) in a 6-week, twice-weekly, remotely delivered format. Study 2 refines the remotely delivered training program to condense twice-weekly sessions to once-weekly, across 8 weeks of intervention, and including live coaching and assigned play-homework to participants. Both studies included a quasi-randomized design where the children randomized to the intervention group are hypothesized to show significant gains in pretend play skills in comparison to those in the waitlist condition following parent-training.

Study 1

Method

Participants

Thirty preschool-aged children diagnosed with Prader-Willi Syndrome, and their parents/guardians, were enrolled in Study 1. To be eligible to participate, families had a child between the ages of 3–5 years old with a confirmed diagnosis of PWS by genetic testing, had reliable computer and internet access, spoke English as their primary language, and were able to complete both the pre and post assessment visits in person. Participating families were excluded if enrolled in any other clinical trials during the time of this study. No other inclusion nor exclusion criteria were used in the present study. Recruitment occurred through newsletters, online postings, and announcements at meetings of state and national chapters of the Foundation for Prader-Willi Syndrome Research (FPWR) and the Prader-Willi Syndrome Association (PWSA-USA). All participating parents/guardians completed informed consent outlining procedures of the study.

Of the 30 participants who enrolled in Study 1 (mean age=4.37, SD=0.85; 60% male), 15 participants were quasi-randomized into the intervention group (INV), while 15 participants were quasi-randomized into the waitlist control group (WC). Participants were randomized based on their gender, age, and genetic subtype in order to ensure that the INV and WC groups had comparable samples. Nonverbal cognitive ability was estimated using the Visual Reception subscale of the Mullen Scales of Early Learning (MSEL) (mean t-score: 32.13 (9.19)), and average receptive language ability was 85.79 (15.72), as measured by the Peabody Picture Vocabulary Test (PPVT-2). These scores fall within the typical low average cognitive range for this population (Angulo et al., 2015). See Table 1 for complete

Table 1 Baseline Characteristics (Study 1)

M (SD)	Total (n=30)	DEL (n=17)	mUPD (n=13)	INV (n=15)	WC (n=15)
Age	4.34 (0.93)	4.62 (0.97) [†]	3.98 (0.75)	4.39 (1.05)	4.28 (0.82)
# male(%)	18 (60)	9 (52.94)	9 (69.23)	8 (53.33)	7 (46.67)
# mUPD(%)	--	--	--	8 (53.33)	7 (46.67)
# Non-White(%)	2 (6)	--	--	--	--
PPVT ^a	85.79 (15.72)	90.41 (13.62) ⁺	79.25 (16.73)	86.57 (12.90)	85.07 (18.41)
MSEL ^b	32.13 (9.79)	36.14 (10.08) ⁺	26.50 (6.15)	31.92 (8.93)	32.36 (11.17)
Child Stress ^{c*}	50.32 (7.62)	52.69 (6.65) ⁺	47.17 (7.94)	51.00 (7.72)	49.54 (7.73)
Parent Stress ^{c*}	48.00 (8.33)	50.56 (7.04) ⁺	44.58 (8.97)	46.73 (6.84)	49.46 (9.86)
Total Stress ^{c*}	49.04 (7.60)	51.63 (5.63) ⁺	45.58 (8.71)	48.60 (6.56)	49.54 (8.91)
Life Stress ^{c*}	47.43 (9.30)	49.81 (11.12)	44.25 (4.92)	46.53 (10.08)	48.46 (8.59)
SCQ ^{d*}	9.41 (6.16)	7.44 (3.35)	11.85 (7.93)	8.71 (5.34)	10.07 (6.95)
SSIS Social Skills ^{e*}	94.25 (13.32)	96.93 (8.12)	91.15 (17.41)	91.50 (12.46)	97.00 (14.03)
SSIS Problem Behaviors ^{f*}	103.14 (12.39)	105.40 (14.08)	100.54 (10.02)	104.64 (14.80)	101.64 (9.75)

^a Peabody Picture Vocabulary Test – Receptive Language (Standard Score)

^b Visual Reception Cognitive Subtest (T Score)

^c Parenting Stress Index (Standard Score)

^d Social Communication Questionnaire (Raw Total Score)

^e Social Skills Improvement System (Standard Score)

* Higher scores = greater impairment

⁺ Significantly greater scores than other subtype

[†] Trending significantly greater scores than other subtype

baseline characteristics of the entire sample, genetic groups, and INV and WC groups.

Measures

Baseline

Mullen Scales of Early Learning (MSEL; Mullen, 1995) Brief, individually administered measure of the overall cognitive functioning in infants and children up to 68 months of age across five domains (gross motor, visual reception, fine motor, expressive language, and receptive language). In the present study, only the visual reception subtest was administered to obtain an overall estimate of nonverbal cognitive ability, given that previous research has shown this to be a

valid and reliable indicator of early cognitive ability in typical and atypical populations (Bishop et al., 2011).

Peabody Picture Vocabulary Test (PPVT-4, Dunn & Dunn 2007) An individually administered measure of receptive vocabulary for standard American English for ages 2 years 6 months to 90 years. The measure has shown good validity and reliability across both typical and atypical populations (Dunn & Dunn, 2007). Overall receptive language ability is reported as a Standard Score (M = 100; SD = 15).

Social Skills Improvement System Rating Scales (SSIS; Gresham and Elliott, 2008) A parent/caregiver survey that evaluates social skills, problem behaviors, and academic competence in children ages 3–18 years of age. The measure includes 12 subscales (communication, cooperation, assertiveness, responsibility, empathy, engagement, self-control, externalizing, bullying, hyperactivity/inattention, internalizing, and autism spectrum) and an overall standard score with national norms for preschool age children. Standard scores and behavior levels (below average, average, above average) are given for each subscale. Reliability and validity evidence have been collected for its use in special populations (Gresham and Elliott, 2008).

Social Communication Questionnaire (SCQ; Rutter et al., 2003) A brief instrument completed by a parent/caregiver that evaluates communication skills and social functioning in screening for an ASD. The survey provides a global cutoff score of 15 with scores above this value indicating a high probability of autism. It is appropriate for children with a mental age over 2.0 years and has been shown to be an efficient, valid, and reliable way to obtain diagnostic information or screen for autistic symptoms (Rutter et al., 2003).

Parent-child interaction task Parent-child dyads were asked to play with a set of toys (i.e., blocks, small figurines, and cars) any way they liked for 5 min. This interaction was recorded and later coded based on a modified tangram task coding system (Hudson & Rapee, 2001) and Mother-Child Structured Interaction Rating Scales (adapted from the Eunice Kennedy Shriver National Institute of Child Development and Human Development [NICHD] Study of Early Child Care Research Network). Coded variables measured aspects of the parent's engagement (Parental Involvement, Unsolicited Help, Response to Child), aspects of the child's ability to engage (Social Interest and Social Competence), and aspects of the dyad's overall interaction (Mutual Enjoyment and General Mood). Scales that measured degree of Parental Involvement, Unsolicited Help, Response to Child, and Mutual Enjoyment and General Mood were adapted from the modified tangram task coding system (Hudson &

Rapee, 2001). Degree of Parental Involvement, Unsolicited Help, Response to Child were scored on an 9-point scale (0–8); Mutual Enjoyment and General Mood were scored on a 6-point scale (0–5). For degree of Parental Involvement and Unsolicited Help, a score of 0 indicated no help or involvement and a score of 8 indicated overinvolvement or unnecessary help. For response to child and mutual enjoyment and general mood, a score of 0 indicated a positive response or interaction, whereas higher scores indicated a negative response or interaction. Scales that measured Social Interest and Social Competence were taken from the Mother-Child Structured Interaction Rating Scale (NICHD) and were coded on a 7-point scale (1–7), where a score of 1 indicated low interest or competence. These measures have been used as valid and reliable measures of the previously mentioned variables across diverse child populations, including those with ASD (McDonald, Baker, & Messinger, 2016).

Parenting Stress Inventory (PSI-4; Johnson, 2015) A 120-item self-report questionnaire that assesses the magnitude of stress in the parent-child system. Items factor into three different domains of stress: child characteristics, parent characteristics, and situational/demographic characteristics. Child characteristics include: Distractibility/Hyperactivity Adaptability Reinforces Parent (i.e., assesses the parent's experience of interactions with his or her child as positively reinforcing), Demandingness, Mood, Acceptability (i.e., assesses the extent to which child characteristics meet expectations of the parent). Parent characteristics include: Competence, Isolation, Attachment, Health, Role Restriction (i.e., assesses the parent's sense of limited freedom and constrained personal identity as a result of the parenting role), Depression, Spouse/Parenting Partner Relationship. Levels of stress across domains are reported as t-scores, with average scores falling between 40 and 54. The four t-score measures reported on in the present study are: Child Domain Stress, Parent Domain Stress, Total Stress (a sum score of child and parent domain stress) and Life Stress. This measure has been used with parents of children with various developmental and psychological disorders, including ASD (Tomanik, Harris, & Hawkins, 2004) and has been found to be a reliable and valid measure of stress across the previously described domains.

