frontiers | Research Topics

Emotion regulation in neurodevelopmental disorders: Current understanding and treatments

Edited by
Ru Ying Cai, Andrea C. Samson and Mirko Uljarevic

Published in
Frontiers in Psychiatry

ISSN 1664-8714
ISBN 978-2-8325-5308-4
DOI 10.3389/978-2-8325-5308-4

About Frontiers

Frontiers is more than just an open access publisher of scholarly articles: it is a pioneering approach to the world of academia, radically improving the way scholarly research is managed. The grand vision of Frontiers is a world where all people have an equal opportunity to seek, share and generate knowledge. Frontiers provides immediate and permanent online open access to all its publications, but this alone is not enough to realize our grand goals.

Frontiers journal series

The Frontiers journal series is a multi-tier and interdisciplinary set of open-access, online journals, promising a paradigm shift from the current review, selection and dissemination processes in academic publishing. All Frontiers journals are driven by researchers for researchers; therefore, they constitute a service to the scholarly community. At the same time, the *Frontiers journal series* operates on a revolutionary invention, the tiered publishing system, initially addressing specific communities of scholars, and gradually climbing up to broader public understanding, thus serving the interests of the lay society, too.

Dedication to quality

Each Frontiers article is a landmark of the highest quality, thanks to genuinely collaborative interactions between authors and review editors, who include some of the world's best academicians. Research must be certified by peers before entering a stream of knowledge that may eventually reach the public - and shape society; therefore, Frontiers only applies the most rigorous and unbiased reviews. Frontiers revolutionizes research publishing by freely delivering the most outstanding research, evaluated with no bias from both the academic and social point of view. By applying the most advanced information technologies, Frontiers is catapulting scholarly publishing into a new generation.

What are Frontiers Research Topics?

Frontiers Research Topics are very popular trademarks of the *Frontiers journals series*: they are collections of at least ten articles, all centered on a particular subject. With their unique mix of varied contributions from Original Research to Review Articles, Frontiers Research Topics unify the most influential researchers, the latest key findings and historical advances in a hot research area.

Find out more on how to host your own Frontiers Research Topic or contribute to one as an author by contacting the Frontiers editorial office: frontiersin.org/about/contact

Emotion regulation in neurodevelopmental disorders: Current understanding and treatments

Topic editors

Ru Ying Cai — Autism Spectrum Australia, Australia
Andrea C. Samson — Unidistance Suisse, Switzerland
Mirko Uljarevic — The University of Melbourne, Australia

Citation

Cai, R. Y., Samson, A. C., Uljarevic, M., eds. (2024). *Emotion regulation in neurodevelopmental disorders: Current understanding and treatments*. Lausanne: Frontiers Media SA. doi: 10.3389/978-2-8325-5308-4

Table of contents

frontiers | Frontiers in Psychiatry

TYPE Editorial
PUBLISHED 31 July 2024
DOI 10.3389/fpsyt.2024.1467834

OPEN ACCESS

EDITED AND REVIEWED BY
Antonio M. Persico,
University of Modena and Reggio Emilia, Italy

*CORRESPONDENCE
Ru Ying Cai
ryingcai@autismspectrum.org.au

RECEIVED 21 July 2024
ACCEPTED 23 July 2024
PUBLISHED 31 July 2024

CITATION
Cai RY, Samson AC and Uljarevic M (2024) Editorial: Emotion regulation in neurodevelopmental disorders: current understanding and treatments.
Front. Psychiatry 15:1467834.
doi: 10.3389/fpsyt.2024.1467834

Editorial: Emotion regulation in neurodevelopmental disorders: current understanding and treatments

Ru Ying Cai[1,2]*, Andrea C. Samson[3,4] and Mirko Uljarevic[2,5,6]

[1]Aspect Research Centre for Autism Practice, Autism Spectrum Australia, Melbourne, VIC, Australia, [2]Olga Tennison Autism Research Centre, La Trobe University, Melbourne, VIC, Australia, [3]Faculty of Psychology, UniDistance Suisse, Brig, Switzerland, [4]Department of Special Education, Université de Fribourg, Fribourg, Switzerland, [5]Psychiatry and Behavioral Sciences, Stanford University, Stanford, CA, Switzerland, [6]Melbourne School of Psychological Sciences, The University of Melbourne, Parkville, VIC, Australia

KEYWORDS

emotion regulation, neurodevelopmental disorders, intervention, mechanism, mental health

Editorial on the Research Topic
Emotion regulation in neurodevelopmental disorders: current understanding and treatments

Emotion regulation is a transdiagnostic process that plays a pivotal role in the development and maintenance of internalizing and externalizing symptoms such as anxiety, depression, or conduct problems. Difficulty with emotion regulation is a prominent feature of neurodevelopmental conditions such as autism spectrum disorder (ASD) and attention deficit hyperactivity disorder (ADHD), as well as neurogenetic syndromes. This Research Topic, *Emotion Regulation in Neurodevelopmental Disorders: Current Understanding and Treatments*, aimed to bring together theoretical insights, methodological developments, and novel empirical findings of emotion regulation in individuals diagnosed with neurodevelopmental disorders. In total, nine articles were published. These papers covered a range of important topics that could be broadly organized into studies focused on the role and mechanisms underlying emotion regulation difficulties and trials evaluating the effectiveness of different intervention modalities on emotion regulation-related outcomes. It was particularly encouraging that one study focused on measurement development and that several studies (both mechanistic and intervention-focused) were conducted with individuals with neurogenetic syndromes. Below, we provide a brief overview of the main findings of the published studies.

Studies focused on interventions related to emotion regulation

A single-subject design study involving four children with Williams Syndromes enrolled in play- and humor-infused exposure therapy approach showed preliminary evidence for improvements in emotion regulation (Young et al.). More specifically, at the

conclusion of the brief intervention, children with Williams Syndrome who presented with baseline co-occurring fears and phobias showed improved tolerance or the ability to engage positively with the previously real-world feared stimulus. A study by Famelart et al. evaluated EMO-T, an intervention aimed at improving emotion expression, recognition, comprehension, and regulation, hypothesized as skills essential for the healthy regulation of emotions that, if impaired, could result in dysregulation difficulties commonly observed in children with Prader–Willi Syndrome (PWS). In addition to improvements in voluntary expression and emotion recognition abilities, school-aged children with PWS enrolled in EMO-T also showed improvements in emotion regulation based on the parent-reported Emotion Regulation Checklist. Enav et al. evaluated whether participation in a four-week reflective/mentalization-based parenting workshop can enhance emotion regulation in parents of children with ASD. Authors specifically focused on different aspects of cognitive reappraisal given its adaptive role in reducing levels of stress, a particularly pertinent issue for parents of children with autism. Relative to the baseline, parents reported higher levels of cognitive reappraisal, as indexed by the Emotion Regulation Questionnaire, as well as higher levels of reflective (but not non-reflective) reappraisal on the Emotion Interaction Questionnaire. Finally, in a large sample of 94 children with attention-deficit/hyperactivity disorder (ADHD), Groves et al. reported that two digital training approaches focused on working memory (Central Executive Training) and inhibitory control (Inhibitory Control Training) both resulted in a significant reduction in emotion dysregulation (measured by the Emotion Dysregulation Inventory) at immediate post-treatment, at 1–2 month and 2–4-month follow-ups.

Studies focused on characterization and the role of emotion regulation

In the only study specifically focused on measurement development, Uljarević et al. described the development, refinement, and initial psychometric evaluation of a new open-source 52-item Executive Functioning Scale (EFS) that was specifically developed to capture theoretically-based facets of executive functioning (EF), including working memory and sequencing, response inhibition, set-shifting, processing speed, risk avoidance, and crucially, emotion regulation. Across two independent data collections encompassing 2,958 children and adolescents, the hypothesized six subscales were derived and confirmed, showing strong classic test and conditional reliability as well as invariance across age, sex, race, and ethnicity groups. Walter et al. explored the association between affective language, as measured by the verbal fluency test, with emotion dysregulation, indexed by the cyclothymic dimension of the Temperament Evaluation of Memphis questionnaire, in adults with ADHD, ASD with co-occurring ADHD, and neurotypical controls. Findings provided very preliminary suggestions that factors related to emotional difficulties in individuals with ADHD and with ASD with co-occurring ADHD might be at least somewhat distinct. More specifically, compared to adults with ADHD and neurotypical controls, individuals with ASD and ADHD produced fewer anger-related words, produced more emotions, and fewer rule breaks. Authors reported associations with emotion regulation only in adults with ADHD, finding that emotional over-reactivity in adults with ADHD was associated with the number of emotions and the frequency of these words. Comparing the effects of the COVID-19 pandemic between 4,138 children with ASD and 711 children with developmental delays (DD), Zhao et al. found that children with ASD had a higher risk of having more emotional and behavioral problems than children with DD. De Blasio et al. explored olfactory processing in 20 youth with profound intellectual and multiple disabilities, demonstrating preserved olfactory preferences in this population and, importantly, that olfactory preferences were associated with mood levels. Samson et al. aimed to characterize emotion regulation strategy use and its associations with anxiety use in individuals with ASD with (n=785) and without ID (n=596), Williams Syndrome (n=261), and Intellectual Disability not otherwise specified (ID-NOS; n=649). Distinct emotion regulation use was identified such that individuals with Williams Syndrome most frequently used parent routine, parent shielding, repetitive behaviors, and distraction; individuals with ASD without ID most frequently utilized isolation/withdrawal; and in general, individuals with ASD and ID and with ID-NOS engaged significantly less frequently in cognitive emotion regulation strategies compared to other groups. Importantly, across all groups, anxiety was linked to a higher use of maladaptive and a lower use of positive strategies.

Conclusion

The research studies included in this Research Topic provided important insights into the transdiagnostic nature of emotion regulation across various neurodevelopmental conditions, providing important data that can be used across both research and clinical contexts. Crucially, several key directions for future research have emerged, including the need for a more careful conceptualization of emotion regulation and the development of new multimodal assessment protocols. Additionally, improving the design of interventions and utilizing adaptive trial frameworks are essential steps toward precision medicine for improving the mental health of people with neurodevelopmental conditions.

Author contributions

RC: Writing – review & editing. AS: Writing – review & editing. MU: Writing – original draft.

Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

Publisher's note

frontiers | Frontiers in Psychiatry

TYPE Original Research
PUBLISHED 17 October 2022
DOI 10.3389/fpsyt.2022.995669

OPEN ACCESS

EDITED BY
Andrea C. Samson,
Unidistance Suisse, Switzerland

REVIEWED BY
Ulrike Rimmele,
Université de Genève, Switzerland
Carolyn Shivers,
Virginia Tech, United States

*CORRESPONDENCE
Yael Enav
yaelenav12@gmail.com

SPECIALTY SECTION
This article was submitted to Autism, a section of the journal Frontiers in Psychiatry

RECEIVED 16 July 2022
ACCEPTED 20 September 2022
PUBLISHED 17 October 2022

CITATION
Enav Y, Hardan AY and Gross JJ (2022) Cognitive reappraisal training for parents of children with autism spectrum disorder.
Front. Psychiatry 13:995669.
doi: 10.3389/fpsyt.2022.995669

Cognitive reappraisal training for parents of children with autism spectrum disorder

Yael Enav[1]*, Antonio Y. Hardan[2] and James J. Gross[2]

[1]Department of Counseling and Human Development, University of Haifa, Haifa, Israel, [2]Department of Psychology, Stanford University, Stanford, CA, United States

Parents of children with autism spectrum disorder (ASD) experience higher stress levels than those of typically developing children. The goal of the current study was to examine whether a mentalization-based intervention would enhance parental cognitive reappraisal, an adaptive form of emotion regulation associated with lower levels of stress. Findings from 27 parents who completed a short training indicated an improvement in cognitive reappraisal. In exploratory analyses, two different types of reappraisal were examined. The intervention-related improvement was found mainly with one type of reappraisal, namely reflective reappraisal that consist of cognitive reappraisal with mentalization characteristics. In light of the evidence indicating that high cognitive reappraisal and high reflective functioning are associated with quality caregiving, findings from the current study suggesting that a brief mentalization-based intervention supports ASD parents' cognitive reappraisal with mentalization characteristics are promising and warrant further investigation.

KEYWORDS

autism, parenting, cognitive reappraisal, reflective functioning, intervention

Introduction

Parents of children with autism spectrum disorder (ASD) experience higher stress levels than parents of children with other neurodevelopmental disorders or children who are typically developing (1–5). Therefore, intervention programs may need to address parental stress, which in turn will benefit the child and the family. Effective emotion regulation (ER) strategies may reduce stress and other negative emotions and increase high-quality parent-child interactions (6, 7).

Cognitive reappraisal

Because different forms of ER have different consequences, it is important to be specific about the types of ER that are (or are not) used in a particular family context. One particularly adaptive form of ER is cognitive reappraisal, a type of cognitive change ER (8, 9). Cognitive reappraisal has been found to change emotions in a sustained manner, and has been associated with lower levels of negative affect (10, 11). Successful cognitive reappraisal leads to better interpersonal functioning along with physical and

psychological well-being (12, 13) and fewer mental health problems (14). In light of its positive effects, there has been a growing interest in the processes that support cognitive reappraisal.

Mentalization and cognitive reappraisal

Mentalizing (also known as reflective functioning) refers to the creation of explanations of one's own or others' mental states, including thoughts, feelings, and intentions during an act of imagining and wondering (15). Mentalizing the experiences of self and others supports emotion regulation (16) and helps transform initial maladaptive thoughts into adaptive ones (15, 17). While mentalizing and wondering about others' states of mind, we imagine their perspective. Considering another person's point of view requires us to pay attention to and actively imagine that person's perspective (18), and this, in turn, promotes the understanding that each mind works differently and helps us become regulated and empathetic (19, 20). Mentalization may play a particularly central role in families of children with ASD, as it is hard to understand the children's thoughts and intentions, and often mentalization lacks a positive response from the child (21).

One reason mentalization may facilitate cognitive reappraisal is that reflective thinking initiates the development of complex representations and symbols (22, 23), leading to reappraisals with reflective characteristics (i.e., reflective appraisals) (24, 25). Reflective reappraisal is the process of reinterpretation of an event's meaning in order to down-regulate the experience of negative emotions that includes reflective characteristics (26, 27). In contrast, non-reflective reappraisals involve reinterpretations of an event's meaning that do not include reflective characteristics. For example: David, a 13-year-old, comes home from school angry and frustrated because he failed his math exam. When his dad asks for help setting the table for dinner, he shouts "Do it yourself" and slams his door. The first thought in his father's mind is: "He is so rude." After focusing on his son's mind and imagining his thoughts, feelings, and intentions, a reappraisal might be: "I know how hard he studied. It must be so frustrating for him to fail the math exam. He is not a rude child." In contrast, a non-reflective reappraisal could be: "He will calm down soon."

The scientific literature has increasingly recognized the relationship between mentalization and cognitive reappraisal (24, 28, 29). For instance, Baylin (28) found mentalization initiated cognitive reappraisal. In a related vein, Fonagy et al. (24) argued mentalization plays a role in appraising an event in a way that promotes resilience. Moreover, in aversive situations, the automatic response is usually a negative thought, and the individual needs to reappraise it in a way that involves reflective, cognitive mentalizing (30). Similarly, Sharp et al. (29) presented a model for trauma intervention that integrated mentalization and cognitive reappraisal.

The current study

This study is part of a larger study (31) that assessed the effect of mentalization based workshop on parental efficacy, parental belief in mailability of emotions and child's symptoms in parents of children with ASD. The goal of the current study was to examine whether and to what extent a mentalization-based intervention, bringing together aspects of psychodynamic and cognitive-behavioral focused on emotion regulation would affect parental cognitive reappraisal (the subjective interpretation made by the parent to an emotional stimuli) in parents of children with ASD. It was hypothesized that the intervention would lead to increased use of cognitive reappraisal, particularly reflective reappraisal.

Method

Participants

Parents of children with ASD were recruited to participate in a 4-week Reflective Parenting Workshop focused on emotion regulation (31). The present report is in an extension of a previous research, in the current investigation, the focus is on the effect of intervention on cognitive reappraisal.

Recruitment for the workshop occurred through distribution of fliers at clinics around the San Francisco Bay Area that provide services to families of children with autism. Twenty-seven intervention-group parents who completed an Emotion Interaction Questionnaire (EIQ) and ERQ (Emotion Regulation Questionnaire) before and after the workshop were included in the current study. Other inclusion criteria for the study included English proficiency, having a child with a diagnosis of autism spectrum disorder between the ages of 3 and 18 years, and completion of the workshop. This study was approved by the university's Institutional Review Board and registered in the Clinical Trials database. Table 1 presents participant demographics. As can be viewed from the table, all participating parents were married, and most were Caucasian women with two children. Over 80% of participants had academic degrees. One third were employed full-time, another one third were employed half-time, and the rest were either self-employed or homemakers. Approximately half of the children were between the ages 5 to 9, and half were between the ages 10 to 17. Most of the ASD children were high functioning.

Procedures

After signing consent forms, participants were screened for meeting the study inclusions criteria. Eligible participants then completed a 4-week face to face group workshop, one and a half hours once a week each session. Parenting workshop focused on reflective functioning and emotion regulation skills of parents

TABLE 1 Frequency distribution on participants' demographic characteristics.

		N (%)
Gender	Male	3 (11.1%)
	Female	24 (88.9%)
Marital status	Married	27 (100%)
Ethnicity	Caucasian	17 (63.0%)
	Pacific Islander	8 (29.6%)
	Other	2 (7.4%)
Number of children	Single child	3 (11.1%)
	Two children	19 (70.4%)
	Three children	4 (14.8%)
	Eight children	1 (3.7%)
Children's ages	5–9	14 (51.85)
	10–17	13 (48.15%)
Children's gender	Male	20 (74.07%)
	Female	7 (25.93%)
Children's functioning	Rating of 5	18 (66.67%)
	Rating of 4	2 (7.41%)
	Rating <= 3	6 (22.22%)
	Absent data	1 (3.70%)
Education	High school or GED	1 (3.7%)
	College or associate's degree	3 (11.1%)
	Bachelor's degree	6 (22.2%)
	Master's degree	13 (48.2%)
	PhD, MD, JD	3 (11.1%)
	Not mentioned	1 (3.7%)
Employment	Full-time	9 (33.3%)
	Part-time	9 (33.3%)
	Self-employed	2 (7.4%)
	Homemaker	6 (22.2%)

of children with ASD based on the literature that associate high parental reflective functioning with high emotional regulation and both are related to high quality caregiving and parent-child relationship (32, 33). The intervention flow and key concepts: Session 1 - emotion, emotion regulation, different emotion regulation strategies with focus on cognitive reappraisal. Session 2 – The effect of parental emotion regulation on children, reflective functioning and holding the mind in mind, the challenges and opportunities in raising children with ASD. Session 3 – Participants are invited to share their emotional scenarios from the past week with the group. Role play of different parent-child emotional scenarios. Session 4 - Participants are invited to share their emotional scenarios from the past week with the group. Participants are invited to share anything about their participation in the workshop with an emphasis on next steps, what they took from the workshop, and anything that was particularly helpful. The participants filled out pre and post questionnaires. Participants were asked to complete emotion interaction questionnaire (EIQ) and an emotion regulation questionnaire (ERQ) in its trait version. The ERQ can be used in two different versions: one version is asking about using cognitive reappraisal during a specific time frame that can be considered as more situational. The other version is asking about using cognitive reappraisal generally, in life, without asking about a specific timeframe. Being non-related to a certain period and or situation, but to a general truth, this version can be considered as more trait related.

Measures

For demographics, the following information was collected: age, gender, race, marital status, education status, and employment status. In addition, participants were asked to specify the number of children they have.

The Emotion Regulation Questionnaire (ERQ); (12) is self-report widely used measure of emotion regulation (34) and it was administered to assess parents' emotion regulation in trait format. It has good psychometric properties with above average internal consistency (Cronbach's α for reappraisal 0.75–0.82, and suppression 0.68–0.76, test–retest reliability across 3 months = 0.69), (12). The ERQ comprises 10 items divided into 2 subscales cognitive reappraisal and suppression. In this study we used the reappraisal subscale that consists of six items (e.g., "I control my emotions by changing the way I think about the situation I'm in"). Participants rated the degree to which they agreed with each statement on a 7-point scale (1 = strongly disagree, 7 = strongly agree).

The Emotion Interaction Questionnaire (EIQ, was developed by the research team) instructed parents first to describe a situation from the past week when they felt their negative emotions had an effect on their behavior, then to describe their reactions and thoughts during the situation and lastly to describe their thoughts after some time had passed.

Data reduction

Two judges were trained to assess cognitive reappraisal, as well as its two subtypes, namely reflective reappraisal and non-reflective reappraisal. Each judge got a detailed explanation with the definition of each subtype, examples of situations that were assessed and coded by the research team and demonstrated each subtype and five situations that the judges were asked to assess and code as part of the training. Cognitive reappraisal was defined as reinterpreting the meaning of an emotional event or stimulus with the goal of influencing one's emotional response (35). Reflective reappraisal was scored as being present when the criteria for cognitive reappraisal were met, and the statement showed one of the following: (1). Awareness of the nature of

mental states (22); (2). Recognition of developmental aspects of mental states (36); and (3). The effort to understand behavior based on mental states (37). An example that received a high score on reflective reappraisal is the following: "We had a doctor appointment for our son during school hours. I prepped him before school so it is not a surprise. He whined for an hour because he was missing recess." After a time passed, "I was thinking how he felt, I gave him time for himself and I was happy that he loves school, I was thinking how change in routine is hard for him." Non-reflective reappraisal was scored as being present when the criteria for cognitive reappraisal were met, but the statement did not meet the criteria for reflective reappraisal (there are no mentalization characteristics). Two judges got parental vignettes and had to assess on 1 to 7 Likert scale how much they saw evidence for cognitive reappraisal based on the vignette, where 1 = there is no evidence and 7 = there is strongly evidence. The judges were blind to the time point of assessment, i.e., whether the parents' description comes from the pre-or post assessment. The correlation between independent raters' scores was high for both reflective reappraisals (pre-intervention: $r = 0.97$, $p < 0.001$; post intervention: $r = 0.93$, $p < 0.001$) and non-reflective reappraisals (pre-intervention: $r = 0.92$, $p < 0.001$; post intervention: $r = 0.91$, $p < 0.001$). For this reason, the ratings were averaged across raters for each reappraisal type, at each time point (pre and post-intervention).

Results

Change in cognitive reappraisal pre- to post-intervention

To examine whether cognitive reappraisal scores, measured by the Emotion Regulation Questionnaire, increased with the intervention, we employed a paired samples *t*-test analysis. Results indicated that there was indeed such increase [pre-intervention cognitive reappraisal: M = 4.71, SE = 0.21; post-intervention cognitive reappraisal: M = 5.15, SE = 0.19; $t_{(26)} = 2.24$, $p < 0.05$, Cohen's D = 0.43].

Next, we examined the increase in cognitive reappraisal scores as measured by narrative scoring. Cognitive reappraisal (CR) at each time point was computed as the sum of the two reappraisal types (CR = NR+RR). The increase in the computed CR measure was then examined using a paired samples *t*-test analysis. Results again indicated an increase between the two measurements [$t_{(26)} = -4.46$, $p < 0.001$, Cohen's D = 0.86; pre-intervention cognitive reappraisal: M = 4.57, SE = 0.53; post-intervention cognitive reappraisal: M = 7.89, SE = 0.46]. Figure 1A presents the results for the ERQ scoring, and Figure 1B presents the results for the narrative scoring. Interestingly, changes in the two indicators of cognitive reappraisal were not correlated ($r = -0.17$, ns), suggesting the difference between trait and situation-based assessments approaches.

Change in reflective and non-reflective reappraisal pre- to post-intervention

Following the primary analyses presented above, we proceeded to some broader, exploratory analyses, that examined change between pre and post-intervention in the two types of cognitive reappraisal, namely, reflective and non-reflective reappraisal (RR and NR, respectively). To this end, we employed a Repeated Measures ANOVA, with two within-subjects variables: time (pre and post) and reappraisal type (RR and NR). The analysis revealed three significant effects. The time effect was significant [$F_{(1,26)} = 19.88$, $p < 0.001$, $\eta^2_p = 0.43$], so that pre-intervention scores ($M = 2.29$, $SE = 0.27$) were significantly lower than post-intervention scores ($M = 3.94$, $SE = 0.23$). The reappraisal type effect was also significant [$F_{(1,26)} = 16.75$, $p < 0.001$, $\eta^2_p = 0.39$], so that NR scores ($M = 2.26$, $SE = 0.24$) were significantly lower than RR scores ($M = 3.97$, $SE = 0.29$). Finally, the time-reappraisal type interaction was significant, [$F_{(1,26)} = 10.90$, $p < 0.01$, $\eta^2_p = 0.30$], so that in NR scores there wasn't a significant difference between the two time points (pre-intervention: $M = 2.02$, $SE = 0.33$; post-intervention: $M = 2.50$, $SE = 0.41$; n.s.), while in RR scores there was a significant difference between the two time points (pre-intervention: $M = 2.56$, $SE = 0.38$; post-intervention: $M = 5.39$, $SE = 0.34$). Figure 2 presents the time*reappraisal type interaction.

Discussion

Our finding of an improvement in cognitive reappraisal after a mentalization-based intervention is in alignment with previous studies that demonstrate positive association between mentalization and successful emotion regulation (16). In particular, studies have shown that higher levels of mentalization are coupled with transformation from non-adaptive thoughts to balanced and adaptive ones (15, 17, 36) similar to the change we found in cognitive reappraisal. The possible improvement in emotion regulation after a short mentalization intervention is especially significant for parents of children with autism as they report higher levels of stress and prolong negative emotions compared to parents of typically developing children (2, 3, 5).

The increase in cognitive reappraisal is especially notable as it was evident using two different assessment methods. One is based on self-report that was given in a trait format. The second is based on a narrative scoring that was given in a state format. The improvement in cognitive reappraisal in a trait format after a short intervention is especially remarkable and might point to a possible underestimation of the true change. The improvement in state format includes ecological characteristics, based on daily situations. The fact that we did not see a correlation between the two methods may be due to the fact that correlations between trait and state measures are often weak (38, 39).

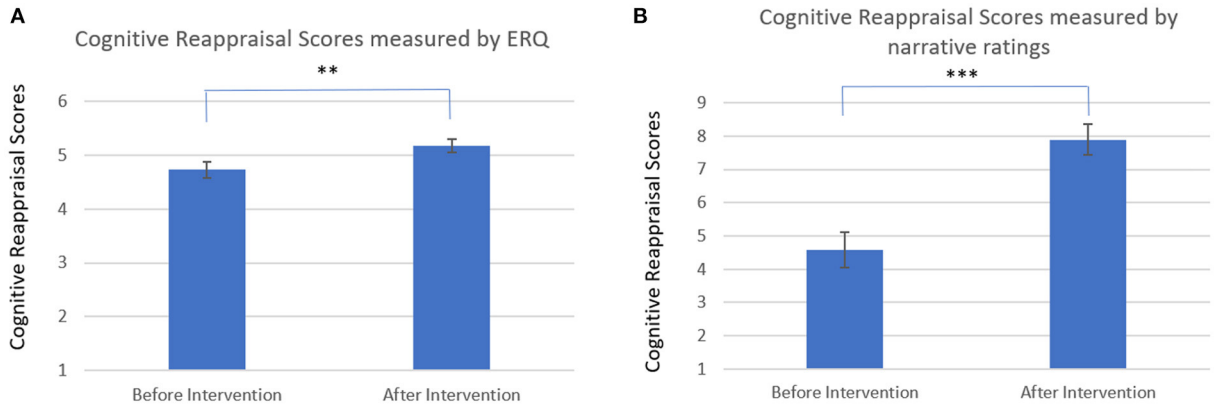


FIGURE 1
(A) Cognitive reappraisal scores measured by ERQ. Results indicate an increase in cognitive reappraisal, as measured by the emotion regulation questionnaire, after intervention. **(B)** Cognitive reappraisal scores measured by narrative ratings. Results indicate an increase in cognitive reappraisal, as measured by narrative ratings (situation-based cognitive reappraisal), after intervention. ***$p < 0.001$, **$p < 0.01$.

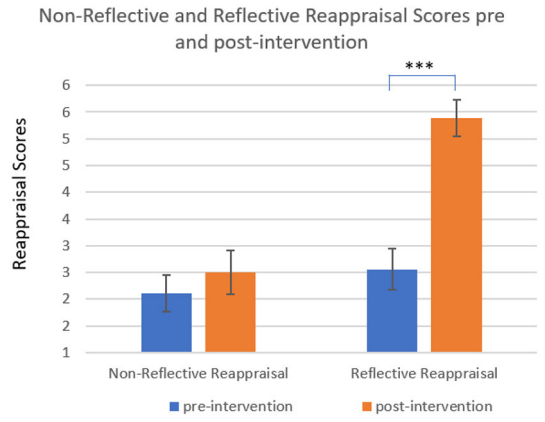


FIGURE 2
Non-reflective and reflective reappraisal scores, pre and post-intervention. Results indicate a significant difference between the two time points in reflective reappraisal scores. The difference in non-reflective reappraisal scores was non-significant. ***$p < 0.001$.

The increase in cognitive reappraisal post intervention was evidenced with reflective reappraisal but not with non-reflective reappraisal. One possible explanation for this specificity in effects could be related to the content of the intervention that is focused on mentalization. As mentalization facilitates reflective thinking, it makes sense that mentalization based intervention is leading to reappraisals with reflective characteristics (i.e., reflective reappraisals). This finding is consistent with previous studies that connect between mentalization and cognitive reappraisal with reflective characteristics. In particular studies present that the capacity to reflect on internal mental states of oneself and others is leading to cognitive appraisals that integrate qualities of mentalization (22, 24).

The improvement in reflective reappraisal following a mentalization-based intervention is important for parents of children with autism. They often have to work harder than parents of typically developing children to understand their children's behavior often in an absence of positive response from the child (21). As there is mounting evidence that high parental mentalization is associated with higher quality caregiving, attachment security, and successful emotion regulation (32, 33, 40) the possibility that short mentalization based intervention supports both parents' mentalization and emotion regulation is encouraging and support using both mentalization and emotion regulation principles in practice with parents of children with ASD.

While the current findings are presenting a potential positive impact of the mentalization-based workshop intervention for parents of children with ASD, this study has several limitations, which should be addressed in the future. First, the investigation does not have a control group, which limits our ability to determine whether change was related to the active intervention. Second, parents of children with ASD were included in the study but the diagnosis of ASD was not confirmed directly by study investigators. Instead, clinicians' reports were used to determine eligibility of parents. Third, the sample size of this study is relatively modest. Fourth, the measurement in this study were based on parents' reports. Finally, the children's ages ranged from 3 to 18, and we were underpowered to detect potential moderation by the children's age and sex.

Author contributions

YE has lead the project and the paper. AH was a mentor and consultant from the Autism perspective. JG was a mentor and

consultant from Emotion Regulation perspective. All authors contributed to the article and approved the submitted version.

Acknowledgments

The authors thank Dana Erhard-Weiss, Mirit Kopleman, Despina Petsagourakis and Jay Webber for their help with this research project.

Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

Publisher's note

All claims expressed in this article are solely those of the authors and do not necessarily represent those of their affiliated organizations, or those of the publisher, the editors and the reviewers. Any product that may be evaluated in this article, or claim that may be made by its manufacturer, is not guaranteed or endorsed by the publisher.