Pre/Post-Intervention

Affect in play scale – preschool (APS-P; Kaugars and Russ, 2009) A standardized play task designed to measure various dimensions of children's pretend play and validated for

preschool children ages 4–5 years, in a number of studies (Kaugars and Russ, 2009; Fehr & Russ 2014). In addition, this scale has been extended for use with 3-year-old children, which has reliably been done in previous work (Yates and Marcelo, 2014). In this task, a variety of structured toys are laid out on a table (cups, stuffed animals, toy car) and children are provided with a story stem and instructions to play with the toys and talk out loud for a 5-minute period. The play is videotaped and then scored according to a detailed manual that assessed both cognitive and affective processes in the play narrative (Kaugars and Russ, 2009). The child's play is scored using a criterion-based rating scale. The variables included ones that captured cognitive processes in play, specifically: (1) Organization of the storyline, (2) Imagination ability to pretend, and (3) Comfort in playing with the toys. These variables were all scored on a 1–5 scale; one being the lowest ability in that domain. Variables that measured affective processes included: (1) Frequency of Affect, a total frequency count of affect units expressed within the play narrative and (2) Variety of Affect, a total count of the number of affect categories out of 11 possible categories expressed during the play. Affect scores relate to child's ability to have mental representations of emotions and then express these emotions in play. For example, a child may recognize that a character is happy to play with another toy during their story and will voice this through expressing “Yay, this is fun!” or having the toys hug. Further, for each 20-second interval, the rater indicates which of three types of play (No Play; Functional Play; Symbolic Play) was the predominant activity – i.e. occurred for greater than or equal to 10 seconds within each 20-second interval. No Play is defined as the child not moving or interacting with the toys. Functional Play relates to a child making simple, repetitive muscle movements with the toys. Symbolic Play is defined as any instance of using toys in an “as-if” manner, substituting an object for another, or using the object in any way other than how it is intended.

Procedure

Baseline visit

During the initial visit, the child completed cognitive and behavioral assessments (MSEL, PPVT-4 and APS-P) while the parent completed demographics and various measures of their child's social cognitive functioning (SCQ and SSIS) and self-report of their own stress related to parenting (PSI-4). Additionally, parents and children completed a measure of parent-child play (PCI). All baseline measures were completed in a lab setting or at an offsite location near the participant's home (e.g. a private library room). Two graduate

level research assistants were used in the current study. Each participating family only worked with one graduate research assistant as their interventionist for the entirety of the study. Upon visit completion, the parent received an intervention folder, which included a parent manual and technological instructions.

Intervention

The PRETEND (**Play-based Remote Enrichment To ENhance Development**) Preschool Program during Study 1 was a 6-week intervention administered remotely via a videoconferencing software to parents of children with PWS. During the intervention period, the parent worked directly with the interventionist via videoconferencing software to complete the PRETEND-Preschool program. The intervention program was adapted from a play-based program aimed at increasing imagination and emotional expression in typically developing children (Moore & Russ, 2008) and a parent-training intervention (PEBM; Tonge et al., 2014). Interventionists followed manualized procedures to ensure fidelity in targeting specific skills throughout the program (see Zyga et al., 2018 for further details on intervention protocol). Interventionists trained individually with one another on parent-training session content in order to adhere with fidelity to the intervention protocol. The PRETEND-Preschool program consisted of individual parent sessions to review play and related cognitive, emotional and behavioral skills, and assigned homework for parents to practice the learned skills from sessions with their children. The PRETEND-Preschool program included 12 sessions, 30–45 min each, delivered twice a week, focused on the following four core skills areas: engagement and play, improving problem behaviors, emotional understanding and coping skills, and social skills and peer interactions. Sessions focused specifically on the following topics: The Social Cognitive Profile, How to be a Play Partner, Sustaining and Maintaining Attention, The ABC's of Behavior, Defining Play, Increasing Complexity in Play, All About Emotions, Emotional Regulation, Defining Social Communication, Building Social Engagement, Putting it All Together, and finally the Program Overview. While parents were encouraged to practice learned skills with their children weekly, the intervention focus was parent training and did not directly involve the child. The PRETEND program has been previously reported on by our group for its feasibility and acceptability (Zyga et al., 2018). All remote sessions were conducted using Adobe Connect videoconferencing software.

Post-Intervention visit

Within four weeks of the completion of the intervention program, the child and parent were seen again for an in-person visit in the lab or at an offsite location near the participant's home. The child underwent the same behavioral assessments as completed at baseline and parents were asked to complete the same surveys. Children were again administered the APS-P as a pre-post intervention measure of their cognitive and affective play skills. The Parent-child interaction task was also re-administered and included as exploratory analysis.

Results

Baseline characteristics

Descriptive statistics of age, gender, receptive language (PPVT Standard Score), and overall cognitive ability (Mullen's T Score) were examined for both the entire sample and by genetic subtype (see Table 1). The DEL subgroup was slightly older than the mUPD group (trending significance, $p = .059$), had higher receptive language skills than the mUPD group (trending significance, $p = .058$, see Table 1), and had significantly higher overall cognitive ability ($p = .014$, see Table 1). Additionally, parents of children in the DEL subgroup demonstrated significantly higher distress (as measured by the PSI-4) reported in the Child Domain (trending significance, $p = .056$), Parent Domain (trending significance, $p = .059$), and higher Total Stress ($p = .035$), as compared with parents of children in the mUPD subgroup (see Table 1). In contrast, parents of participating children in the mUPD subgroup endorsed higher social cognitive deficits (SCQ scores) for their children, as compared to the children in the DEL subgroup (trending significance, $p = .053$, see Table 1). No differences were observed between the DEL or mUPD subgroups on reported social skills or problem behaviors (as measured by the SSIS, see Table 1).

Coding and reliability

The Parent-child interaction task and the APS-P were coded by two independent coders (undergraduate research assistants) blind to group membership and participant subtype. Coders were trained directly from the original manual of the measures (Hudson & Rapee, 2001; Kaugars and Russ, 2009; Fehr & Russ 2014). Reliability data was scored for 100% of the participant sample. Interrater agreement was calculated by an interclass correlation coefficient for each Parent-child interaction task and APS-P variable scored by

Table 2 Mixed Factorial ANOVA results for INV vs. WC (Whole Sample) from Baseline to Post-Intervention (Study 1)

M (SD) beneath APS-P Variable	INV Baseline (n = 11)	INV Outcome (n = 11)	WC Baseline (n = 10)	WC Outcome (n = 10)	p-value	Effect Size
Imagination	2.45 (1.13)	2.36 (0.92)	3.60 (0.70)	3.00 (0.94)	0.390	0.039
Organization	2.64 (1.03)	2.73 (0.91)	2.90 (0.88)	2.50 (0.97)	0.279	0.061
Affect Frequency	7.18 (5.88)	5.36 (4.23)	7.00 (5.25)	7.80 (5.63)	0.347	0.047
Affect Variety	1.82 (1.17)	1.82 (1.40)	2.60 (1.43)	1.90 (1.29)	0.336	0.049
No Play^a	0.12 (0.15)	0.09 (0.09)	0.21 (0.16)	0.23 (0.19)	0.496	0.025
Functional Play	0.53 (0.33)	0.54 (0.25)	0.42 (0.20)	0.38 (0.20)	0.636	0.012
Symbolic Play	0.35 (0.31)	0.28 (0.17)	0.36 (0.21)	0.38 (0.21)	0.462	0.029

^aDecreases = improvement

the two raters, to ensure interrater reliability of scoring. A two-way random effects model tested for agreement using a 95% confidence interval. All variables on the Parent-child interaction task and APS-P had interrater agreement above 0.80 (acceptable).