References

1. Davis RF III, Kiang L. Parental stress and religious coping by mothers of children with autism. *Psycholog Relig Spiritual.* (2020) 12:137–48. doi: 10.1037/rel0000183

2. DesChamps TD, Ibañez LV, Edmunds SR, Dick CC, Stone WL. Parenting stress in caregivers of young children with ASD concerns prior to a formal diagnosis. *Autism Res.* (2020) 13:82–92. doi: 10.1002/aur.2213

3. Estes A, Olson E, Sullivan K, Greenson J, Winter J, Dawson G, et al. Parenting-related stress and psychological distress in mothers of toddlers with autism spectrum disorders. *Brain Dev.* (2013) 35:133–8. doi: 10.1016/j.braindev.2012.10.004

4. Olson L, Chen B, Ibarra C, Wang T, Mash L, Linke A, et al. Behaviors are associated with increased parenting stress in caregivers of young children with autism. *J Autism Dev Disord.* (2022) 52:975–86. doi: 10.1007/s10803-021-04995-w

5. Zablotsky B, Bradshaw CP. The association between mental health, stress, and coping supports in mothers of children with autism spectrum disorders. *J Autism Dev Disord*. (2013) 43:1380–93. doi: 10.1007/s10803-012-1693-7

6. Hughes EK. Reciprocal relationships between parent and adolescent internalizing symptoms. *J Fam Psychol.* (2010) 24:115. doi: 10.1037/a0018788

7. Morelen D, Shaffer A. Maternal emotion regulation: links to emotion parenting and child emotion regulation. *J Fam Issues.* (2014) 37:1891–916. doi: 10.1177/0192513X14546720

8. Gross JJ. Emotion regulation: conceptual and empirical foundations.? In: Gross JJ, editors. *Handbook of Emotion Regulation*. The Guilford Press (2014). p. 3–20.

9. Urry HL. Emotion regulation in older age. *Curr Dir Psychol Sci.* (2010) 19:352–7. doi: 10.1177/0963721410388395

10. Buhle JT, Silvers JA, Wager TD, Lopez R, Onyemekwu C, Kober H, et al. Cognitive reappraisal of emotion: a meta-analysis of human neuroimaging studies. *Cerebral Cortex.* (2013) 24:2981–90. doi: 10.1093/cercor/bht154

11. Morawetz C, Bode S, Baudewig J. Effective amygdala-prefrontal connectivity predicts individual differences in successful emotion regulation. *Soc Cogn Affect Neurosci.* (2017) 12:569–85. doi: 10.1093/scan/nsw169

12. Gross JJ. Individual differences in two emotion regulation processes: implications for affect, relationships, and well-being. *J Pers Soc Psychol.* (2003) 85:348. doi: 10.1037/0022-3514.85.2.348

13. Picó-Pérez M, Radua J, Steward T, Menchón JM. Emotion regulation in mood and anxiety disorders: a meta-analysis of fMRI cognitive reappraisal studies. *Prog Neuro Psychopharmacol Biol Psychiatr.* (2017) 79:96–104. doi: 10.1016/j.pnpbp.2017.06.001

14. Troy AS, Wilhelm FH, Shallcross AJ. Seeing the silver lining: cognitive reappraisal ability moderates the relationship between stress and depressive symptoms. *Emotion.* (2010) 10:783. doi: 10.1037/a0020262

15. Fonagy P. Progress in the treatment of borderline personality disorder. *Br J Psychiatr.* (2006) 188:1–3. doi: 10.1192/bjp.bp.105.012088

16. Kivity Y, Levy KN, Kelly KM. In-session reflective functioning in psychotherapies for borderline personality disorder: the emotion regulatory role of reflective functioning. *J Consult Clin Psychol.* (2021) 89:751. doi: 10.1037/ccp0000674

17. Beaulieu-Pelletier G, Bouchard MA, Philippe FL. Mental States Task (MST): development, validation, and correlates of a self-report measure of mentalization. *J Clin Psychol.* (2013) 69:671–95. doi: 10.1002/jclp.21942

18. Allen JG, Fonagy P. *Mentalizing in Clinical Practice*. American Psychiatric Pub (2008).

19. Hooker CI, Verosky SC, Germine LT, Knight RT. Mentalizing about emotion and its relationship to empathy. *Soc Cogn Affect Neurosci.* (2008) 3:204–17. doi: 10.1093/scan/nsn019

20. Swan P. Social connection: empathy and mentalization for teachers. *Pastor Care Educ.* (2015) 33:220–33. doi: 10.1080/02643944.2015.1094120

21. Slade A. Mentalizing the unmentalizable: parenting children on the spectrum. *J Infant Child Adolesc Psychother.* (2009) 8:7–21. doi: 10.1080/15289160802683054

22. Fonagy P, Gergely G, Jurist EL. *Affect Regulation, Mentalization, and the Development of the Self*. Routledge (2018).

23. Jurist EL. Mentalized affectivity. *Psychoanal Psychol.* (2005) 22:426. doi: 10.1037/0736-9735.22.3.426

24. Fonagy P, Luyten P, Allison E. What we have changed our minds about: part 2. Borderline personality disorder, epistemic trust and the developmental significance of social communication. *Borderline Personal Disord Emot Dysregul.* (2017) 4:1–12. doi: 10.1186/s40479-017-0062-8

25. Lecours S. Dimensions of mentalisation: outlining levels of psychic transformation. *Int J Psycho Anal.* (1997) 78:855–75.

26. Katznelson H. Reflective functioning: a review. *Clin Psychol Rev.* (2014) 34:107–17. doi: 10.1016/j.cpr.2013.12.003

27. Levy KN, Meehan KB, Kelly KM, Reynoso JS, Weber M, Clarkin JF, et al. Change in attachment patterns and reflective function in a randomized control trial of transference-focused psychotherapy for borderline personality disorder. *J Consult Clin Psychol.* (2006) 74:1027. doi: 10.1037/0022-006X.74.6.1027

28. Baylin J. Reflective functioning and parenting. In: Alper J, Howe D, editors. *Assessing Adoptive Parents, Foster Carers, and Kinship Carers: Improving Analysis and Understanding of Parenting Capacity*. London and Philadelphia: Jessica Kingsley Publishers (2017). p. 175–92.?

29. Sharp C, Fonagy P. Posttraumatic stress disorder: a social-cognitive perspective. *Clin Psychol Sci Pract.* (2012) 19:229. doi: 10.1111/cpsp.12002

30. Fonagy P, Luyten P, Allison E, Campbell C. What we have changed our minds about: part 1. Borderline personality disorder as a limitation of resilience. *Borderline Personal Disord Emot Dysregul.* (2017) 4:1–11. doi: 10.1186/s40479-017-0061-9

31. Enav Y, Erhard-Weiss D, Kopelman M, Samson AC, Mehta S, Gross JJ, et al. A non-randomized mentalization intervention for parents of children with autism. *Autism Res.* (2019) 12:1077–86. doi: 10.1002/aur.2108

32. Heron-Delaney M, Kenardy JA, Brown EA, Jardine C, Bogossian F, Neuman L, et al. Early maternal reflective functioning and infant emotional regulation in a preterm infant sample at 6 months corrected age. *J Pediatr Psychol.* (2016) 41:906–14. doi: 10.1093/jpepsy/jsv169

33. Smaling HJ, Huijbregts SC, Suurland J, van der Heijden KB, Mesman J, van Goozen SH, et al. Prenatal reflective functioning and accumulated risk as predictors of maternal interactive behavior during free play, the still-face paradigm, and two teaching tasks. *Infancy*. (2016) 21:766–84. doi: 10.1111/infa.12137

34. Preece DA, Becerra R, Robinson K, Gross JJ. The emotion regulation questionnaire: psychometric properties in general community samples. *J. Pers. Assess.* (2020) 102:348–56. doi: 10.1080/00223891.2018.1564319

35. Gross JJ. *Handbook of Emotion Regulation*. Guilford publications (2013).

36. Slade A. Parental reflective functioning: an introduction. *Attach Hum Dev.* (2005) 7:269–81. doi: 10.1080/14616730500245906

37. Allen JG. Mentalizing in practice. In: Allen JG, Fonagy P, editors. *Handbook of Mentalization-Based Treatment*. John Wiley & Sons Inc (2006). p. 3–30.

38. Brockman R, Ciarrochi J, Parker P. Emotion regulation strategies in daily life: mindfulness, cognitive reappraisal and emotion suppression. *Cogn Behav Ther*. (2017) 46:91–113. doi: 10.1080/16506073.2016.1218926

39. Nezlek JB. A multilevel framework for understanding relationships among traits, states, situations and behaviours. *Euro J Pers.* (2007) 21:789–810. doi: 10.1002/per.640

40. Ensink K, Bégin M, Normandin L. Maternal and child reflective functioning in the context of child sexual abuse: pathways to depression and externalising difficulties. *Euro J Psychotraumatol.* (2016) 7:30611. doi: 10.3402/ejpt.v7.30611

frontiers | Frontiers in Psychiatry

TYPE Original Research
PUBLISHED 23 November 2022
DOI 10.3389/fpsyt.2022.940872

OPEN ACCESS

EDITED BY
Chung Hyuk Park,
George Washington University,
United States

REVIEWED BY
Adriana Sampaio,
University of Minho, Portugal
Susana Mouga,
Coimbra Institute for Biomedical Imaging and Translational Research (CIBIT), Portugal

*CORRESPONDENCE
Andrea C. Samson
andrea.samson@unifr.ch

SPECIALTY SECTION
This article was submitted to Autism, a section of the journal Frontiers in Psychiatry

RECEIVED 10 May 2022
ACCEPTED 31 October 2022
PUBLISHED 23 November 2022

CITATION
Samson AC, Sokhn N, Van Herwegen J and Dukes D (2022) An exploratory study on emotion regulation strategy use in individuals with Williams syndrome, autism spectrum disorder and intellectual disability. *Front. Psychiatry* 13:940872. doi: 10.3389/fpsyt.2022.940872

An exploratory study on emotion regulation strategy use in individuals with Williams syndrome, autism spectrum disorder and intellectual disability

Andrea C. Samson [1,2]*, Nayla Sokhn [2,3,4], Jo Van Herwegen [5] and Daniel Dukes [1,6]

[1]Institute of Special Education, University of Fribourg, Fribourg, Switzerland, [2]Faculty of Psychology, UniDistance Suisse, Brig, Switzerland, [3]Department of Psychology, University of Fribourg, Fribourg, Switzerland, [4]Eye and Brain Mapping Laboratory (iBMLab), Department of Psychology, University of Fribourg, Fribourg, Switzerland, [5]Department of Psychology and Human Development, UCL Institute of Education, London, United Kingdom, [6]Swiss Center for Affective Sciences, University of Geneva, Geneva, Switzerland

Background: Individuals with neurodevelopmental disorders often have atypical emotion profiles, but little is known about how they regulate their emotions. While several studies have examined emotion regulation strategy use in autism spectrum disorder (ASD), only a few have included individuals with intellectual disability (ID) or focused on specific syndromes such as Williams syndrome (WS).

Methods: A parent-reported survey launched during the first months of the COVID-19 pandemic allowed to exploratorily study emotion regulation strategy use and its link to anxiety in individuals with ASD with (N=785) and without ID (N=596), WS (N=261), and Intellectual Disability not otherwise specified (N=649).

Results: Using multilevel analyses, besides revealing specific group differences in emotion regulation strategy use, a variety of strategies (e.g., rumination, avoiding information, repetitive behaviors) were found to be linked to elevated levels of anxiety, while focusing on the positive was linked to lower anxiety levels in all groups. Moreover, only autistic people without ID used humor more frequently while experiencing lower anxiety levels.

Conclusion: This study sheds light on an underexplored area of emotion regulation strategy use in different neurodevelopmental disorders. It also paves the way to further examine emotion regulation in more rigorous ways to better understand emotion regulation in different neurodevelopmental disorders as

well as the impact on outcome measures such as anxiety. This exploratory study may help to develop and validate adequate measures to study a broad array of ER strategies used by individuals with neurodevelopmental disorders.

KEYWORDS

emotion regulation, COVID-19, Williams syndrome, autism, anxiety, intellectual disability

Introduction

In a variety of contexts, it has been shown that the ability to regulate one's own emotions is important for mental health and wellbeing (1, 2). Emotion regulation (ER) has been defined as the strategies people use to alter the trajectory of an emotion at different points in the emotion generative process to facilitate progress toward a desired goal (2). People use more or less consciously a variety of ER strategies, at times in combination or in sequence, and ideally in a flexible way, to attenuate or itensify their emotional experience. While the context will determine how adaptive the choice and implementation of an ER strategy will be, the adaptiveness of a strategy can also be seen in terms of how often it is used on a habitual basis and its association with long-term outcomes on mental health (2). In this sense, a few strategies are linked to positive long-term outcomes, such as cognitive reappraisal, humor, or problem solving, while others (including avoidance, rumination, or expressive suppression) have been linked to rather negative long-term consequences. Emotion dysregulation, i.e., the inflexible use of more maladaptive ER strategies, has been linked to a variety of internalizing (e.g., anxiety and depression) and externalizing (e.g., aggression) problems (3).

While individuals with neurodevelopmental disorders frequently experience emotional disturbances such as higher internalizing or externalizing problems (4), the link between these problems and ER strategy use has only explicitly been made in a few studies (5, 6). Several studies revealed a tendency for autistic individuals to use maladaptive instead of adaptive ER strategies (7). However, little is known about ER strategy use and efficacy in other neurodevelopmental disorders, such as Williams syndrome (WS), and there are only a few studies about ER strategy use in ASD with intellectual disability (ID) (8). For individuals with ID, more cognitively challenging strategies such as cognitive reappraisal may be less accessible, and the flexibility that is required in different phases of ER (being aware of potential ER strategies, selecting and implementing them appropriately to the context) may be restricted. Furthermore, one needs to constantly monitor ER efficacy in order to maintain, stop or change a specific strategy (2). Limited access to a variety of self-focused ER strategies may render individuals with neurodevelopmental disorders, particularly with ID, more dependent on other people to support their ER, i.e., extrinsic ER (9).

For the purpose of this study, we are interested in comparing the ER strategy use of autistic people with individuals with WS. WS is a rare genetic disorder with an estimated prevalence rate of 1 in 7,500 children (10). In contrast, ASD is relatively more frequent: a recent systematic review indicates a median prevalence of 100 in 10'000 (11). ASD and WS are both neurodevelopmental disorders associated with distinct but overlapping social phenotypes as well as associated psychopathologies, including anxiety. In terms of what distinguishes the social phenotypes, individuals with WS have a socio-emotional profile that can be characterized as being more than usually inclined to approach strangers, being gregarious and hyper-sociable, and being attracted to others' faces (12–15), while the socio-emotional profile of individuals with ASD may be characterized in certain domains as opposite to that (16, 17) with socially-avoidant behavior, a lack of desire to engage with others, and a reduced attention or interest toward social stimuli (15, 17, 18). Interestingly, and in terms of the similarities between the social phenotypes, individuals with WS and autism also share some characteristics, such as difficulties in social cognition and social information processing (e.g., Theory of Mind), social communication, autistic mannerisms and the need to adhere to routines in daily life (16, 17, 19–22).

It is likely that some of these socio-emotional characteristics and difficulties in social cognition and social information processing impact self-focused and extrinsic ER in individuals with neurodevelopmental disorders. Hyper-sociability, positivity bias [which is prominent in WS (23)], and high social approach may, on the one hand, protect individuals from experiencing negative emotions too intensely for too long, but on the other, impact on the availability and choice of ER strategies. Low social motivation and a limited Theory of Mind may render extrinsic ER less accessible to individuals. In addition, further characteristics may impact ER, such as alexithymia [i.e., difficulty to recognize and express emotions and to distinguish between different emotions, as well as thoughts focused on external rather than internal experience (24)], which is frequently present in autistic people (25). Alexithymia may make it difficult to identify the need to regulate, select and implement appropriate strategies or monitor

ER efficacy. Finally, the need to adhere to routines may increase the need to regulate. To summarize, several factors may play a role in ER strategy use in individuals with different neurodevelopmental disorders.

In terms of ASD and WS specifically, both are characterized by elevated levels of anxiety (26). Many autistic individuals experience higher trait anxiety (27) and as many as 40% are estimated to meet the diagnostic criteria for anxiety disorder (28). Anxiety is also reported to be one of the most common psychopathologies in individuals with WS (29, 30) with higher levels of anxiety reported than in individuals with Down syndrome, Prader Willi-syndrome, and Intellectual Disability, for example (30–33). Some studies also point to higher anxiety levels in autistic individuals compared to individuals with WS (26, 34). Interestingly, the type of anxieties appears to be different: while autistic people may have social and non-social anxieties, individuals with WS less often show social anxieties compared to autistic people (26).

As already mentioned, while there are some studies concerning ER in ASD, more research is necessary to understand the ER profile in WS. Some studies suggest that individuals with WS may have difficulties with ER affecting anxiety and specific phobias (35–37), but little research has examined ER strategy use in WS. Gaining more insight into ER strategy use in individuals with WS could have important implications for interventions (36).

ER was particularly challenging during the COVID-19 pandemic which presumably almost universally generated high stress and elevated levels of negative emotions. Several studies suggested that maladaptive strategy use such as catastrophizing, rumination, or excessive health-related information seeking was correlated with increased perceived distress, negative emotions, depression, and anxiety, while adaptive strategies (including positive refocusing and acceptance) have acted as a buffer to alleviate emotional distress and negative emotions during COVID-19 in the general population (38–44). It has even been suggested that individuals with neurodevelopmental disorders and their families have been disproportionally impacted (45–47). For example, in a study including individuals with attention deficit and hyperactivity disorder, poorer pre-COVID-19 ER abilities were linked to increased mental health symptoms (48). However, to the best of our knowledge, no study to date has focused on ER strategy use and anxiety in individuals with WS compared to autistic people. A survey launched in the early months of the COVID-19 pandemic presented an opportunity to do just that.

The present study

The primary goal of this study was to explore the caregiver-reported anxiety and use of a broad array of ER strategies and how the two may be linked in neurodevelopmental disorders. We aimed to include a broad array of ER strategies in an exploratory manner since to the best of our knowledge there is no current validated questionnaire that includes a comparable broad range of behaviors and strategies used in the attempt to deal with anxiety in neurodevelopmental disorders. We were specifically interested in comparing the ER strategy use of individuals with WS and autistic people. Since ID may impact the access to and use of ER strategies, particularly in relation to more cognitively demanding strategies such as cognitive reappraisal, we split the ASD group into individuals with and without ID. Moreover, since it was of interest to study the impact of ID without ASD, we also included individuals with ID not otherwise specified (ID-NOS). First, we compared the level of anxiety in these groups. Second, we described the use of 14 ER strategies representing a variety of adaptive and maladaptive but also rather cognitive and behavioral strategies in these four groups. Twelve strategies were self-focused, while two described co-regulation by parents or caregivers (extrinsic ER). Third, we examined the link between ER strategy use and anxiety across the four groups. Finally, we examined potential group differences in the link between strategy use and anxiety.

Method

Participants

For the present study, we will focus on a subsample of the large international sample collected (see procedures section) of 2,288 individuals, in total: 261 individuals with WS ($M = 17.78$ years, $SD = 10.22$, 119 (45.6%) female), 785 autistic people with ID ($M = 12.42$ years, SD=7.00, 153 (19.5%) female), 596 autistic people without ID ($M = 11.49$ years, $SD = 5.92$, 124 (20.8%) female), and 649 individuals with ID-NOS ($M = 19.06$ years, $SD = 12.07$, 281 (43.3%) female). The individuals resided in 51 countries (see Supplementary material A). Specific data on race/ethnicity has not been recorded. 65.12% of the respondents were mothers, 27.00% were fathers, and a minority (7.88%) were caregivers or other relatives. The education attainment levels of the respondents were 2.80% with no formal qualification, 13.30% with further vocational training, 20.91% with school-leaving certificate, 35.14% with a university bachelor's degree or equivalent, 21.79 with a university master's degree or equivalent and 6.03% who wrote "Other".

We found a significant difference in age of the four groups; $F(3, 2284) = 105.48$, $p<0.001$. *Post-hoc* tests revealed that the groups of ASD without and with ID did not differ in age ($p = 0.22$), nor did the groups of WS and ID-NOS ($p = 0.20$). However, autistic people with ID were significantly younger than individuals with ID-NOS ($p < 0.001$, CI = [5.44 7.86]) and individuals with WS ($p < 0.001$, CI = [3.73 7.00]); and autistic people without ID were significantly younger than individuals with ID-NOS ($p < 0.001$, CI = [6.27 8.87]) and those with WS

($p < 0.001$, CI = [4.58 7.99]). The χ^2-test on distribution of males and females in the four groups revealed a significant effect; $\chi^2(3) = 151.98$, $p < 0.001$. There were more males than females in each group except in the WS group in which gender was distributed equally ($p = 0.06$). As a consequence, we included age and gender as covariates for the multilevel analyses.

The survey

The parent or caregiver reported survey was developed at the beginning of the COVID-19 pandemic in order to better understand how individuals with special educational needs and disabilities (SEND; neurodevelopmental disorders constitute a subgroup of SEND) were affected by the pandemic [see (49, 50)]. First, demographic information was requested of the respondent (parents or caregivers) and individual with SEND. The respondent also reported the primary diagnosis and the presence (or absence) of ID in the individual with SEND, allowing us, for example, to distinguish between autistic people with and without ID. Then, anxiety was assessed with a single item ["How anxious was/is your child?", similar to Turon et al. (51)] on a scale from 1 (not at all) to 5 (extremely) at three time points: before the pandemic, at the start of the pandemic and at the time of survey completion—in the "now" moment (between April and August 2020). Here, we will focus only on the "now" moment since data relating to ER strategy use was only collected about then. Finally, the survey included a broad range of 14 potential ER strategies and asked with single items how frequently from 1 (very rarely) to 5 (very frequently) the individual with SEND used each strategy to deal with the potentially elevated levels of anxiety (the wording of the instructions and ER items can be found in the Supplementary material B, as well as here (49)[1].

To explore the ER strategy use, we included a range of rather maladaptive and adaptive ER strategies (taking into consideration their habitual use and long-term consequences). Furthermore, we included some cognitive and behavioral strategies, since a broad range of strategies was more likely to capture the relative strengths of each group (2). Twelve of the strategies concerned self-regulation. The following may be considered as rather maladaptive in relation to their long-term consequences: *isolation/withdrawal, information avoidance, information search* [excessively searching for information about the COVID-19 may be an (unsuccessful) attempt to attenuate anxiety levels (41)], *rumination* which refers to continuously thinking about one's own experience, and the causes and consequences of one's negative emotion without calling for action (52), and *expressive suppression*, which refers to inhibiting the outward expression of an emotion (53). Two further possibly maladaptive strategies concerned behavior: *aggressive behaviors* and *repetitive behaviors*. The latter two might not typically be seen as ER strategies but could also be considered as symptoms linked to elevated negative emotions (i.e., anxiety). However, they may also function at times as a response-focused ER strategy. Moreover, repetitive behaviors have been reported to be used as an attempt to deal with negative emotions in several studies (45, 54). In this light, it is important to note that the survey asked which strategies were shown in response to deal with elevated levels of anxiety and stress and not if an individual shows these behaviors in general.

Some strategies may be considered as either adaptive or maladaptive depending on the context such as *sharing/talking about* COVID-19 and *distraction* as one of the attentional deployment strategies (i.e., shifting one's attention to something else in order to avoid or reduce unwanted emotions) (1). Finally, some strategies can be seen as rather adaptive considering their habitual use and long-term consequences: *cognitive reappraisal* (53), *focusing on the positive* (55), and *humor* (56, 57).

Furthermore, we asked about two strategies that we imagined may have been employed by parents or caregivers to regulate the child's emotion: shielding the child from negative information about the pandemic (*parent shielding*) and establishing a routine in their daily life (*parent routine*)[2].

Procedures

The survey, developed at the beginning of the COVID-19 pandemic, was available in 16 languages (49, 50). With the help of more than 60 international collaborators, flyers were sent to associations and cohorts to invite parents and caregivers to report about their child with SEND. There was no age limit for the child with SEND, so respondents were also able to report about their adult child. Ethical approval for this anonymous survey was obtained by the institutional review board of Unidistance Suisse.

Due to the necessity of rapidly developing our COVID-19 specific questionnaire in March 2020, we did not include community members or people with SEND in the development of the questionnaire. However, we had several parent associations help us recruit and we are continuing to work with them with regards to the dissemination of the survey results.

1 The survey included other questions about other domains, such as a variety of specific worries or ER efficacy, which will be reported elsewhere.

2 Although we asked not only about parents or caregivers establishing a routine ("I try or my child tries to establish a routine in his/her daily life to lower the experienced stress"), for the purpose of this analysis, we classified this item as extrinsic ER strategy as establishing a routine is likely to involve the parents' consent, advice or help. The survey is fully available here: Van Herwegen et al. (49).

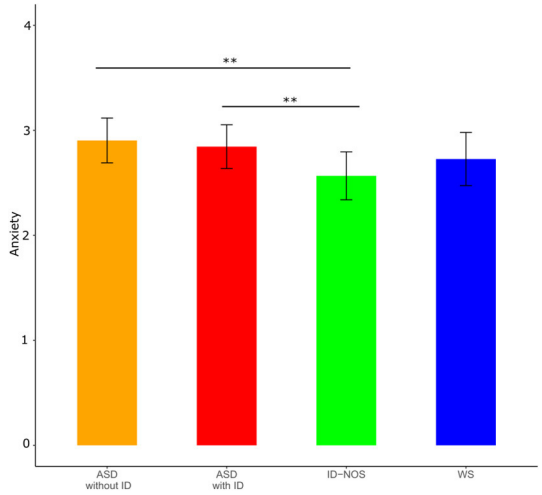


FIGURE 1
Mean anxiety levels with confidence intervals for each group. Significant effects of *post hoc* tests are indicated (black lines). ASD, autism spectrum disorder; ID, intellectual disability; ID-NOS, intellectual disability not otherwise specified; WS, Williams syndrome. Significance levels of *post-hoc* tests: $^{**}p < 0.01$.

Data analysis plan

We included data of families based on the following selection criteria, resulting in the above-mentioned $N = 2{,}288$: primary diagnosis of WS, ASD, or ID-NOS, available information about country, age, gender, available information about presence of ID (which allowed us to distinguish between autistic people with and without ID), age 5 and older (since self-focused cognitive strategies are less likely to occur in younger participants). We excluded cases in which the respondents provided inconsistent information (e.g., primary diagnosis ID-NOS, but a negative response when asked if their child had ID). We did not impute missing values for the ER strategy use; instead, we opted for the complete-cases-analysis approach: a participant is removed if an answer is missing for any of the 12 ER strategies.

We performed linear mixed models to investigate (1) the level of anxiety in the different groups, (2) ER strategy use in the different groups, (3) ER profiles linked to anxiety across the four groups, and (4) group differences in the link between ER strategies and anxiety. The independent variable group for all models except for the third is coded as a categorical variable with four modalities (ASD without ID, ASD with ID, ID-NOS, and WS), the reference level is the ASD without ID. In all four models, age and gender were added as covariates and the country was used as a random factor to account for dependency within each country. Finally, all models are estimated using the restricted maximum likelihood (REML). Preprocessing of the data was conducted using MATLAB software (58) (R2018b, The MathWorks, Natick, MA). Analyses were conducted using R statistical software version 4.0.3 (59), with the packages *lme4* and *lmerTest* for multilevel regression (60, 61), *emmeans* (62) for pairwise comparisons using the tukey method for p-value correction, *effects* (63) and *ggplot 2* (64) *packages* for visualization of main effects of the model.

Results

Level of anxiety in the different groups

The analyses of anxiety levels revealed a main effect of group; $F(3, 2229) = 5.48$, $p < 0.001$ and age; $F(1, 2240) = 33.29$, $p < 0.001$, indicating increased anxiety with increasing age, but no effect for gender; $F(1, 2226) = 0.02$, $p > 0.05$. *Post-hoc* tests revealed significant differences between individuals with ID-NOS and autistic people without ID ($t = 3.68$, $p < 0.01$) and autistic people with ID ($t = 3.46$, $p < 0.01$), respectively, showing that autistic people with and without ID had higher levels of anxiety than individuals with ID-NOS (see Figure 1).

Emotion regulation strategy use in the different groups

On a descriptive level, when sorting the employed ER strategies for each group according to their frequency (see Supplementary material C), it can be observed that *parent routine* and *parent shielding* were among the most frequently parent-reported strategies in all groups. Distraction, repetitive behaviors, and isolation/withdrawal were also frequently employed.

The analyses of parent-reported use of ER strategies revealed significant group effects in 12 out of 14 ER strategies (see Figure 2). No significant group differences were found for *parent shielding* and *cognitive reappraisal*. Generally speaking, no gender effects were found but there was almost always an age effect indicating that the use of a specific ER strategy increased with increasing age except for *repetitive behaviors, parent shielding,* and *parent routine*, where no age effects were found. This suggests that extrinsic ER and repetitive behaviors do not seem to change in our sample as much as other strategies examined here. For the statistics of group, age, and gender effects (see Table 1). In the following, we focus on *post-hoc* tests within the 12 ER strategies in which significant group effects were found.

Isolation/withdrawal

We found significant differences between all the groups except between ID-NOS and WS (see Supplementary material D for *t*-values) with the highest scores for autistic people without ID, followed by autistic people with ID, and the lowest levels for individuals with ID-NOS and WS.

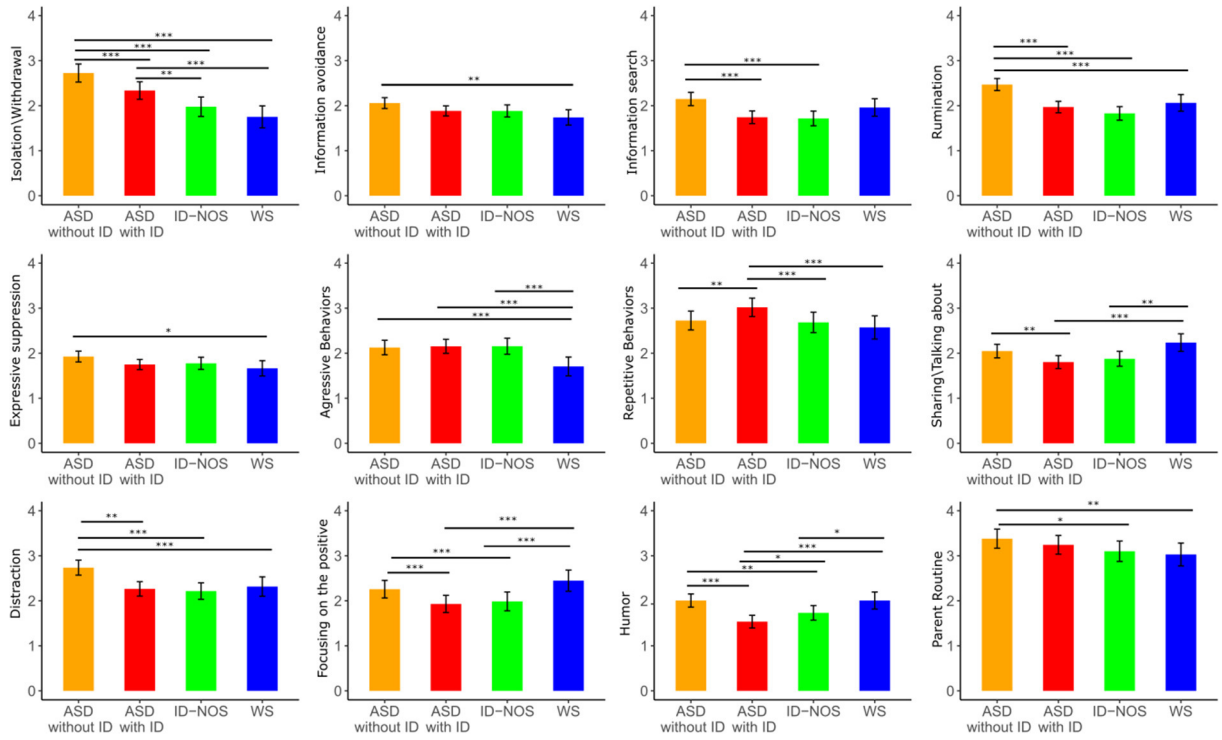


FIGURE 2
Group differences were found in the use of 12 of the 14 emotion regulation (ER) strategies (frequency: from 1 = very rarely to 5 = very frequently). Mean levels with confidence intervals are shown per group for each ER strategy. Significant effects of *post hoc* tests are indicated (black lines). ASD, autism spectrum disorder; ID, intellectual disability; ID-NOS, intellectual disability not otherwise specified; WS, Williams syndrome; Significance levels of *post-hoc* tests: * p <0.05, ** p <0.01, *** p <0.001.