Baseline to Post-Intervention analyses

A series of 2 × 2 (Group [INV, WC] X Time [timepoint 1, timepoint 2]) mixed factorial repeated measures analyses of variance (ANOVAs) were used to evaluate within and between group differences of participating children's scores on the APS-P variables. Hypotheses were not supported, in that no significant interactions were observed between group or time on any of the APS-P variables (Imagination, Organization, Affect Frequency, Affect Variety, No Play, Functional Play, and Symbolic Play, see Table 2). Controlling for cognition did not affect the model. A significant main effect of group was observed for the Imagination variable, in that the WC group had greater overall scores than the INV group ($p = .007$). In order to investigate possible subtype effects, Genetic Subtype was included as an additional independent variable in the analysis of pre-post results between the INV and WC groups. This additional series of 2 × 2 × 2 (Group [INV, WC] X Genetic Subtype [DEL, mUPD] X Time [timepoint 1, timepoint 2]) ANOVAs revealed no significant interactions between any of the independent variables. There were also no significant group differences baseline to post-intervention for participants' scores on the Parent-child interaction task.

Summary

In Study 1, a remote, 6-week, play-based intervention was delivered weekly to parents of preschoolers with PWS. Results indicated no significant changes in children's play skills from pre to post-intervention, nor any significant impact of genetic subtype on children's play skills. Baseline analyses demonstrated that the DEL group had slightly

higher average age, higher average receptive language skills, higher overall cognitive ability, fewer social cognitive deficits, and greater reported distress from parents, compared with the mUPD group. No significant differences were observed at baseline between the groups in terms of reported social skills or problem behaviors. Further analysis indicated that no improvements were seen by genetic subtype.

Study 2

Results of Study 1 on the efficacy of the PRETEND parent-training program demonstrated that hypotheses were not confirmed, given that no changes were observed in the pretend play skills of children whose parents underwent the intervention. However, recent findings from our school-age sample (where intervention occurred directly with the child with PWS) demonstrated that children with PWS can significantly improve their cognitive and affective play skills and associated cognitive flexibility following the remote pretend play intervention (Dimitropoulos et al., 2021). This study directly involved both the children in play and the parents via parent-training sessions focused on play-based skills, and suggests that further involvement of children into the pretend play intervention may be advantageous.

Previous research supported that in typical and atypical preschool populations, the involvement of adults in play is key in facilitating success of improved cognitive and affective skills in play in children, therefore it was important to keep parents involved in the delivery of the play intervention (Fehr & Russ, 2016, Zyga et al., 2015). While parents in Study 1 were advised to practice learned play skills at home with their children, there was no controlled element of practice built into the study. Additionally, previous research indicates that parent-child play can help to improve both parent-child relationships and children's play skills (Wallace, 2018; Kasari et al., 2006). Given the success of the PRETEND intervention for school-aged children with PWS, and the lack of observed change in play skills following the parent-training intervention for preschoolers with PWS in

Table 3 Baseline Characteristics (Study 2)

M (SD)	Total (n=20)	DEL (n=8)	mUPD (n=12)	INV (n=10)	WC (n=10)
Age	3.70 (0.73)	3.75 (0.89)	3.67 (0.65)	3.90 (0.74)	3.50 (0.71)
# male(%)	10 (50)	4 (50)	6 (50)	5 (50)	5 (50)
# mUPD(%)	--	--	--	6 (60)	6 (60)
# Non-White(%)	3 (15)	--	--	--	--
PPVT ^a	89.82 (14.64)	91.17 (20.19)	89.09 (11.73)	93.25 (10.78)	86.78 (17.46)
PTI-2 ^b	89.75 (18.14)	101.50 (25.40) ⁺	82.70 (6.52)	86.63 (6.65)	92.88 (25.26)
Child Stress ^{c*}	49.20 (6.62)	51.75 (6.92)	47.50 (6.11)	51.80 (6.16)	46.60 (6.29)
Parent Stress ^{c*}	47.65 (9.14)	51.00 (11.40)	45.42 (6.92)	51.40 (8.59)	43.90 (8.44)
Total Stress ^{c*}	48.10 (7.75)	51.13 (8.94)	46.08 (6.46)	51.50 (6.80)	44.70 (7.39)
Life Stress ^{c*}	44.25 (4.80)	46.25 (5.37)	42.92 (4.08)	43.40 (3.89)	45.10 (5.65)
SCQ ^{d*}	7.45 (3.20)	9.00 (3.96) ⁺	6.42 (2.19)	7.40 (2.76)	7.50 (3.75)
SSIS Social Skills ^{e*}	89.45 (14.30)	84.38 (15.20)	92.83 (12.90)	85.20 (9.18)	93.70 (17.24)
SSIS Problem Behaviors ^{f*}	99.15 (14.39)	108.00 (10.92) ⁺	93.25 (13.67)	100.60 (9.25)	97.70 (18.62)

^aPeabody Picture Vocabulary Test – Receptive Language (Standard Score)

^bPictorial Test of Intelligence IQ (Standard Score)

^cParenting Stress Index (Standard Score)

^dSocial Communication Questionnaire (Raw Total Score)

^eSocial Skills Improvement System (Standard Score)

*Higher scores = greater impairment

⁺Significantly greater scores than other subtype

the present study, adjustments were made to further involve the children of participating parents into the PRETEND-Preschool intervention.

Method

Participants

Twenty participants were enrolled in Study 2 that had not previously participated in Study 1. Two participants were lost to follow up after the baseline visit and did not complete the intervention, therefore their data was not used for pre/post analysis. The final sample for complete pre/post analyses was $n=18$ (mean age = 3.70, $SD=0.73$; 50% male). Average Full-Scale IQ for the sample was 89.75 (18.41), and average receptive language ability was 89.92 (14.64), as measured by the Pictorial Test of Intelligence (PTI-2) and the Peabody Picture Vocabulary Test (PPVT-2),

respectively. See Table 3 for complete demographics for participants from Study 2.

Measures

Baseline and pre-post intervention

All measures used in Study 1 were used again in Study 2, with the exception of the MSEL, which was replaced by the PTI-2. In Study 1, due to time constraints at the baseline visit, only the Visual Reception subtest of the MSEL was administered as an estimate of cognitive ability. In order to obtain a more robust cognitive measure for preschool-aged children, the PTI-2 was administered for Study 2.

Pictorial test of intelligence (PTI-2; French, 2001) an individually administered test of general intelligence for both typical and disabled children ages 3–10 through 8–11 years. The three subtests (verbal ability, form discrimination, and quantitative reasoning) are combined to produce an Intelligence Quotient (IQ). The PTI-2 was deemed suitable for this study as it requires little to no verbal or motor skill effort from examinees, and reliability and validity evidence have been collected for its use in special populations (Twum and Hayford, 2019; Williams et al., 2014).

Procedure

Baseline visit

During the initial visit, the child completed cognitive and behavioral assessments (PTI-2, PPVT-4 and APS-P) while the parent completed demographics and various measures of their child's social and social cognitive functioning (SCQ and SSIS) and self-report of their own stress related to parenting (PSI-4). Additionally, parents and children completed a measure of parent-child play (PCI). All baseline measures were completed in a lab setting or offsite location near the participant's home. Two interventionists were used in the present study (advanced doctoral students in clinical psychology). Upon visit completion, the parent was given an intervention folder, which included a parent manual and technological instructions. Additionally, all parents were provided with a Dell Bluetooth Earpiece for the live-coaching sessions, that easily connected to their phone or computer to receive audio from the interventionist at the time of the coaching play session. All remote sessions were conducted via using Zoom videoconferencing software.

Intervention

The PRETEND Preschool Program for Study 2 was an 8-week intervention administered remotely via a videoconferencing software to parents of children with PWS. As in Study 1, interventionists followed the same manualized procedures to ensure fidelity in targeting specific parent-training skills throughout the program. The PRETEND-Preschool program for Study 2 had eight parent-training sessions, 45–60 min each, delivered once a week, focused on the following topics: The Social Cognitive Profile, How to be a Good Play Partner, Building Skills in Play, The ABC's of Behavior, All about Emotions, Increasing Complexity in Play, Improving Social Engagement, and Putting it all Together. These eight sessions covered the same content that was focused on across the 12 sessions in Study 1, but with slightly longer sessions to cover more topics within a single parent session. The program for Study 2 also contained an additional three live-coaching sessions, completed after Sessions 3, 5, and 7, and play-based activities assigned for homework. The live coaching sessions consisted of the parent and child playing independently for 15–20 min in front of the Zoom video, while the interventionist watched with their video camera off, and provided interactive coaching of play skills to the parent via a Bluetooth earpiece (connected either to the computer audio or a separate phone call such that the parent could hear the interventionist, but the child could not). In these live-coaching sessions, interventionists encouraged the same play techniques that were taught to the parent during the parent-training sessions, including modeling, scaffolding, reinforcing the child, and following the child's lead. In Study 1, children were not directly involved in the parent intervention, therefore the incorporation of the live-coaching sessions in Study 2 was the most notable procedural adjustment.