TABLE 1 Effects of group, age, and gender within the multilevel analyses on ER strategy use.

ER strategies	Effect of group	Effect of age	Effect of gender
Isolation/withdrawal	$F(3, 2251) = 34.53^{***}$	$F(1, 2280) = 27.15^{***}$	$F(1, 2267) = 0.00$
Information avoidance	$F(3, 1584) = 4.11^{***}$	$F(1, 1997) = 14.86^{***}$	$F(1, 1983) = 0.27$
Information search	$F(3, 2062) = 12.44^{***}$	$F(1, 2216) = 35.53^{***}$	$F(1, 2277) = 1.00$
Rumination	$F(3, 1681) = 22.87^{***}$	$F(1, 2038) = 35.05^{***}$	$F(1, 2281) = 2.55$
Expressive suppression	$F(3, 1536) = 3.28^{*}$	$F(1, 1966) = 16.93^{***}$	$F(1, 2282) = 0.00$
Aggressive behaviors	$F(3, 2147) = 7.04^{***}$	$F(1, 2249) = 7.95^{***}$	$F(1, 2276) = 0.17$
Repetitive behaviors	$F(3, 2245) = 8.43^{***}$	$F(1, 2279) = 0.40$	$F(1, 2272) = 0.09$
Sharing/talking about COVID-19	$F(3, 2136) = 8.33^{***}$	$F(1, 2244) = 15.16^{***}$	$F(1, 2276) = 0.18$
Distraction	$F(3, 2127) = 15.07^{***}$	$F(1, 2244) = 11.29^{***}$	$F(1, 2275) = 0.40$
Cognitive reappraisal	$F(3, 2026) = 0.97$	$F(1, 2206) = 10.84^{**}$	$F(1, 2275) = 0.00$
Focusing on the positive	$F(3, 2248) = 12.38^{***}$	$F(1, 2282) = 8.78^{**}$	$F(1, 2258) = 1.35$
Humor	$F(3, 2123) = 18.27^{***}$	$F(1, 2242) = 6.56^{*}$	$F(1, 2275) = 1.80$
Parent shielding	$F(3, 2278) = 0.72$	$F(1, 2277) = 1.45$	$F(1, 2264) = 1.41$
Parent routine	$F(3, 2268) = 4.70^{**}$	$F(1, 2282) = 0.20$	$F(1, 2270) = 0.09$

ER = emotion regulation.
*p < 0.05, **p < 0.01, ***p < 0.001.

Information avoidance

Autistic people without ID more frequently avoided information compared to individuals with WS ($t = 3.22$, $p < 0.01$).

Information search

Autistic people without ID search for more information compared to autistic people with ID ($t = 5.54$, $p < 0.001$) and individuals with ID-NOS ($t = 5.02$, $p < 0.001$).

Rumination

Autistic people without ID ruminate more frequently than all other groups (compared to autistic people with ID: $t = -6.77$, $p < 0.001$, to individuals with ID-NOS: $t = -7.42$, $p < 0.001$, and to individuals with WS; $t = -3.92$, $p < 0.001$).

Expressive suppression

Autistic people without ID used this ER strategy more frequently than individuals with WS ($t = 2.67$, $p < 0.05$).

Aggressive behaviors

Individuals with WS used *aggressive behaviors* less frequently than autistic people with ID ($t = 4.31$, $p < 0.001$), autistic people without ID ($t = 3.93$, $p < 0.001$) and individuals with ID-NOS ($t = 4.07$, $p < 0.001$).

Repetitive behaviors

Autistic people with ID used *repetitive behaviors* significantly more frequently than all other groups (compared to autistic people without ID: $t = -3.43$, $p < 0.01$, to individuals with ID-NOS: $t = 3.80$, $p < 0.001$, and to individuals with WS: $t = 3.82$, $p < 0.001$).

Sharing/talking about COVID-19

Individuals with WS used this ER strategy significantly more frequently than autistic people with ID ($t = -4.43$, $p < 0.001$) and individuals with ID-NOS ($t = -3.46$, $p < 0.01$). Moreover, autistic people without ID used this ER strategy more frequently than autistic people with ID ($t = 3.41$, $p < 0.01$).

Distraction

Autistic people without ID used *distraction* more frequently than the other three groups (compared to autistic people with ID: $t = 6.02$, $p < 0.001$, to individuals with ID-NOS: $t = 5.65$, $p < 0.001$, and to individuals with WS: $t = 3.82$, $p < 0.01$).

Focusing on the positive

Turning to the rather adaptive ER strategies, a significant group effect was found for *focusing on the positive*. While autistic people without ID and WS did not differ, both groups used this strategy more frequently than the other two groups: Autistic people without ID used *focusing on the positive* more frequently than autistic people with ID ($t = 4.42$, $p < 0.001$) and individuals with ID-NOS ($t = 3.11$, $p < 0.001$). In addition, individuals with WS used this strategy more frequently than autistic people with ID ($t = -5.09$, $p < 0.001$) and individuals with ID-NOS ($t = -4.27$, $p < 0.001$).

Humor

Autistic people without ID and WS did not differ, but both used *humor* as ER strategy more frequently compared to the other groups. Autistic individuals with ID seemed to be using this strategy less often than all other groups (autistic people without ID used humor significantly more frequently than autistic people with ID: $t = 6.78$, $p < 0.001$, and individuals with ID-NOS: $t = 3.36$, $p < 0.01$. Autistic people with ID used this ER strategy less frequently than individuals with ID-NOS: $t = -2.74$, $p < 0.05$, and then individuals with WS: $t = -4.96$, $p < 0.001$. Individuals with ID-NOS used this ER strategy less frequently than individuals with WS: $t = -2.72$, $p < 0.05$).

Parent routine

Finally, a significant group effect was found for *parent routine*. Parents and caregivers of autistic people without ID used this strategy more frequently than the parents of individuals with ID-NOS ($t = 3.00$, $p < 0.05$) and individuals with WS ($t = 3.16$, $p < 0.01$).

Emotion regulation strategy use linked to anxiety across the four groups

The analyses of ER strategies across the four groups revealed significant effects for seven strategies that were linked to higher anxiety: *information avoidance*; $F(1, 2217) = 4.40$, $p < 0.05$, *information search;* $F(1,2224) = 5.50$, $p < 0.05$, *distraction*; $F(1, 2225) = 20.24$, $p < 0.001$, *rumination*; $F(1, 2223) = 47.60$, $p < 0.001$, *aggressive behaviors*; $F(1, 2221) = 12.31$, $p < 0.001$, *repetitive behaviors*; $F(1, 2224) = 17.45$, $p < 0.001$, and *parent shielding*; $F(1, 2150) = 22.97$, $p < 0.001$. Only one strategy, i.e., *focusing on the positive* was linked to lower anxiety; $F(1, 2227) = 12.69$, $p < 0.001$ (see Figure 3). A significant effect was found for age; $F(1, 2227) = 14.77$, $p < 0.001$, indicating a stronger association between strategy use and anxiety with increasing age, but no gender effect was found; $F(1, 2221) = 0.01$, $p > 0.05$.

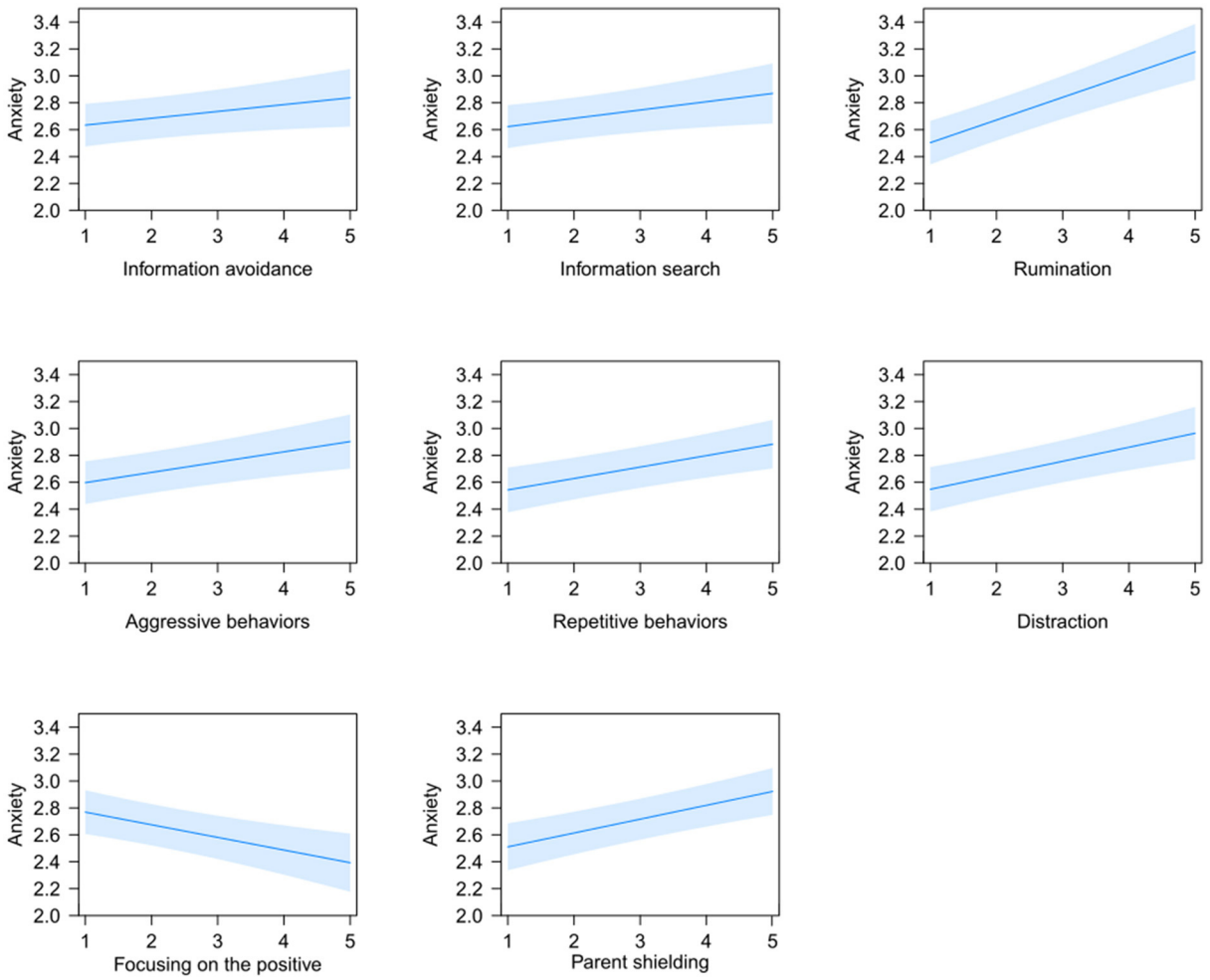


FIGURE 3
Illustration of the significant association between the frequency (from 1 = very rarely to 5 = very frequently) of emotion regulation strategy use and anxiety levels across the four groups. Blue bands denote the 95% confidence interval.

Group differences in the link between emotion regulation and anxiety

We found significant effects of age and of the same seven ER strategies as presented in section Emotion regulation strategy use linked to anxiety across the four groups but not for gender or group (see Supplementary material E for the statistics). However, most interestingly, we found a significant interaction between humor and group; $F(3, 2163) = 3.67$, $p < 0.05$. *Post-hoc* comparisons revealed a significant difference between autistic people without ID and with ID; $t = -3.27$, $p < 0.01$. This may suggest that only autistic people without ID used humor more frequently while experiencing lower levels of anxiety, since *humor* was linked to lower anxiety in this group. While we did not find an association between humor and anxiety in individuals with WS and ID-NOS, *humor* was more frequently used while experiencing increased levels of anxiety in autistic people with ID (see Figure 4).

Discussion

Our survey, launched in the first months of the pandemic, allowed us to study ER strategy use and the link to anxiety in neurodevelopmental disorders, namely, in individuals with WS, autistic people with and without ID, and individuals with ID-NOS. Individuals with neurodevelopmental disorders have been disproportionally affected by the COVID-19 pandemic with increased levels of anxiety and other mental health outcomes (46, 47, 65, 66). While our current analysis does not allow a comparison with typically developing individuals, we observed that autistic people (with and without ID) had elevated anxiety

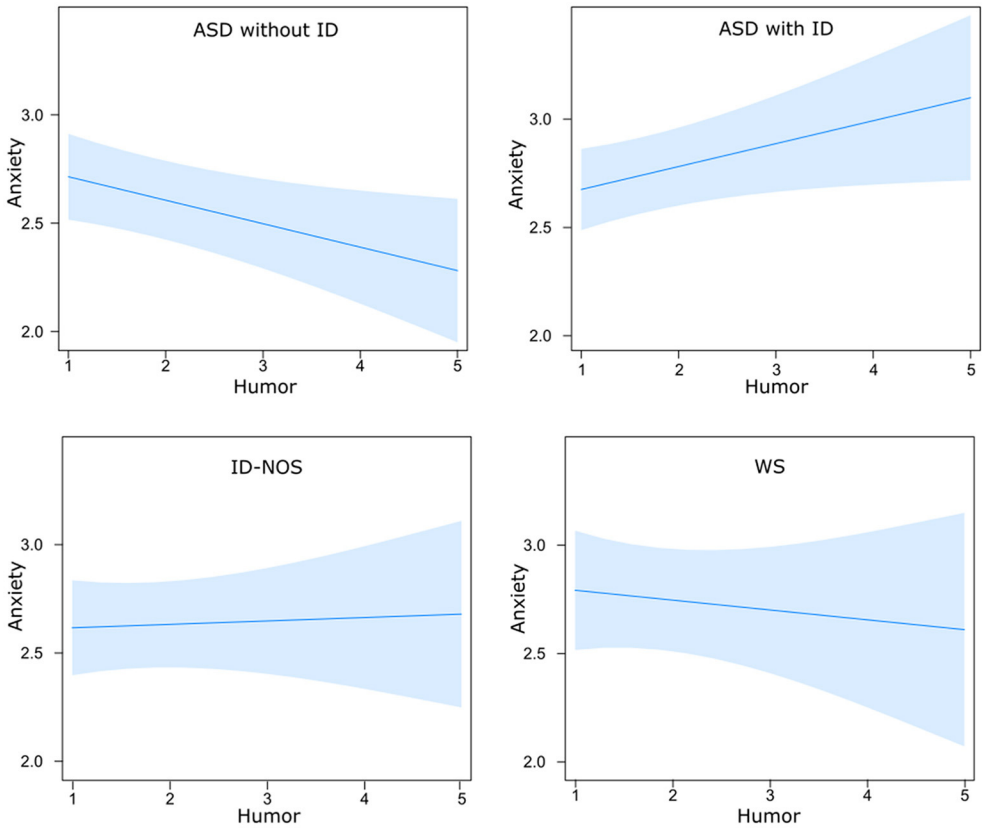


FIGURE 4
Differential effects of humor on anxiety in relation to group membership: Using humor as an emotion regulation strategy was linked to lower anxiety in autistic individuals without ID and to higher anxiety in autistic individuals with ID. ASD, autism spectrum disorder; ID, intellectual disability; ID-NOS, intellectual disability not otherwise specified; WS, Williams syndrome. Blue bands denote the 95% confidence interval.

levels compared to individuals with ID-NOS, in line with previous studies reporting prevalence rates of 42–79% in autistic people (67) while prevalence rates seem lower in individuals with ID [3–22%, (68)]. While individuals with WS experienced increased anxiety in previous studies when compared to people with ID (30), their anxiety levels in the present study were reported to be between the other three groups. Our study mainly aimed to increase our knowledge about parent-reported ER strategy use and how ER strategy use was linked to anxiety.