As in Study 1, interventionists followed manualized procedures to ensure fidelity in targeting specific skills throughout the program – see Zyga et al. 2018 for further details on original intervention protocol. The manual was adjusted slightly in terms of (a) number of sessions administered to parents, (b) the length of sessions, and (c) the addition of the 3 live-coaching sessions. All content for the manual in Study 2 remained the same for Study 1, therefore the same steps to train interventionists to fidelity were used: interventionists trained individually with one another on parent-training session content in order to adhere with fidelity to the intervention protocol.

Post-Intervention visit

Within four weeks of the completion of the intervention program, the child and parent were seen again for an in-person

visit in the lab or at an offsite location near the participant's home. The child underwent the same behavioral assessments as completed at baseline and the parent was asked to complete the same surveys. Children were again administered the APS-P as a pre/post intervention measure of their cognitive and affective play skills.

Results

Baseline characteristics

Descriptive statistics of age, gender, receptive language (PPVT Standard Score), and overall cognitive ability (PTI-2 Standard Score) were examined for both the entire sample and the genetic groups (see Table 3). Analyses of baseline characteristics of the two PWS groups in Study 2 indicated that the mUPD subgroup and the DEL subgroup differed significantly on several social and cognitive variables. Results from a series of independent samples *t* tests indicated that the DEL subgroup had slightly increased social communication impairments (as measured by the SCQ) at baseline compared with the mUPD subgroup (trending, $p = .076$). Additionally, the DEL subgroup had significantly increased problem behaviors (as measured by the SSIS) compared with the mUPD subgroup ($p = .020$). Notably, the mUPD subgroup has significantly lower overall cognitive scores compared with the DEL subgroup ($p = .040$), thus the PTI-2 score was used as a covariate in the primary analysis.

Baseline to Post-Intervention analyses

Overall Group Analysis

A series of 2×2 (Group [INV, WC] X Time [timepoint 1, timepoint 2]) mixed factorial repeated measures ANOVAs were used to evaluate within and between group differences of participating children's scores on the APS-P variables. Hypotheses were not supported, in that no significant interactions were observed between group or time on any of the APS-P variables (Imagination, Organization, Affect Frequency, Affect Variety, No Play, Functional Play, and Symbolic Play, see Table 4). In order to investigate possible subtype effects, Genetic Subtype was included as an additional independent variable in the analysis of pre-post results between the INV and WC groups. This additional series of $2 \times 2 \times 2$ (Group [INV, WC] X Genetic Subtype [DEL, mUPD] X Time [timepoint 1, timepoint 2]) ANOVAs revealed a significant interaction for the Organization variable between group, genetic subtype, and time ($p = .046$). In order to further investigate possible subtype effects, an additional series of 2×2 (Group [INV, WC] X Time [timepoint

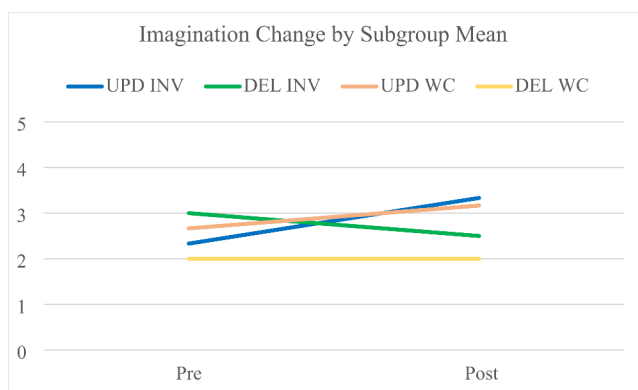


Fig. 1 APS-P Imagination Score Change from Baseline to Outcome Between Subgroups

1, timepoint 2]) mixed factorial repeated measures ANOVAs were conducted examining possible changes in the APS-P variables separately in the mUPD and DEL groups. Results indicated a significant interaction within the mUPD subgroup, such that the INV participants made significant improvements in their Organization scores from baseline to outcome ($p = .049$), whereas the WC participants in the mUPD subgroup made no significant changes (see Tables 5 and 6, and Fig. 1). A main effect of timepoint was observed for the Imagination, Functional Play, and Symbolic Play variables ($p = .024, 0.057, 0.015$, respectively), such that all participants in the mUPD subgroup made improvements on

these variables at the outcome visit. Notably, no significant changes were observed on any APS-P variables within the DEL subgroup pre to post intervention.

There were also no significant group differences baseline to post-intervention for participants' scores on the Parent-child interaction task. As the Parent-child interaction task was not a primary variable to examine for play-skill change in this study, it was not evaluated further for subtype differences.

Individual, descriptive analysis by subtype

Given the small sample sizes of the subgroups based on PWS subtype, a series of individual, descriptive analyses were used to further examine individual change between the INV and WC participants within the mUPD and DEL subtypes. A meaningful difference from baseline to outcome was defined by 0.5 of the standard deviation value of each variable within the sample. A deficit in any play process on the original APS was defined by the criterion agreement of one standard deviation below the typical mean for each play process (Zyga et al., 2015), which typically is about 0.5. By this metric of individual change, results indicated that the INV participants in the mUPD subgroup demonstrated improvement above and beyond that of the WC participants on the following APS-P variables: Imagination, Organization, Affect Frequency, No Play, Functional

Table 4 Mixed Factorial ANOVA results for INV vs. WC (Whole Sample) from Baseline to Post-Intervention (Study 2)

M (SD) underneath APS-P Variable	INV Baseline (n = 10)	INV Outcome (n = 10)	WC Baseline (n = 8)	WC Outcome (n = 8)	p-value	Effect Size
Imagination	2.60 (0.97)	3.00 (1.25)	2.50 (1.20)	2.88 (0.99)	0.959	0.000
Organization	2.70 (0.95)	2.70 (1.06)	2.38 (0.74)	2.13 (0.64)	0.382	0.048
Affect Frequency	7.90 (4.01)	10.70 (7.78)	5.75 (5.15)	7.38 (9.77)	0.773	0.005
Affect Variety	1.90 (1.45)	2.30 (1.64)	1.50 (1.07)	2.00 (1.51)	0.887	0.001
No Play^a	0.17 (0.18)	0.13 (0.20)	0.09 (0.16)	0.12 (0.18)	0.311	0.064
Functional Play	0.43 (0.24)	0.35 (0.19)	0.57 (0.30)	0.54 (0.29)	0.732	0.008
Symbolic Play	0.40 (0.28)	0.52 (0.28)	0.35 (0.23)	0.34 (0.20)	0.385	0.047

^a Decreases = improvement

Table 5 Mixed Factorial ANOVA results for INV vs. WC (mUPD) from Baseline to Post-Intervention (Study 2)

APS-P Variable	mUPD INV Baseline (n = 6)	mUPD INV Outcome (n = 6)	mUPD WC Baseline (n = 6)	mUPD WC Outcome (n = 6)	p-value	Effect Size
Imagination	2.33 (1.03)	3.33 (1.37)	2.67 (1.37)	3.17 (0.98)	0.395	0.073
Organization	2.50 (1.05)	2.83 (1.17)	2.50 (0.84)	2.17 (0.75)	0.049*	0.333
Affect Frequency	7.00 (4.82)	12.00 (7.92)	7.33 (5.01)	9.67 (10.41)	0.635	0.023
Affect Variety	2.17 (1.72)	3.00 (1.79)	1.67 (1.21)	2.50 (1.38)	1.00	0.000
No Play^a	0.25 (0.19)	0.15 (0.22)	0.09 (0.18)	0.13 (0.21)	0.163	0.185
Functional Play	0.45 (0.21)	0.27 (0.15)	0.59 (0.26)	0.47 (0.29)	0.724	0.013
Symbolic Play	0.31 (0.27)	0.59 (0.32)	0.32 (0.22)	0.40 (0.20)	0.136	0.209

^a Decreases = improvement

* $p < .05$

Table 6 Mixed Factorial ANOVA results for INV vs. WC (DEL) from Baseline to Post-Intervention (Study 2)