It seems striking that *parent routine* and *parent shielding* as extrinsic ER were among the top three most frequently employed strategies in all groups, which shows that co-regulation of negative emotions seems to play an important role in all groups. Further studies may help to better understand if other, more cognitive extrinsic ER strategies, such as *other-employed cognitive reappraisal*, are of greater relevance to individuals with neurodevelopmental disorders than to individuals without. It is also interesting that *repetitive behaviors* are among the four most frequently used strategies in all groups, except for autistic people without ID.

Emotion regulation profiles in the different groups

Age effects were found for most of the ER strategies indicating increased use of ER strategies as individuals get older. Although we did not find specific age effects for the different groups in relation to ER strategy use, we would like to mention that the use of adaptive or maladaptive ER strategies may be considered as potential protective or risk factors for socio-emotional outcome measures in individuals with neurodevelopmental disorders across the life span. For example, while children and adolescents with WS are often described as overfriendly and of portraying high approach behavior, adults with WS are often described as socially isolated [see, for example Davies et al. (69)]. It may be important to further study the use of ER strategies in different age groups in relation to various outcome measures including anxiety or social isolation in individuals with neurodevelopmental disorders.

Importantly, we identified differences in the ER profiles of the groups. As far as we are aware, this is the first

time that the strategy use of individuals with WS has been studied. At a descriptive level, individuals with WS used most frequently *parent routine, parent shielding, repetitive behaviors* and *distraction*, suggesting that extrinsic ER, as well as behavioral ER strategies, are of great relevance. We would like to highlight some characteristics in ER potentially linked to the socio-emotional profile in this rare genetic disorder. In comparison with the other groups with ID, individuals with WS used *focusing on the positive* significantly more frequently and *aggressive behaviors* significantly less. This might be linked to their socio-emotional profile such as the positivity bias in individuals with WS who are often described as gregarious and cheerful (70). In addition, individuals with WS seemed to use humor as frequently as autistic people without ID—which may be again linked to their positivity bias. However, the use of humor was not linked to lower levels of anxiety in individuals with WS (in contrast to the effects of using humor on anxiety in autistic individuals without ID) which may be possibly linked to the particular cognitive challenges related to humor appreciation and production (71). Participants with WS also reportedly used the strategy *sharing/talking about COVID-19* more frequently than the other groups with ID, and the lowest levels of *isolation/withdrawal*, as well as less *information avoidance* and *expressive suppression* than autistic people without ID, which may be linked to the pro-social nature widely reported in WS, characterized by high social interest, social approach behavior and high sociability (70).

Autistic people without ID seemed to have a different ER pattern. Compared to the other groups, autistic people without ID had the highest scores in *isolation/withdrawal* and seemed to *avoid information* more than other groups which seems to be in line with reports about their socio-emotional profile (15, 16). They also seemed to use several cognitive strategies more frequently than the other groups which are all characterized by cognitive impairment: *Search for more information about COVID-19*, which may be linked to the detail-oriented cognitive style in autistic individuals (72), and *rumination*. Interestingly, *focusing on the positive* and *humor* were as frequently employed as by individuals with WS. We may conclude that autistic people without ID are more likely to use and potentially benefit from cognitive strategies (which are potentially too challenging for individuals with ID) because of their typical or better than typical cognitive functioning levels. In addition, establishing a routine by parents and caregivers seems to be highly relevant for autistic people without ID (but also for autistic people with ID, as there were no significant differences between these two groups). This fits well to the known elevated need for routines of autistic people (73).

Our study suggests that autistic people with ID and individuals with ID-NOS have a more limited ER repertoire than the other two groups, by not frequently engaging in cognitive strategies. Despite the limited research on ER in ID [see McClure et al., (74)], this aligns with previous observations of a limited repertoire of coping and ER strategies in individuals with ID (75, 76). In the present study, *Repetitive behaviors* were used most frequently by autistic people with ID compared to the other groups. In the absence of a broader range of (cognitive) strategies available to deal with elevated levels of anxiety, it may be possible that extrinsic ER is more important for autistic people with ID and individuals with ID-NOS. While our study only asked for two extrinsic ER strategies, future studies should include a broader range (e.g., extrinsic cognitive reappraisal, distraction) to better understand which strategies employed by parents or caregivers are used most frequently and are most efficacious. Future research is needed, not only to more rigorously examine self-reported and observed ER strategy use linked to ID and cognitive flexibility, but also to examine, at a more fine-grained scale, in which stages of the ER cycle [identification, strategy selection, strategy implementation, and monitoring (2)] ER might be most affected in individuals with ID. To our knowledge, no research has attempted to conceptualize and tease apart these different stages within the ER cycle with the link to ID (2). While our study focused on the parent-reported implementation of ER strategies, the other stages linked to self- and other-focused ER need to be better understood.

Besides cognitive functioning levels, positivity bias, social approach behaviors/sociability, and the need to adhere to routines, as discussed above, alexithymia and theory of mind are potential factors that may impact ER (see Introduction). With the current study design, we are not able to determine the extent to which these factors impact ER in our target groups. However, future studies assessing these abilities in neurodevelopmental disorders may further elucidate their differential impact on ER.

Emotion regulation strategy use and its link to anxiety

Several of the ER strategies included in this study were linked to increased anxiety. Many of these (*information avoidance, information search, rumination, aggressive behaviors*) could be considered as rather maladaptive when a strategy is used frequently on a habitual basis, while *distraction* might be adaptive or maladaptive depending on the context (77). *Repetitive behaviors* may actually have an important soothing effect for individuals with neurodevelopmental disorders, particularly if other (cognitive) strategies are less available or accessible (45, 54), even though it may have negative long-term consequences. In general, the literature has shown that if certain strategies are used too frequently, there may have negative long-term consequences on mental health (3). Interestingly, we cannot determine whether these strategies had necessarily a negative effect on emotions, despite their link to increased anxiety levels. It may also be possible that higher anxiety *triggered* the use of a broad range of strategies, including strategies that are considered as rather maladaptive. Also, we need to be clear that we did not ask what the people do on

a habitual basis (we rather asked, "which strategies does your child use these days to cope with potentially elevated levels of anxiety") and we did not employ a longitudinal research design which would allow us to draw conclusions on mid- or long-term consequences. Therefore, the increased use of these strategies may not only reflect unsuccessful attempts to attenuate increased anxiety levels, but they may also be elicited by higher anxiety, suggesting that the increased use of these strategies may also reflect an increased *need* to regulate. Also, our study assessed ER strategies at one time point only. Future studies targeting individuals with different neurodevelopmental disorders should attempt to monitor changes in ER strategy use related to varying levels of anxiety and stress to be able to draw conclusions about potential changes and adaptations of ER strategy selection and implementation. Such studies could be done using ecological momentary assessments (78) if they have been adapted for individuals with neurodevelopmental disorders with and without ID.

Interestingly, only one strategy was more frequently employed when experiencing lower levels of anxiety: *Focusing on the positive*. Even if this might suggest that this could be a powerful strategy to alleviate anxiety in individuals with neurodevelopmental disorders, in line with findings in individuals with typical development (55), we would like to reiterate that we cannot draw conclusions about a causal link in the present study. Future studies should try to shed more light on the question of whether only people with low anxiety levels are actually able to *focus on the positive,* in the face of difficult life circumstances, or if this strategy actually reduces anxiety in neurodevelopmental disorders. This could prove very important. Nevertheless, several intervention-based studies were published in recent years that successfully implemented positive elements such as play and humor to address fear and anxieties in children with WS (79) or that were able to increase the use of positive emotion regulation strategies including *Focusing on the Positive* in individuals with ASD (80).

Humor as an emotion regulation strategy

While previous studies have shown that humor can be a powerful strategy to regulate emotions [see Samson and Gross (57) for an overview], we found that the use of *humor* as an ER strategy was only linked to lower anxiety in autistic people without ID, suggesting that they may potentially benefit from this strategy, while *humor* was linked to increased anxiety in autistic people with ID. However, again, we need to be cautious in drawing conclusions about a causal link. As shown in a series of studies, autistic individuals have no difficulty to appreciate simpler forms of humor but have more difficulties in understanding and appreciating more complex jokes that require, for example, a Theory of mind to be understood (jokes based on false beliefs) (81). Moreover, lower cheerfulness as a trait and higher seriousness seemed to dampen the susceptibility to humor and humor appreciation in autistic individuals without ID [Samson (82) for a review]. The present study suggests there might be important differences between autistic people with and without ID in terms of their use of humor. It may be possible that producing humor to regulate emotions might have been too overwhelming for autistic people with ID, while it could serve as a resource for autistic people without ID (71). However, this study also opens up room for future research such as examining the difference between producing (which may be more challenging) and consuming humor (for example, watching funny movies) to regulate emotions. Further research could also examine the extent to which autistic people with ID may benefit from humor as extrinsic ER.

Limitations and future implications

Our exploratory study with a large sample of individuals with different neurodevelopmental disorders included autistic people with ID who are often underrepresented in studies on ER in ASD and, for the first time in such a study of ER strategies, individuals with WS. Future research should ideally attempt to replicate our findings with validated measures (for example, by first developing adequate measures that allow the assessment of a variety of ER strategies in neurodevelopmental disorders) in better characterized samples, and focus on specific ER strategies in more detail in experimental settings. These steps would help formulate concrete implications for application in daily life or interventional settings.

There are some limitations to this study that need to be mentioned. With a view to including a broad range of ER strategies but keeping an already long survey manageable for the families, we assessed ER strategy use with single items rather than including existing ER questionnaires. This helped to gain a better understanding of ER strategy use in neurodevelopmental disorders since, to the best of our knowledge, no existing questionnaire includes such a wide range of strategies. Future studies should therefore develop this battery further for the study of ER in individuals with neurodevelopmental disorders. Moreover, as our motivation was to access as many families with a child with SEND as possible and within a short time frame to cover the early effects of the COVID-19 pandemic, we were neither able to include individuals with SEND in the research process, nor to verify the primary diagnosis, relying instead on parent and caregiver reports. Given that not everyone may have had access to clinics or professionals using gold standard measures for diagnosis [e.g., the Autism Diagnostic Observation Schedule, second edition; ADOS-2; Lord et al. (83) for ASD], this remains a weakness of the study. That said, we hope that given the large group size, the findings are nevertheless representative of the different syndromes. In the same vein, we also relied entirely on parent-reported anxiety and ER strategy use. While short anxiety scales exist with 10 items (84), eight items (68), and four items (85), to name a few examples, assessment of

anxiety with a single item can also be found in the literature (51, 86). Such measures have often been developed in the context of particularly difficult circumstances, such as assessing anxiety in critically ill patients. Given that parents and caregivers of children with SEND experienced a difficult time in the early months of the COVID-19 pandemic [for example Toseeb et al. (47)], it was our goal to keep the survey as short as reasonably possible. We thus opted for a single item to assess anxiety, similar in wording to Turon et al. (51).

Regarding the parent-reported ER strategies, it may be the case that certain ER strategies are less visible to a third party (e.g., a parent or caregiver) leading to under-reporting of that strategy (e.g., *expressive suppression*, perhaps), and may even have led to an over-reporting of the extrinsic ER strategies. This may bias parent and caregiver's responses particularly in individuals that may be less able to communicate about more cognitive, less behavioral strategies which may be more likely in individuals with ID. Nonetheless, parent reports lead to insight about ER in individuals who may not have been able to report in such detail about their own situation, emotional experience, and ER during the first months of the COVID-19 pandemic. As it stands, parent-reported ER questionnaires are not uncommon in the literature (87).

One particular characteristic of this study is that the results are taken from a subsample of responses from participants collected during the first few months of the COVID pandemic. But since we have not collected data before the pandemic, we do not claim that these strategies are either pandemic specific, or that they can be generalized to non-pandemic times. While we assume that similar patterns may be found in other times of individual, family or societal crises, future studies should examine changes in ER strategy use in relation to varying levels of anxiety, for example by using ecological momentary assessment approaches (see above). Since it is likely challenging for parents to know whether a particular ER strategy is used as an attempt to regulate COVID-specific anxieties and stress or other factors not related to the pandemic encountered in daily life, we explicitly asked parents and caregivers to report about ER strategy use in general "these days", rather than anything COVID-specific. This exploratory study suggests some interesting similarities and differences across groups that future researchers can use to build hypotheses for more structured, less exploratory studies. For example, it would be interesting to extend knowledge about ER strategy choice. Previous research has shown that under highly negative situations, cognitive reappraisal is less effective than distraction (88). It would be interesting to discover if individuals with neurodevelopmental disorders are able to adapt the strategy choice in relation to the negativity of experienced events.

Also, while we focused here on ER strategy *use*, we must emphasize that ER involves other processes that were not considered here, but which would be relevant for further study in neurodevelopmental disorders with and without ID (see above). These processes include the ability to recognize emotions, set regulatory goals, identify and select potential strategies, the ability to implement one or more strategies in combination or in sequence, and other dynamic processes related to ER such as maintaining or disrupting the use of a particular ER strategy in relation to the context and its efficacy (89). In short, the ER field is large, and there is a huge potential for ER studies in SEND, and, more specifically, in neurodevelopmental disorders. Furthermore, although we included a variety of countries ("country" was included as random factor in the analysis), the current study did not focus on the study of cultural or regional differences in the either the caregiver reports or the use of ER strategies. However, other analyses originating from this international collaborative study from which the present data comes will focus on country-specific differences. We hope that the relative strengths of this exploratory study stimulate and further motivate research in this domain, in spite of its relative limitations.

Conclusions

The present study elucidated different patterns of ER strategy use in different neurodevelopmental disorders. Autistic people without ID were reported to have the largest ER repertoire, including more self-focused cognitive strategies, and were the only group in which we found humor as ER strategy to be linked with lower anxiety levels. For the first time, this study was able to shed light on ER strategy use in individuals with WS, which differed from those of autistic people, which can possibly be accounted for by the well-researched differences in socio-emotional profiles. Moreover, several rather maladaptive strategies were linked to increased anxiety, while *focusing on the positive* was linked to lower anxiety levels across all groups. As such, this exploratory study provides increased insight into ER in WS, ASD with and without ID, and ID-NOS, and highlights the need for further studies on ER strategy use in individuals with neurodevelopmental disorders, using more rigorous assessment tools and that monitor changes of ER strategy use over time. Future studies are required as knowledge about the impact of different ER strategies as potential protective and risk factors on outcomes such as anxiety may also inform interventions to support individuals with neurodevelopmental disorders in their daily lives and in future times of crisis.

Data availability statement

The raw data supporting the conclusions of this article will be made available by the authors, without undue reservation.

Ethics statement

The studies involving human participants were reviewed and approved by Ethics Committee of Unidistance Suisse. The

patients/participants provided their written informed consent to participate in this study.

Author contributions

AS, DD, and JV designed and directed the project. NS and AS analyzed the data. AS, NS, DD, and JV wrote the paper. All authors discussed the results, commented on the manuscript, and provided approval for publication of the content.

Funding

Research Funds of Unidistance Suisse, the European Federation of Williams Syndrome (FEWS) and Swiss National Science Foundation (PP00P1_176722 for AS).

Acknowledgments

The authors would like to thank the families who participated in this research study. We would like to thank all colleagues who helped recruit participants [see Dukes et al. (50)], as well as the WS-iREACH group.

Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

Publisher's note

Supplementary material

The Supplementary Material for this article can be found online at: https://www.frontiersin.org/articles/10.3389/fpsyt.2022.940872/full#supplementary-material

References

1. Gross JJ. The emerging field of emotion regulation: an integrative review. *Rev Gen Psychol.* (1998) 2:271–99. doi: 10.1037/1089-2680.2.3.271

2. Gross JJ. The extended process model of emotion regulation: elaborations, applications, future directions. *Psychol Inq.* (2015) 26:130–7. doi: 10.1080/1047840X.2015.989751

3. Gross JJ, Jazaieri H. Emotion, emotion regulation, and psychopathology: An affective science perspective. *Clin Psychol Sci.* (2014) 2:387–401. doi: 10.1177/2167702614536164

4. Tajik-Parvinchi D, Farmus L, Tablon Modica P, Cribbie RA, Weiss JA. The role of cognitive control and emotion regulation in predicting mental health problems in children with neurodevelopmental disorders. *Child Care Health Dev.* (2021) 47:608–17. doi: 10.1111/cch.12868

5. England-Mason G. Emotion regulation as a transdiagnostic feature in children with neurodevelopmental disorders. *Curr Dev Disord Rep.* (2020) 7:130–8. doi: 10.1007/s40474-020-00200-2

6. Mazefsky CA. Emotion regulation and emotional distress in autism spectrum disorder: Foundations and considerations for future research. *J Autism Dev Disord.* (2015) 45:3405–8. doi: 10.1007/s10803-015-2602-7

7. Cai RY., Richdale AL, Uljarevi,ć M, Dissanayake C, Samson AC. Emotion regulation in autism spectrum disorder: where we are and where we need to go. *Autism Res.* (2018) 11:962–78. doi: 10.1002/aur.1968

8. Sáez-Suanes GP, García-Villamisar D, del Pozo Armentia A, Dattilo J. Emotion regulation as a mediator between depressive symptoms and autism spectrum disorder (ASD) in adults with ASD and intellectual disabilities. *Res Autism Spectr Disord.* (2020) 78:101654. doi: 10.1016/j.rasd.2020.101654

9. Nozaki Y, Mikolajczak M. Extrinsic emotion regulation. *Emotion.* (2020) 20:10–5. doi: 10.1037/emo0000636

10. Strømme P, Bjørnstad PG, Ramstad K. Prevalence estimation of Williams syndrome. *J Child Neurol.* (2002) 17:269–71. doi: 10.1177/088307380201700406

11. Zeidan J, Fombonne E, Scorah J, Ibrahim A, Durkin MS, Saxena S, et al. Global prevalence of autism: a systematic review update. *Autism Res.* (2022) 15:778–90. doi: 10.1002/aur.2696

12. Jones W, Bellugi U, Lai Z, Chiles M, Reilly J, Lincoln A, et al. Hypersociability in Williams syndrome. *J Cogn Neurosci.* (2000) 12:30–46. doi: 10.1162/089892900561968

13. Riby DM, Hancock PJ. Viewing it differently: Social scene perception in Williams syndrome and autism. *Neuropsychologia.* (2008) 46:2855–60. doi: 10.1016/j.neuropsychologia.2008.05.003

14. Riby D, Hancock PJ. Looking at movies and cartoons: Eye-tracking evidence from Williams syndrome and autism. *J Intellect Disabil Res.* (2009) 53:169–81. doi: 10.1111/j.1365-2788.2008.01142.x

15. Riby DM, Doherty-Sneddon G, Bruce V. Atypical unfamiliar face processing in Williams syndrome: what can it tell us about typical familiarity effects? *Cogn Neuropsychiatry.* (2008) 13:47–58. doi: 10.1080/13546800701779206

16. Vivanti G, Hocking DR, Fanning P, Dissanayake C. The social nature of overimitation: Insights from autism and Williams syndrome. *Cognition.* (2017) 161:10–8. doi: 10.1016/j.cognition.2017.01.008

17. Lai PT, Ng R, Bellugi U. Parents' perspective on the social traits observed in school-age children with autism and children with Williams syndrome. *Res. Pract. Intellect. Dev. Disabil.* (2021) 2:174–84. doi: 10.1080/23297018.2021.1934893

18. Loveland KA, Tunali-Kotoski B. The School-Age Child with an Autistic Spectrum Disorder. In: Volkmar FR, Paul R, Klin A, Cohen D. (Eds.). *Handbook of autism and pervasive developmental disorders: Diagnosis, development, neurobiology, behavior.* John Wiley and Sons Inc. (2005) p. 247–287. doi: 10.1002/9780470939345.ch9

19. Asada K, Itakura S. Social phenotypes of autism spectrum disorders and Williams syndrome: similarities and differences. *Front Psychol.* (2012) 3:247. doi: 10.3389/fpsyg.2012.00247

20. Glod M, Riby DM, Rodgers J. Relationships between sensory processing, repetitive behaviors, anxiety, and intolerance of uncertainty in autism spectrum disorder and Williams syndrome. *Autism Res.* (2019) 12:759–65. doi: 10.1002/aur.2096

21. Klein-Tasman BP, Phillips KD, Lord CE, Mervis CB, Gallo F. Overlap with the autism spectrum in young children with Williams syndrome. *J Dev Behav Pediatr.* (2009) 30:289. doi: 10.1097/DBP.0b013e3181ad1f9a

22. Klein-Tasman BP, Li-Barber KT, Magargee ET. Honing in on the social phenotype in Williams syndrome using multiple measures and multiple raters. *J Autism Dev Disord.* (2011) 41:341–51. doi: 10.1007/s10803-010-1060-5

23. Boulton KA, Porter MA. Extending the positive bias in Williams syndrome: the influence of biographical information on attention allocation. *Dev Psychopathol.* (2020) 32:243–56. doi: 10.1017/S0954579418001712

24. Sifneos PE. The prevalence of 'alexithymic' characteristics in psychosomatic patients. *Psychother Psychosom.* (1973) 22:255–62. doi: 10.1159/000286529

25. Kinnaird E, Stewart C, Tchanturia K. Investigating alexithymia in autism: a systematic review and meta-analysis. *European Psychiatry.* (2019) 55:80–9. doi: 10.1016/j.eurpsy.2018.09.004

26. Rodgers J, Riby DM, Janes E, Connolly B, McConachie H. Anxiety repetitive behaviours in autism spectrum disorders and Williams syndrome: a cross-syndrome comparison. *J Autism Dev Disord.* (2012) 42:175–80. doi: 10.1007/s10803-011-1225-x

27. Jolliffe R, Adams D, Simpson K. Trait anxiety in individuals on the autism spectrum: a systematic review. *Rev J Autism Dev Disord.* (2022). doi: 10.1007/s40489-022-00308-8

28. Van Steensel FJ, Bögels SM, Perrin S. Anxiety disorders in children and adolescents with autistic spectrum disorders: a meta-analysis. *Clin Child Fam Psychol Rev.* (2011) 14:302–17. doi: 10.1007/s10567-011-0097-0

29. Leyfer OT, Woodruff-Borden J, Klein-Tasman BP, Fricke JS, Mervis CB. Prevalence of psychiatric disorders in 4 to 16-year-olds with Williams syndrome. *Am J Med Genet Part B Neuropsychiatr Genet.* (2006) 141B:615–22. doi: 10.1002/ajmg.b.30344

30. Royston R, Howlin P, Waite J, Oliver C. Anxiety disorders in Williams syndrome contrasted with intellectual disability and the general population: a systematic review and meta-analysis. *J Autism Dev Disord.* (2017) 47:3765–77. doi: 10.1007/s10803-016-2909-z

31. Dykens EM. Anxiety, fears and phobias in persons with Williams syndrome. *Dev Neuropsychol.* (2003) 23:291–316. doi: 10.1207/S15326942DN231&2_13

32. Graham JM, Rosner B, Dykens E, Visootsak J. Behavioral features of CHARGE syndrome (Hall-Hittner syndrome) comparison with Down syndrome, Prader-Willi syndrome, Williams syndrome. *Am J Med Genet.* (2005) 133:240–7. doi: 10.1002/ajmg.a.30543

33. Dimitropoulos A, Ho A, Klaiman C, Koenig K, Schultz R. A comparison of behavioral and emotional characteristics in children with autism, Prader-Willi syndrome, Williams syndrome. *J Ment Health Res Intellect Disabil.* (2009) 2:220–43. doi: 10.1080/19315860903052204

34. South M, Hanley M, Normansell-Mossa K, Russel NCC, Cawthorne T, Riby DM. "Intolerance of uncertainty" mediates the relationship between social profile and anxiety in both Williams syndrome and autism. *Autism Res.* (2021) 14:1986–95. doi: 10.1002/aur.2554

35. Phillips KD. *Emotion regulation and dysregulation in children and adolescents with Williams syndrome (Publication No. 3332181) [Doctoral dissertation]*. University of Wisconsin-Milwaukee. ProQuest Dissertations and Theses Global. (2008).

36. Pitts C, Klein-Tasman B, Osborne J, Mervis C. Predictors of specific phobia in children with Williams syndrome. *J Intellect Disabil Res.* (2016) 60:1031–42. doi: 10.1111/jir.12327

37. Woodruff-Borden J, Kistler DJ, Henderson DR, Crawford NA, Mervis CB. Longitudinal course of anxiety in children and adolescents with Williams syndrome. *Am J Med Genet C Semin Med Genet.* (2010) 154:277–90. doi: 10.1002/ajmg.c.30259

38. Dubey N, Podder P, Pandey D. Knowledge of COVID-19 and its influence on mindfulness, cognitive emotion regulation and psychological flexibility in the Indian community. *Front Psychol.* (2020) 11:589365. doi: 10.3389/fpsyg.2020.589365

39. Ferreira F, de O, Lopes-Silva JB, Siquara GM, Manfroi EC, De Freitas. PM. Coping in the Covid-19 pandemia: how different resources and strategies can be risk or protective factors to mental health in the Brazilian population. *Health Psychol Behav Med.* (2021) 9:182–205. doi: 10.1080/21642850.2021.1897595

40. Kim JH, Shim Y, Choi I, Choi E. The role of coping strategies in maintaining well-being during the COVID-19 outbreak in South Korea. *Soc Psychol Personal Sci.* 1–13. (2021). doi: 10.1177/1948550621990595

41. Petrocchi S, Levante A, Bianco F, Castelli I, Lecciso F. Maternal distress/coping and children's adaptive behaviors during the COVID-19 lockdown: mediation through children's emotional experience. *Front Public Health.* (2020) 8:587833. doi: 10.3389/fpubh.2020.587833

42. Wang H, Xia Q, Xiong Z, Li Z, Xiang W, Yuan Y, et al. The psychological distress and coping styles in the early stages of the 2019 coronavirus disease (COVID-19) epidemic in the general mainland Chinese population: A web-based survey. PLOS ONE. (2020) 15:e0233410. doi: 10.1371/journal.pone.0233410

43. Wang QQ, Fang YY, Huang HL, Lv WJ, Wang XX, Yang TT, et al. Anxiety, depression and cognitive emotion regulation strategies in Chinese nurses during the COVID-19 outbreak. *J Nurs Manag.* (2021) 29:1263–74. doi: 10.1111/jonm.13265

44. Yan L, Gan Y, Ding X, Wu J, Duan H. The relationship between perceived stress and emotional distress during the COVID-19 outbreak: effects of boredom proneness and coping style. *J Anxiety Disord.* (2021) 77:102328. doi: 10.1016/j.janxdis.2020.102328

45. Di Renzo M, Di Castelbianco FB, Vanadia E, Petrillo M, D'Errico S, Racinaro L, et al. (2020). Parent-reported behavioural changes in children with autism spectrum disorder during the COVID-19 lockdown in Italy. *Continuity in Education*. 1:117–25. doi: 10.5334/cie.20

46. Sideropoulos V, Dukes D, Hanley M, Palikara O, Rhodes S, Riby D, et al. The impact of COVID-19 on anxiety and worries for families of individuals with special education needs and disabilities in the UK. *J Autism Dev Disord.* (2021) 52:2656–69. doi: 10.31234/osf.io/gyhd9

47. Toseeb U, Asbury DK, Code A, Fox L, Deniz E. *Supporting families with children with special educational needs and disabilities during COVID-19*. PsyArXiv. (2020). Available online at: https://psyarxiv.com/tm69k/

48. Breaux R, Dvorsky MR, Marsh NP, Green CD, Cash AR, Shroff DM, et al. Prospective impact of COVID-19 on mental health functioning in adolescents with and without ADHD: protective role of emotion regulation abilities. *J Child Psychol Psychiatry.* (2021) 62:1132–9. doi: 10.1111/jcpp.13382

49. Van Herwegen J, Dukes D, Samson A. *COVID19 Crisis Response Survey for families of Individuals with Special Needs [Data set]*. OSFHOME. (2020). Available online at: osf.io/5nkq9 (accessed May 27, 2020).

50. Dukes D, Van Herwegen J, Alessandri M, Alnemary F, Rad JA, Lavenex PB, et al. Introducing the COVID-19 crisis special education needs coping survey. *PsyArXiv*. 1–15. (2021). doi: 10.31234/osf.io/rtswa

51. Turon H, Carey M, Boyes A, Hobden B, Dilworth S, Sanson-Fisher R. Agreement between a single-item measure of anxiety and depression and the Hospital Anxiety and Depression Scale: a cross-sectional study. *PLoS ONE*. (2019) 14: e0210111. doi: 10.1371/journal.pone.0210111

52. Lewis EJ, Yoon KL, Joormann J. Emotion regulation and biological stress responding: associations with worry, rumination, and reappraisal. *Cogn Emot.* (2018) 32:1487–98. doi: 10.1080/02699931.2017.1310088

53. Gross JJ, John OP. Individual differences in two emotion regulation processes: implications for affect, relationships, and well-being. *J Pers Soc Psychol.* (2003) 85:348–62. doi: 10.1037/0022-3514.85.2.348

54. Samson AC, Wells WM, Phillips JM, Hardan AY, Gross JJ. Emotion regulation in autism spectrum disorder: Evidence from parent interviews and children's daily diaries. *J Child Psychol Psychiatry.* (2015) 56:903–13. doi: 10.1111/jcpp.12370

55. Quoidbach J, Berry EV, Hansenne M, Mikolajczak M. Positive emotion regulation and well-being: comparing the impact of eight savoring and dampening strategies. *Pers Individ Dif.* (2010) 49:368–73. doi: 10.1016/j.paid.2010.03.048

56. Kugler L, Kuhbandner C. That's not funny!–But it should be: Effects of humorous emotion regulation on emotional experience and memory. *Front Psychol*. (2015) 6:1296. doi: 10.3389/fpsyg.2015.01296

57. Samson AC, Gross JJ. The dark and light sides of humor: An emotion-regulation perspective. In: Gruber J, Moskowitz TJ. (Eds.). *Positive emotion: Integrating the light sides and dark sides*. Oxford University Press. (2014) p. 169–182. doi: 10.1093/acprof:oso/9780199926725.003.0010

58. MATLAB. *Version 9.5.0 (R2018b). The MathWorks Inc*. (2018). Available online at: https://ch.mathworks.com/de/products/matlab.html

59. R Core Team. *R: A language and environment for statistical computing*. Vienna, Austria: R Foundation for Statistical Computing. (2020). Available online at: https://www.R-project.org/

60. Bates D, Maechler M, Bolker B, Walker S. Fitting linear mixed-effects models using lme4. *J Stat Softw.* (2015) 67:1–48. doi: 10.18637/jss.v067.i01

61. Kuznetsova A, Brockhoff PB, Christensen RHB. lmerTest package: tests in linear mixed effects models. *J Stat Softw*. (2017) 82:1–26. doi: 10.18637/jss.v082.i13

62. Lenth RV. *Emmeans: Estimated Marginal Means, aka Least-Squares Means. R package version 1.6.2-1.* (2021). Available online at: https://cran.r-project.org/web/packages/emmeans/index.html

63. Fox J, Weisberg S. Visualizing fit and lack of fit in complex regression models with predictor effect plots and partial residuals. *J Stat Softw.* (2018) 87:1–27. doi: 10.18637/jss.v087.i09

64. Wickham H. (2016). *ggplot2: Elegant Graphics for Data Analysis.* New York: Springer-Verlag. Available online at: https://ggplot2.tidyverse.org

65. Guller B, Yaylaci F, Eyuboglu D. Those in the shadow of the pandemic: impacts of the COVID-19 outbreak on the mental health of children with neurodevelopmental disorders and their parents. *Int J Dev Disabil.* (2021) 1–13. doi: 10.1080/20473869.2021.1930827