APS-P Variable	DEL INV Baseline (n=4)	DEL INV Outcome (n=4)	DEL WC Baseline (n=2)	DEL WC Outcome (n=2)	p-value	Effect Size
Imagination	3.00 (0.82)	2.50 (1.00)	2.00 (0.00)	2.00 (0.00)	0.312	0.250
Organization	3.00 (0.82)	2.50 (1.00)	2.00 (0.00)	2.00 (0.00)	0.312	0.250
Affect Frequency	9.25 (2.36)	8.75 (8.26)	1.00 (0.00)	0.50 (0.71)	1.00	0.000
Affect Variety	1.50 (1.00)	1.25 (0.50)	1.00 (0.00)	0.50 (0.71)	0.633	0.062
No Play^a	0.25 (0.19)	0.15 (0.22)	0.10 (0.14)	0.07 (0.09)	0.274	0.287
Functional Play	0.40 (0.30)	0.49 (0.18)	0.49 (0.51)	0.77 (0.04)	0.476	0.134
Symbolic Play	0.53 (0.27)	0.42 (0.23)	0.41 (0.37)	0.17 (0.05)	0.634	0.062

^a Decreases = improvement

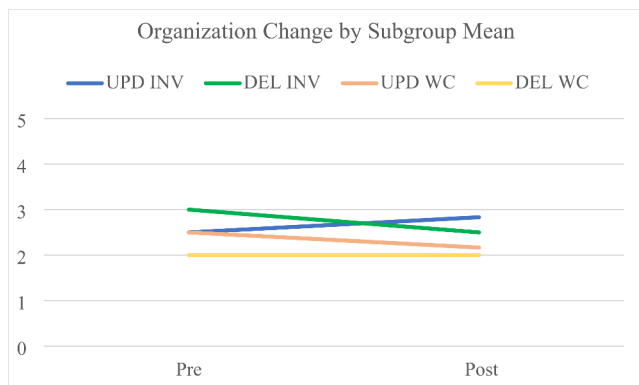


Fig. 2 APS-P Organization Score Change from Baseline to Outcome Between Subgroups

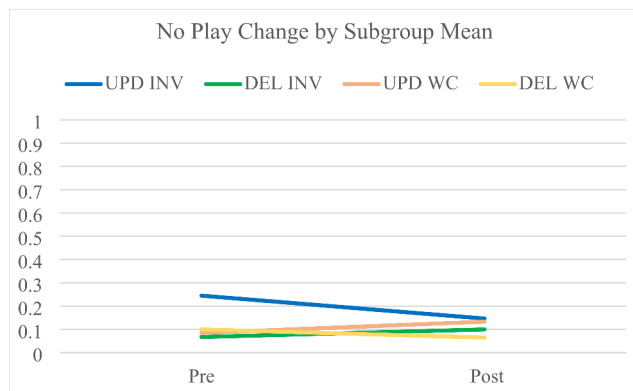


Fig. 4 APS-P No Play^a Score Change from Baseline to Outcome Between Subgroups. ^a Decreases = improvement

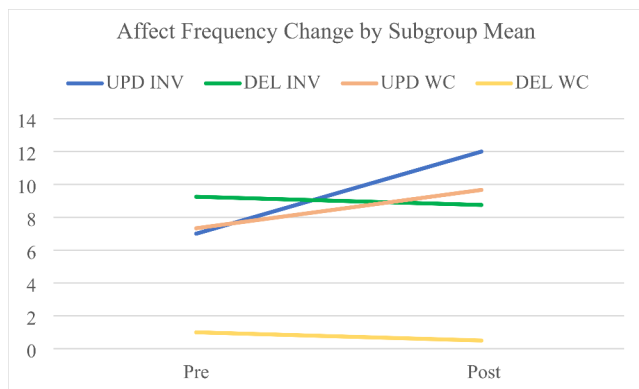


Fig. 3 APS-P Affect Frequency Score Change from Baseline to Outcome Between Subgroups

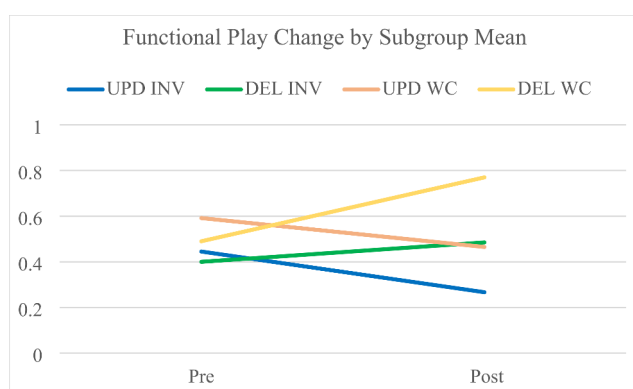


Fig. 5 APS-P Functional Play^a Score Change from Baseline to Outcome Between Subgroups. ^a Decreases = improvement

Play, and Symbolic Play. In contrast, the INV participants in the DEL subgroup demonstrated improvement above and beyond the WC participants on only the Affect Frequency and Functional Play variables. Specifically, results from this individual, descriptive analysis indicate that for children who underwent the intervention, a greater percentage of children in the mUPD subgroup showed improvement across all variables of the APS-P (see Table 7). Notably, children in the mUPD INV subgroup demonstrated more improvement in comparison to both the DEL INV subgroup

and the mUPD WC subgroup on variables of Imagination, Organization, Affect Frequency, No Play, Functional Play, and Symbolic Play (see Figs. 1, 2, 3, 4, 5 and 6).

Interaction Effects

In order to investigate possible variables contributing to the differential effects of the intervention on the two PWS subtypes, baseline measures were further examined by subtype. Zyga and Dimitropoulos (2020) indicated that

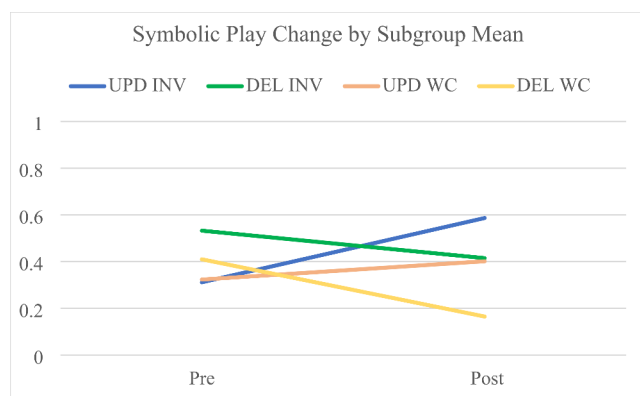


Fig. 6 APS-P Symbolic Play Score Change from Baseline to Outcome Between Subgroups

parenting stress can significantly relate to parent-child interaction, therefore a series of Pearson-product moment correlations were conducted between parent-reported stress (as measured by the PSI-4) and parent-child play skills (as measured by the PCI) at baseline. For the DEL subgroup, findings indicated the presence of significant relationships between domains of parent-reported stress, and overall impairment in dyadic variables in parent-child play. Significant positive correlations were observed among General Mood (reversed scored) in play and reported Child Domain Stress ($r = .759, p = .029$) and General Mood in play and reported Total Stress ($r = .712, p = .047$), such that participants and their parents who were rated as having greater impairments in their overall mood in their dyadic play also had increased stress in both the child domain and total stress of life. Additionally, a significant negative correlation was observed among Parent Involvement in play and reported Life Stress ($r = -.864, p = .005$), and significant positive correlations were observed among Mutual Enjoyment in play and reported Children Domain Stress, Parent Domain Stress, and Total Stress ($r = .803, p = .017, r = .732, p = .039$, and $r = .820, p = .013$, respectively), indicating that participants with greater impairments of parent involvement (overinvolvement) in dyadic parent-child play also had higher reported stress in their lives, and participants with

greater impairment in their Mutual Enjoyment in dyadic parent-child play had greater stress in both parent and child domains, and greater total stress. Additionally, results from these correlations indicated a significant negative correlation among children's Social Interest during parent-child play and reported Parent Domain stress ($r = -.713, p = .047$), and a significant positive correlation among children's Social Interest during parent-child play and reported Life Stress ($r = .810, p = .015$), indicating that participating children showing lower social interest in dyadic parent-child play had parents with higher stress in the parenting domain, but lower overall reported stress in life. Notably, no significant relationships were found among parenting stress variables and parent-child dyadic play variables in the mUPD group.

Summary

In Study 2, a remote, 8-week, play-based intervention was delivered weekly to parents of preschoolers with PWS, with live-coaching play sessions and assigned play-homework to parent-child dyads. Results indicated that there was a significant interaction between group, genetic subtype, and time for the Organization variable on the APS-P. Further analysis examining change in the INV versus WC groups specifically for participants with mUPD indicated significant improvement in organizational skills in play for the INV group at post-intervention, whereas the WC group made no change. Comparatively, no changes were observed for the INV group versus the WC group for participants with DEL. Baseline analyses demonstrated that the DEL group had higher overall cognitive ability, greater social cognitive deficits, and greater reported distress from parents, compared with the mUPD group. No significant differences were observed at baseline between the groups in terms of reported social skills, receptive language skills, or age.

Based on small sample size, data for the mUPD group and DEL group were separately examined for individual improvements across variables. The participants with mUPD in the INV group (6 participants) demonstrated

Table 7 Qualitative Improvement from Baseline to Outcome

	mUPD INV % (ratio ^b)	DEL INV % (ratio ^b)	mUPD WC % (ratio ^b)	DEL WC % (ratio ^b)	INV % (ratio ^b)	WC % (ratio ^b)
Imagination	66.7% (4/6)	0% (0/4)	50% (3/6)	0% (0/2)	40% (4/10)	37.5% (3/8)
Organization	33.3% (2/6)	0% (0/4)	0% (0/6)	0% (0/2)	20% (2/10)	0% (0/8)
Affect Frequency	66.7% (4/6)	25% (1/4)	16.7% (1/6)	0% (0/2)	50% (5/10)	12.5% (1/8)
Affect Variety	66.7% (4/6)	0% (0/4)	66.7% (4/6)	0% (0/2)	40% (4/10)	50% (4/8)
No Play ^a	50% (3/6)	0% (0/4)	16.7% (1/6)	0% (0/2)	30% (3/10)	12.5% (1/8)
Functional Play	66.7% (4/6)	25% (1/4)	33.3% (2/6)	0% (0/2)	50% (5/10)	25% (2/8)
Symbolic Play	83.3% (5/6)	25% (1/4)	50% (3/6)	0% (0/2)	60% (6/10)	37.5% (3/8)

^a Decreases = improvement

^b Ratio indicates how many children improved relative to number of children in each subgroup

improvement above and beyond that of the mUPD WC participants (6 participants) across variables of cognitive play, affective play, and increased time spent in symbolic play, whereas very few children in the DEL INV group (4 participants) made improvements relative to the DEL WC group (2 participants). Overall, results from this individual, descriptive analysis indicate that for children who underwent the PRETEND-Preschool intervention in Study 2, with live-coaching of parent-child play alongside parent-training sessions, a greater percentage of children in the mUPD subgroup showed improvement across all variables of pretend play. Further analyses of baseline levels of parent-reported stress and parent-child dyadic play indicated significant relationships among multiple variables for the DEL group, whereas the mUPD group had no observed significant relationships between reported parent stress and parent-child play.

Discussion

Collectively, findings from Study 1 and Study 2 on the preliminary efficacy of the PRETEND-Preschool program indicate that this remote, parent-training, play-based intervention did not lead to improvements in play skills for children when they were not involved in the intervention, but involving children via parent-child play into the intervention led to significant improvements in cognitive and affective play skills for some of the children with PWS. Specifically, in Study 2, the addition of live coaching-play sessions between parents and children facilitated significant improvements for participants with mUPD who received the intervention. Conversely, participants in the DEL group did not appear to make any significant improvements in play skills following the modified intervention in Study 2.

Previous research has shown that the ability for parent and child to interact and practice skills together leads to more persistent behavioral change for children with developmental disabilities, and that parent-training can be used feasibly and effectively over telehealth (Scahill et al., 2016; Bearss et al., 2018). Given this, we expect that the addition of the coaching-play sessions for parent-child play in Study 2 may have led to the observed improvements in play skills observed for the participants with mUPD who underwent the intervention. There may be several reasons why a portion of the participants responded to the PRETEND-Preschool program in Study 2 rather than in Study 1. First, the coaching-play sessions allowed for a controlled dosage of practice where parents and children had four built-in opportunities in the 8-week program to play together and practice pretend playing. Second, the coaching-play sessions provided a space in which a parent could utilize and expand

upon their pretend play repertoire with guided communication from the play interventionist. Parents were taught in their parent-training sessions how to use modeling and scaffolding as key components for demonstrating and increasing pretend play skills for their children, and also how to use positive reinforcement to build upon adaptive skills the child exhibits in the play sessions. By practicing these skills with a play interventionist there for support and prompting, the children were likely able to engage more fully in interactions with parents and practice skills more successfully, perhaps then leading to the improvements found in the current study. Third, the coaching-play sessions had a play interventionist present, and therefore the parent-child sessions echoed the skill-building that trained play interventionists have used successfully directly with school-aged children in other studies (Dimitropoulos et al., 2021; Doernberg et al., 2021). Taken together, it can be suggested that these coaching-play sessions were the likely mechanism of change for the PRETEND-Preschool intervention in Study 2, given the lack of any improvement in Study 1 where all other procedures were kept the same. Thus, a main takeaway from the current study suggests that it appears to be necessary to incorporate child involvement via parent-child play in addition to the parent-training sessions.

It is important to consider why the DEL group did not respond to the modified intervention in Study 2. Notably, parents of the DEL group reported significantly higher stress at baseline, which related to their scores on a measure of baseline parent-child dyadic play. These findings are consistent with previous research indicating parenting stress is associated with poorer parent-child interaction in play (Zyga and Dimitropoulos, 2020), and indicates that dyadic play skills between parents and children may be negatively impacted by parenting distress in our sample. For the DEL group in Study 2, given that poorer parent-child dyadic play was related to increased parenting stress, it may indicate that coaching-play sessions were not as effective, and therefore participants with DEL did not make improvements in the INV group. In addition, the increased number of problem behaviors for the DEL group may serve as a barrier towards increases in play skills during the parent-child play sessions. Finally, the DEL group in Study 2 had greater social cognitive impairments compared to the mUPD group at baseline. This finding differs from the baseline characteristics of the groups from Study 1, wherein participants with DEL had fewer social-cognitive impairments at baseline. Collective research indicates that children with the DEL subtype of PWS typically have increased levels of social cognitive ability compared with the mUPD subtype (Key et al., 2013; Dimitropoulos et al., 2013; Whittington and Holland, 2017). Therefore, the findings of greater social cognitive impairments in the DEL group in Study 2 are inconsistent with

the existing literature, and are likely specific to this current sample. In general, we would expect that all children would respond to specific techniques used in the PRETEND-Preschool intervention, such as the guided coaching sessions. However, as previous and current research has shown, there may be a difference in social cognitive skills, cognitive functioning, behavioral difficulties, and parental characteristics such as parent stress between the two main PWS subtypes (Whittington et al., 2010, Hogart et al., 2010, Hartley et al., 2005; Ihara et al., 2014). This may suggest that interventions for this population may need to be tailored for each subtype in order to see similar improvements in functioning.

There are several important implications of these findings. By examining results across both studies, it appears that the addition of live coaching-play sessions for parent-child play was an important mechanism of change to facilitate improvement following the play intervention. While the parent-training sessions remained consistent from Study 1 to Study 2 in terms of their content, the addition of parent-child play sessions to the intervention led to significant improvements for participants with mUPD who underwent the intervention. The controlled parent-child play provided a platform wherein children were more actively involved in the study, and parents could refine their learned play techniques with live coaching from a trained interventionist. These results suggest that learning the play techniques through parent-training is not sufficient for preschool children with PWS to gain skills, but the controlled and monitored practice of play directly with the children may be what facilitates improvements. As demonstrated with the success of the PRETEND program administered to school-age children with PWS (Dimitropoulos et al., 2021), direct involvement of the children within the play training appears to be important. Additionally, the PRETEND-Preschool program may be less effective for parents who are experiencing greater distress, and possibly also less effective for children with increased problem behaviors and greater social cognitive impairments. The impact of PWS caregiver burden and parenting stress is a recent growing area of research in the field (Kayadjanian et al., 2018; Mao et al., 2019; Wong et al., 2021), and it may be a barrier to proper implementation of the present intervention program. Further investigation into these areas will be important in refining the current intervention in increasing efficacy across both subtype groups.

There were several important limitations to the present study. First, small sample sizes limited the number of quantitative analyses that could be performed, especially for subtype analyses. Given that the mUPD group had data from only 12 participants (6 in INV and 6 in WC) and the DEL group had data from only 6 participants (4 in INV and 2 in WC) for pre-post analyses, quantitative comparisons within these groups were very limited in power and should

be interpreted with caution. Given the small sample size, it is possible that variance in baseline characteristics became confounding variables that may have been interfering with the direct comparison of intervention efficacy between the groups. Second, the small sample lacked diversity, making it more challenging to generalize the results of the study to a larger population of preschool children with PWS. Finally, given that the only pre-post variable evaluated was pretend play skills in children, results from the present study cannot determine whether change in pretend play abilities are meaningful outside of this context.

Conclusions

The present study indicated that the PRETEND-Preschool intervention was impactful for some children with PWS, allowing for significant increases in their cognitive and affective skills in play. There are several important areas of research that should be investigated to refine and improve this intervention to increase efficacy for all children with PWS. First, future studies should focus on the extension of the PRETEND-Preschool program to a wider, more representative sample of preschool children with PWS, with greater sample size allowing for more rigorous analysis of potential subtype effects. Second, future research should also include additional measures of emotional, social, cognitive, and behavioral functioning pre-post intervention, to better understand how improvements in pretend play may transfer to other improvements in more generalized developmental areas. Finally, given the barrier that parenting stress appeared to have on parent-child play and improvement child play skills overall in the DEL group, it is essential that accommodations be put in place in the PRETEND-Preschool intervention to identify and mitigate parenting stress in PWS, and improve parent-child engagement throughout the study. Future studies should also investigate other potential barriers to amelioration from the PRETEND-Preschool intervention, such as social cognitive impairments, child problem-behaviors, and specifically cognitive and behavioral rigidity across subtypes. Despite the limitations from the present study, the PRETEND-Preschool intervention is a promising avenue of intervention for parents of children with PWS, that can be successfully administered remotely via telehealth to increase access to treatment for any child regardless of location or SES.

References

- Angulo, M. A., Butler, M. G., & Cataletto, M. E. (2015). Prader-Willi syndrome: a review of clinical, genetic, and endocrine findings. *Journal of endocrinological investigation*, 38(12), 1249–1263

- Barnett, L. A. (1990). Developmental benefits of play for children. *Journal of Leisure Research*, 22(2), 138–153
- Barton, E. E., & Wolery, M. (2008). Teaching pretend play to children with disabilities: A review of the literature. *Topics in Early Childhood Special Education*, 28(2), 109–125
- Barton, E. E., & Wolery, M. (2010). Training teachers to promote pretend play in young children with disabilities. *Exceptional Children*, 77(1), 85–106
- Bennett, J. A., Germani, T., Haqq, A. M., & Zwaigenbaum, L. (2015). Autism spectrum disorder in Prader–Willi syndrome: a systematic review. *American Journal of Medical Genetics Part A*, 167(12), 2936–2944
- Bodrova, E., Germeroth, C., & Leong, D. J. (2013). Play and self-regulation: lessons from Vygotsky. *American journal of play*, 6(1), 111–123
- Butler, M. G., Miller, J. L., & Forster, J. L. (2019). Prader-Willi syndrome-clinical genetics diagnosis and treatment approaches: an update. *Current pediatric reviews*, 15(4), 207–244
- Charman, T. (2003). Why is joint attention a pivotal skill in autism? *Philosophical Transactions of the Royal Society of London. Series B: Biological Sciences*, 358(1430), 315–324
- Cox, N. S., Alison, J. A., Rasekaba, T., & Holland, A. E. (2012). Telehealth in cystic fibrosis: a systematic review. *Journal of telemedicine and telecare*, 18(2), 72–78
- Dansky, J. L. (1999). “Play.” In *Encyclopedia of Creativity*, edited by Mark A. Runco and Steven, R., & Pritzker, 393–408
- Debladis, J., Valette, M., Strenilkov, K., Mantoulan, C., Thuilleaux, D., & Laurier, V., ... Tauber M. (2019) Face processing and exploration of social signals in Prader-Willi syndrome: a genetic signature. *Orphanet journal of rare diseases*, 14(1), 1–13
- Dimitropoulos, A., Ho, A., & Feldman, B. (2013). Social responsiveness and competence in Prader-Willi syndrome: Direct comparison to autism spectrum disorder. *Journal of autism and developmental disorders*, 43(1), 103–113
- Dimitropoulos, A., Zyga, O., & Russ, S. W. (2019). Early social cognitive ability in preschoolers with Prader–Willi syndrome and autism spectrum disorder. *Journal of autism and developmental disorders*, 49(11), 4441–4454
- Dimitropoulos, A., Zyga, O., Doernberg, E., & Russ, S. W. (2021). Show me what happens next Preliminary efficacy of a remote play-based intervention for children with Prader-Willi syndrome. *Research in Developmental Disabilities*, 108, 103820
- Doernberg, E. A., Russ, S. W., & Dimitropoulos, A. (2021). Believing in Make-Believe: Efficacy of a Pretend Play Intervention for School-Aged Children with High-Functioning Autism Spectrum Disorder. *Journal of Autism and Developmental Disorders*, 51(2), 576–588
- Dunn, L. M., & Dunn, D. M. (2007). *PPVT-4: Peabody picture vocabulary test*. Pearson Assessments.
- Dykens, E. M., Roof, E., & Hunt-Hawkins, H. (2017). Cognitive and adaptive advantages of growth hormone treatment in children with Prader-Willi syndrome. *Journal of Child Psychology and Psychiatry*, 58(1), 64–74
- Dykens, E. M., Roof, E., Hunt-Hawkins, H., Daniell, C., & Jurgensmeyer, S. (2019). Profiles and trajectories of impaired social cognition in people with Prader-Willi syndrome. *PloS one*, 14(10), e0223162
- Fairchild, R. M., Ferng-Kuo, S. F., Rahmouni, H., & Hardesty, D. (2020). Telehealth increases access to care for children dealing with suicidality, depression, and anxiety in rural emergency departments. *Telemedicine and e-Health*, 26(11), 1353–1362
- Fehr, K. K., & Russ, S. W. (2014). Assessment of pretend play in preschool-aged children validation and factor analysis of the affect in play scale–preschool version. *Journal of Personality Assessment*, 96(3), 350–357
- Fehr, K. K., & Russ, S. W. (2016). Pretend play and creativity in preschool-age children Associations and brief intervention. *Psychology of Aesthetics, Creativity, and the Arts*, 10(3), 296
- Fein, G. G. (1981). Pretend play in childhood: An integrative review. *Child development*, 1095–1118
- Feldman, B. H., & Dimitropoulos, A. (2014). Face discrimination skills in Prader-Willi syndrome and autism Spectrum disorder. *Journal of Mental Health Research in Intellectual Disabilities*, 7(3), 264–285.
- French, J. L. (2001). *Pictorial Test of Intelligence–Second Edition (PTI-2)*.
- Greenspan, S. I., & Wieder, S. (1997). Developmental patterns and outcomes in infants and children with disorders in relating and communicating: A chart review of 200 cases of children with autistic spectrum diagnoses. *Journal of Developmental and Learning disorders*, 1, 87–142.
- Gresham, F. M., Elliott, S. N., Vance, M. J., & Cook, C. R. (2011). Comparability of the Social Skills Rating System to the Social Skills Improvement System: Content and psychometric comparisons across elementary and secondary age levels. *School Psychology Quarterly*, 26(1), 27.
- Harris, P. L. (2000). *The work of the imagination*. Blackwell Publishing.
- Hartley, S. L., MacLean Jr, W. E., Butler, M. G., Zarccone, J., & Thompson, T. (2005). Maladaptive behaviors and risk factors among the genetic subtypes of Prader–Willi syndrome. *American Journal of Medical Genetics Part A*, 136(2), 140–145.
- Hogart, A., Wu, D., LaSalle, J. M., & Schanen, N. C. (2010). The comorbidity of autism with the genomic disorders of chromosome 15q11. 2-q13. *Neurobiology of disease*, 38(2), 181–191.
- Hudson, J. L., & Rapee, R. M. (2001). Parent–child interactions and anxiety disorders: an observational study. *Behaviour research and therapy*, 39(12), 1411–1427.
- Ihara, H., Ogata, H., Sayama, M., Kato, A., Gito, M., Murakami, N., ... Nagai, T. (2014). QOL in caregivers of Japanese patients with Prader–Willi syndrome with reference to age and genotype. *American Journal of Medical Genetics Part A*, 164(9), 2226–2231.
- Jeffrey, J., Marlotte, L., & Hajal, N. J. (2020). Providing telebehavioral health to youth and families during COVID-19: Lessons from the field. *Psychological Trauma: Theory, Research, Practice, and Policy*, 12(S1), S272.
- Johnson, A. O. (2015). Test review: parenting stress index, (PSI-4).
- Kasari, C., Freeman, S., & Paparella, T. (2006). Joint attention and symbolic play in young children with autism: A randomized controlled intervention study. *Journal of Child Psychology and Psychiatry*, 47(6), 611–620.
- Kasari, C., Gulsrud, A., Paparella, T., Hellemann, G., & Berry, K. (2015). Randomized comparative efficacy study of parent-mediated interventions for toddlers with autism. *Journal of consulting and clinical psychology*, 83(3), 554.
- Kaugars, A. S., & Russ, S. W. (2009). Assessing preschool children’s pretend play: Preliminary validation of the Affect in Play Scale–Preschool version. *Early Education and Development*, 20(5), 733–755.
- Kayadjanian, N., Schwartz, L., Farrar, E., Comtois, K. A., & Strong, T. V. (2018). High levels of caregiver burden in Prader-Willi syndrome. *PloS one*, 13(3), e0194655.
- Key, A. P., & Dykens, E. M. (2017). Incidental memory for faces in children with different genetic subtypes of Prader-Willi syndrome. *Social cognitive and affective neuroscience*, 12(6), 918–927.
- Krasnor, L. R., & Pepler, D. J. (1980). The study of children’s play: Some suggested future directions. *New Directions for Child and Adolescent Development*, 1980(9), 85–95.
- Lifter, K., & Bloom, L. (1989). Object knowledge and the emergence of language. *Infant Behavior and Development*, 12(4), 395–423.

- Lifter, K., Mason, E. J., & Barton, E. E. (2011). Children's play: Where we have been and where we could go. *Journal of Early Intervention*, 33(4), 281–297.
- Lo, S. T., Siemensma, E., Collin, P., & Hokken-Koelega, A. (2013). Impaired theory of mind and symptoms of autism spectrum disorder in children with Prader–Willi syndrome. *Research in developmental disabilities*, 34(9), 2764–2773.
- Mao, S. J., Shen, J., Xu, F., & Zou, C. C. (2019). Quality of life in caregivers of young children with Prader–Willi syndrome. *World Journal of Pediatrics*, 15(5), 506–510.
- McDonald, N. M., Baker, J. K., & Messinger, D. S. (2016). Oxytocin and parent–child interaction in the development of empathy among children at risk for autism. *Developmental psychology*, 52(5), 735.
- McGeary, D. D., McGeary, C. A., & Gatchel, R. J. (2012). A comprehensive review of telehealth for pain management: where we are and the way ahead. *Pain Practice*, 12(7), 570–577.
- Mercer, J. (2017). Examining DIR/Floortime™ as a treatment for children with autism spectrum disorders: A review of research and theory. *Research on Social Work Practice*, 27(5), 625–635.
- Miller, J. L., Lynn, C. H., Driscoll, D. C., Goldstone, A. P., Gold, J. A., Kimonis, V., ... Driscoll, D. J. (2011). Nutritional phases in Prader–Willi syndrome. *American journal of medical genetics Part A*, 155(5), 1040–1049.
- Moore, M., & Russ, S. W. (2008). Follow-up of a pretend play intervention: Effects on play, creativity, and emotional processes in children. *Creativity Research Journal*, 20(4), 427–436.
- Mullen, E. M. (1995). *Mullen scales of early learning* (pp. 58–64). Circle Pines, MN: AGS.
- Rice, L. J., Woodcock, K., & Einfeld, S. L. (2018). The characteristics of temper outbursts in Prader–Willi syndrome. *American Journal of Medical Genetics Part A*, 176(11), 2292–2300.
- Rosner, B. A., Hodapp, R. M., Fidler, D. J., Sagun, J. N., & Dykens, E. M. (2004). Social competence in persons with Prader–Willi, Williams and Down's Syndromes. *Journal of Applied Research in Intellectual Disabilities*, 17(3), 209–217.
- Russ, S. W., & American Psychological Association. (2014). *Pretend play in childhood: Foundation of adult creativity* (pp. 45–62). Washington, DC: American Psychological Association.
- Rutter, M., Bailey, A., & Lord, C. (2003). SCQ. *The Social Communication Questionnaire*. Torrance, CA: Western Psychological Services.
- Scahill, L., Bearss, K., Lecavalier, L., Smith, T., Swiezy, N., Aman, M. G., ... Johnson, C. 2016. Effect of parent training on adaptive behavior in children with autism spectrum disorder and disruptive behavior: results of a randomized trial. *Journal of the American Academy of Child & Adolescent Psychiatry*, 55(7), 602–609.
- Singer, D. G., & Singer, J. L. (1992). *The house of make believe: Children's play and the developing imagination*. Cambridge, MA: Harvard University Press
- Solomon, R., Van Egeren, L. A., Mahoney, G., Huber, M. S. Q., & Zimmerman, P. (2014). PLAY Project Home Consultation intervention program for young children with autism spectrum disorders: a randomized controlled trial. *Journal of Developmental and Behavioral Pediatrics*, 35(8), 475.
- Strain, P. S., Hoyson, M., & Jamieson, B. (1985). Normally developing preschoolers as intervention agents for autistic-like children: Effects on class department and social interaction. *Journal of the Division for Early Childhood*, 9(2), 105–115.
- Tomanik, S., Harris, G. E., & Hawkins, J. (2004). The relationship between behaviours exhibited by children with autism and maternal stress. *Journal of Intellectual and Developmental Disability*, 29(1), 16–26.
- Tonge, B., Brereton, A., Kiomall, M., Mackinnon, A., & Rinehart, N. J. (2014). A randomised group comparison controlled trial of 'preschoolers with autism': A parent education and skills training intervention for young children with autistic disorder. *Autism*, 18(2), 166–177.
- Twum, F., & Hayford, S. K. Motor Skills as Predictors of Intellectual ability of Pupils with Cerebral Palsy.
- Vismara, L. A., McCormick, C., Young, G. S., Nadhan, A., & Monlux, K. (2013). Preliminary findings of a telehealth approach to parent training in autism. *Journal of autism and developmental disorders*, 43(12), 2953–2969.
- Wallace, C. E. (2018). *Improving the parent-child relationship in ADHD: A pretend playintervention* (Doctoral dissertation, Case Western Reserve University).
- Whittington, J., & Holland, A. (2010). Neurobehavioral phenotype in Prader–Willi syndrome. In *American Journal of Medical Genetics Part C: Seminars in Medical Genetics* (Vol. 154, No. 4, pp. 438–447). Hoboken: Wiley Subscription Services, Inc., A Wiley Company.
- Whittington, J., & Holland, T. (2011). Recognition of emotion in facial expression by people with Prader–Willi syndrome. *Journal of Intellectual Disability Research*, 55(1), 75–84.
- Whittington, J., & Holland, A. (2017). Cognition in people with Prader–Willi syndrome: Insights into genetic influences on cognitive and social development. *Neuroscience & Biobehavioral Reviews*, 72, 153–167.
- Williams, M. E., Sando, L., & Soles, T. G. (2014). Cognitive tests in early childhood: Psychometric and cultural considerations. *Journal of Psychoeducational Assessment*, 32(5), 455–476.
- Wong, S. B., Wang, T. S., Tsai, W. H., Tzeng, I. S., & Tsai, L. P. (2021). Parenting stress in families of children with Prader–Willi syndrome. *American Journal of Medical Genetics Part A*, 185(1), 83–89.
- Yates, T. M., & Marcelo, A. K. (2014). Through race-colored glasses: Preschoolers' pretend play and teachers' ratings of preschooler adjustment. *Early Childhood Research Quarterly*, 29(1), 1–11.
- Zyga, O., Russ, S., Ievers-Landis, C. E., & Dimitropoulos, A. (2015). Assessment of pretend play in Prader–Willi syndrome: A direct comparison to autism spectrum disorder. *Journal of autism and developmental disorders*, 45(4), 975–987.
- Zyga, O., Russ, S. W., & Dimitropoulos, A. (2018). The PRETEND Program: Evaluating the feasibility of a remote parent-training intervention for children with Prader–Willi syndrome. *American journal on intellectual and developmental disabilities*, 123(6), 574–584.
- Zyga, O., & Dimitropoulos, A. (2020). Preliminary Characterization of Parent–Child Interaction in Preschoolers with Prader–Willi Syndrome: The Relationship Between Engagement and Parental Stress. *American journal on intellectual and developmental disabilities*, 125(1), 76–84.

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