66. Mutluer T, Doenyas C, Aslan Genc H. Behavioral implications of the covid-19 process for autism spectrum disorder, and individuals' comprehension of and reactions to the pandemic conditions. *Front Psychiatry.* (2020) 11. doi: 10.3389/fpsyt.2020.561882

67. Kent R, Simonoff E. Prevalence of anxiety in autism spectrum disorders. In: Kerns CM, Renno P, Storch EA, Kendall PC, Wood JJ, editors. *Anxiety in Children and Adolescents with Autism Spectrum Disorder: Evidence-Based Assessment and Treatment.* Academic Press (2017). p. 5–32. doi: 10.1016/B978-0-12-805122-1.00002-8

68. Reardon T, Spence SH, Hesse J, Shakir A, Creswell C. Identifying children with anxiety disorders using brief versions of the Spence Children's Anxiety Scale for children, parents, and teachers. *Psychol Assess.* (2018) 30:1342–55. doi: 10.1037/pas0000570

69. Davies M, Udwin O, Howlin P. Adults with Williams syndrome: preliminary study of social, emotional, behavioural difficulties. *Br J Psychiatry.* (1998) 172:273–6. doi: 10.1192/bjp.172.3.273

70. Ng R, Järvinen A, Bellugi U. Toward a deeper characterization of the social phenotype of Williams syndrome: the association between personality and social drive. *Res Dev Disabil.* (2014) 35:1838–49. doi: 10.1016/j.ridd.2014.04.015

71. Treichel N, Dukes D, Barisnikov K, Samson AC. How cognitive, social, and emotional profiles impact humor appreciation: Sense of humor in autism spectrum disorder and Williams syndrome. *Humor Res.* (2022) 35:113–33. doi: 10.1515/humor-2021-0038

72. Valla JM, Belmonte MK. Detail-oriented cognitive style and social communicative deficits, within and beyond the autism spectrum: Independent traits that grow into developmental interdependence. *Dev Rev.* (2013) 33:371–98. doi: 10.1016/j.dr.2013.08.004

73. Gotham K, Bishop SL, Hus V, Huerta M, Lund S, Buja A, et al. Exploring the relationship between anxiety and insistence on sameness in autism spectrum disorders. *Autism Res.* (2013) 6:33–41. doi: 10.1002/aur.1263

74. McClure KS, Halpern J, Wolper PA, Donahue JJ. Emotion regulation and intellectual disability. *J Develop Disabil.* (2009) 15:38–44.

75. Benson BA, Fuchs C. Anger-arousing situations and coping responses of aggressive adults with intellectual disability. *J Intellect Develop Disabil.* (1999) 24:207–14. doi: 10.1080/13668259900033991

76. te Brinke LW, Schuiringa HD, Matthys W. Emotion regulation and angry mood among adolescents with externalizing problems and intellectual disabilities. *Res Develop Disabil.* (2021) 109:103833. doi: 10.1016/j.ridd.2020.103833

77. Sheppes G, Gross JJ. Is timing everything? Temporal considerations in emotion regulation. *Pers Soc Psychol Rev.* (2011) 15:319–31. doi: 10.1177/1088868310395778

78. Stinson L, Liu, Dallery J. Ecological momentary assessment: A systematic review of validity research. *Pers Behav Sci.* (2022) 45:469–93. doi: 10.1007/s40614-022-00339-w

79. Klein-Tasman BP, Young BN, Levine K, Rivera K, Miecielica EJ, Yund BD, et al. Acceptability and effectiveness of humor- and play-infused exposure therapy for fears in Williams syndrome. Evidence-based practice in child and adolescent. *MentHealth.* (2022) 1:94–111. doi: 10.1080/23794925.2021.1950080

80. Zaharia A, Noir-Kahlo K, Bressoud N, Sander D, Dukes D, Samson AC. Proof of concept: a brief psycho-educational training program to increase the use of positive emotion regulation strategies in individuals with autism spectrum disorder. *Front Psychol.* (2021) 12:705937. doi: 10.3389/fpsyg.2021.705937

81. Samson AC, Hegenloh M. Stimulus properties affect humor processing in individuals with Asperger syndrome. *J Autism Dev Disord.* (2010) 40:438–47. doi: 10.1007/s10803-009-0885-2

82. Samson AC. Humor(lessness) elucidated - sense of humor in individuals with Autism Spectrum Disorders: Review and introduction. *Int J Humor Res.* (2013) 26:393–409. doi: 10.1515/humor-2013-0027

83. Lord C, Rutter M, DiLavore P, Risi S, Gotham K, Bishop S. *Autism diagnostic observation schedule—2nd edition (ADOS-2).* Los Angeles, CA: Western Psychological Corporation. (2012).

84. Bergua V, Meillon C, Potvin O, Ritchie K, Tzourio C, Bouisson J, et al. Short STAI-Y anxiety scales: validation and normative data for elderly subjects. *Aging Ment health.* (2016) 20:987–95. doi: 10.1080/13607863.2015.1051511

85. Kroenke K, Spitzer RL, Williams JBW, Löwe B. An ultra-brief screening scale for anxiety and depression: the PHQ–4. *Psychosomatics.* (2009) 50: 613–21. doi: 10.1016/S0033-3182(09)70864-3

86. McKinley S, Coote K, Stein-Parbury J. Development testing of a Faces Scale for the assessment of anxiety in critically ill patients. *J Adv Nurs.* (2003) 41:73–9. doi: 10.1046/j.1365-2648.2003.02508.x

87. Gullone E, Hughes EK, King NJ, Tonge B. The normative development of emotion regulation strategy use in children and adolescents: a 2-year follow-up study. *J Child Psychol Psychiatry.* (2010) 51:567–74. doi: 10.1111/j.1469-7610.2009.02183.x

88. Sheppes G, Scheibe S, Suri G, Gross JJ. Emotion regulation choice. *Psychological Sci.* (2011) 22:1391–6. doi: 10.1177/0956797611418350

89. Cai RY, Samson AC. Emotion regulation in individuals on the autism spectrum. In: Gross JJ, Ford B. (Eds). *Handbook of Emotion Regulation. [Manuscript accepted for publication].* (2022).

frontiers | Frontiers in Psychiatry

TYPE Clinical Trial
PUBLISHED 06 December 2022
DOI 10.3389/fpsyt.2022.1034722

OPEN ACCESS

EDITED BY
Ru Ying Cai,
Autism Spectrum Australia, Australia

REVIEWED BY
Anouk Schrantee,
Amsterdam University Medical Center, Netherlands
Delshad Shroff,
Virginia Tech, United States
Ursula Pauli-Pott,
Philipps-University Marburg, Germany

*CORRESPONDENCE
Michael J. Kofler
kofler@psy.fsu.edu

SPECIALTY SECTION
This article was submitted to Autism, a section of the journal Frontiers in Psychiatry

RECEIVED 01 September 2022
ACCEPTED 17 November 2022
PUBLISHED 06 December 2022

CITATION
Groves NB, Chan ESM, Marsh CL, Gaye F, Jaisle EM and Kofler MJ (2022) Does central executive training and/or inhibitory control training improve emotion regulation for children with attention-deficit/hyperactivity disorder? A randomized controlled trial.
Front. Psychiatry 13:1034722.
doi: 10.3389/fpsyt.2022.1034722

Does central executive training and/or inhibitory control training improve emotion regulation for children with attention-deficit/hyperactivity disorder? A randomized controlled trial

Nicole B. Groves[1], Elizabeth S. M. Chan[1], Carolyn L. Marsh[1], Fatou Gaye[1], Emma M. Jaisle[2] and Michael J. Kofler[1]*

[1]Department of Psychology, Florida State University, Tallahassee, FL, United States, [2]Department of Psychology, Florida International University, Miami, FL, United States

Introduction: Approximately 48–54% of children with attention-deficit/hyperactivity disorder (ADHD) have impairing difficulties with emotion regulation, and these difficulties are not ameliorated by first-line ADHD treatments. Working memory and inhibitory control represent promising intervention targets given their functional, if not causal, links with ADHD-related emotion dysregulation.

Methods: This preregistered randomized controlled trial tested whether two digital therapeutic training protocols that have been previously shown to improve working memory (Central Executive Training [CET]) and inhibitory control (Inhibitory Control Training [ICT]) can improve emotion regulation in a sample of 94 children with ADHD aged 8–13 years ($M = 10.22$, $SD = 1.43$; 76% White/non-Hispanic; 29 girls).

Results: Results of Bayesian mixed model ANOVAs indicated both treatment groups demonstrated significant decreases in emotion dysregulation relative to pre-treatment at immediate post-treatment (parent report; $d = 1.25$, $BF_{10} = 8.04 \times 10^{13}$, $p < 0.001$), at 1–2 months after completing treatment (teacher report; $d = 0.99$, $BF_{10} = 1.22 \times 10^{6}$, $p < 0.001$), and at 2–4-months follow-up (parent report; $d = 1.22$, $BF_{10} = 1.15 \times 10^{14}$, $p < 0.001$). Contrary to our hypotheses, the CET and ICT groups demonstrated equivalent reductions in emotion dysregulation and maintenance of effects. Exploratory analyses revealed that results were robust to control for informant expectancies, ADHD medication status/changes, in-person vs. at-home treatment, child age, and time from treatment completion to post-treatment ratings.

Discussion: To determine whether working memory and inhibitory control are causally linked with ADHD-related emotion dysregulation, future studies

should include active control conditions that do not train executive functions prior to making decisions about the clinical utility of CET/ICT for the treatment of emotion dysregulation in ADHD.

Clinical trial registration: [https://clinicaltrials.gov/], identifier [NCT03324464].

KEYWORDS

attention-deficit/hyperactivity disorder (ADHD), emotion regulation, working memory, inhibitory control, executive function

Introduction

Attention-deficit/hyperactivity disorder (ADHD) is a neurodevelopmental disorder characterized by impairing symptoms of inattention, hyperactivity, and impulsivity (1). ADHD affects approximately 5% of school-aged children (2), and most children with ADHD experience difficulties with emotion regulation that, in turn, portend significantly greater distress and impairment than ADHD symptoms alone (3). Unfortunately, the limited available literature suggests that evidence-based treatments for ADHD (4)—including psychostimulants (5) and behavioral parent training (6)—often do not reduce emotion dysregulation, suggesting the need for interventions that directly target factors that underlie ADHD-related emotion dysregulation. Working memory and potentially inhibitory control have been linked functionally with ADHD-related emotion dysregulation (7–10), suggesting that they may reflect promising intervention targets for producing downstream improvements in emotion regulation for children with ADHD. The present randomized controlled trial tests the extent to which two cognitive training protocols that have been previously shown to improve working memory (Central Executive Training [CET]) and inhibitory control (Inhibitory Control Training [ICT]) (11) can improve emotion regulation for children with ADHD.

Emotion regulation and executive functioning in attention-deficit/hyperactivity disorder

Emotion regulation refers to the ability to modulate the speed and intensity of emotional escalation and de-escalation, and it involves complex physiological, experiential, and behavioral processes (12, 13). Approximately 48–54% of children with ADHD exhibit comorbid difficulties with emotion regulation based on recent meta-analytic evidence [$d = 0.80$–0.95; (3)]. Mitigating emotion dysregulation in ADHD is imperative given that it increases the already large burden of illness associated with ADHD (14), including predicting greater academic and social impairment (15–17), higher rates of healthcare utilization (16), and higher daily parenting stress (18) than ADHD symptoms alone (19). Additionally, emotion dysregulation persists into adulthood for many people with ADHD (20–22), and portends increased risk for the development of comorbid psychopathology [e.g., oppositional defiant disorder, anxiety, depression; (23)].

Several conceptual models have been proposed to explain the high prevalence and adverse outcomes of emotion dysregulation in children with ADHD (3, 24), with growing acknowledgment that the phenomenology, etiology, course, and correlates of emotion dysregulation are likely as heterogenous as most other ADHD-related symptoms (25–28). Of the proposed mechanisms linking ADHD and emotion regulation, underdeveloped executive functions—particularly working memory and inhibitory control—reflect promising investigative targets. Working memory refers to the active, top-down manipulation of information held in short-term memory (29), and is impaired in 68–85% of children with ADHD (30–32). Inhibitory control refers to a set of interrelated cognitive processes that underlie the ability to withhold (action restraint) or stop (action cancelation) an ongoing behavioral response (33), and is impaired in 21–46% of children with ADHD (28, 34, 35).

Working memory and inhibitory control have each been theorized as core deficits underlying ADHD-related behavioral symptoms and functional outcomes [e.g., (36, 37)], including emotion dysregulation (38). Conceptually, intact executive functions are necessary to regulate the generation and expression of emotion at each of its theorized stages. That is, emotion generation and expression require one to select and modify situations, actively attend to stimuli, form cognitive interpretations of events, and modulate responses accordingly (39). Deficits in working memory and/or inhibitory control may lead to a breakdown in any of these complex processes. Indeed, working memory predicts emotion regulation in samples of children with ADHD with and without common comorbidities (7, 9). Additionally, experimental evidence

indicates that increasing working memory demands produces disproportionate increases in negative emotional expression for children with ADHD relative to their typically developing counterparts, suggesting that working memory is functionally related to emotion dysregulation in ADHD (10). Inhibitory control has also been associated with emotion regulation in children with ADHD (9, 40), and there is some evidence suggesting this relation is causal in other populations [i.e., emotionally dysregulated adults; (41)]. Interestingly, however, when considered together, only working memory uniquely predicted emotion regulation, whereas inhibitory control did not (8).

Executive function interventions for emotion dysregulation in other populations

Given the established links among working memory, inhibitory control, and emotion regulation, there has been increased interest in the extent to which training these executive functions can improve emotion regulation in various populations. For example, training working memory using emotional/affective stimuli has improved emotion regulation in samples of neurotypical adults (42–44) as well as clinical samples of adolescents and adults (45, 46). In contrast, findings are more mixed for inhibitory control, such that training inhibitory control improved emotion regulation in adults with elevated emotional reactivity (41) but did not affect emotion regulation in neurotypical adults (47). Finally, one study trained both working memory and inhibitory control, and reported improved emotion regulation in typically developing preschoolers relative to waitlist controls (48). Taken together, the evidence supporting training working memory and/or inhibitory control for improving emotion regulation in non-ADHD samples is promising, albeit mixed.

Executive function interventions for emotion dysregulation in children with attention-deficit/hyperactivity disorder

Both working memory and potentially inhibitory control may be functionally related to emotion regulation difficulties in children with ADHD (9, 10), but, to our knowledge, no study to date has examined whether emotion regulation improves in children with ADHD following targeted training of executive functioning (49). In a partial exception, Tamm et al. (50–52) reported that a play-based (non-computerized) metacognitive attention training for preschoolers with ADHD failed to improve emotion regulation based on parent report. Similarly, in a randomized controlled trial comparing the efficacy of training all, some, or no executive functions, impacts on most cognitive and behavioral outcomes were non-specific and not attributable to any particular treatment target (53). Additionally, meta-analytic evidence suggests that, even though direct training may improve working memory and inhibitory control for preschoolers with ADHD and related externalizing behaviors, effects on behavioral outcomes are not significant (54). However, the extent to which similar findings would be obtained by training specific executive functions in school-aged samples of children with ADHD remains unknown.

For children with ADHD, evaluating the efficacy of executive function training protocols is further complicated by target misspecification and related issues of first-generation protocols that significantly limit their potential to produce downstream effects on behavior and functioning [for reviews see (49, 55)]. Specifically, most executive function training protocols have historically not produced the intended improvements in the executive function(s) they were intended to train (49, 56, 57). Indeed, most executive function training protocols have been shown to train cognitive abilities—such as short-term memory capacity rather than central executive working memory processes (29)—that are not impaired in most children with ADHD and, in most cases, are unrelated to ADHD symptoms and functional outcomes even cross-sectionally (11, 49). Thus, their lack of downstream effects on behavior and functioning is unsurprising to the extent that they are training cognitive abilities that generally do not support the behaviors we are trying to modify [e.g., (55)].

To address these limitations, our group created two translational, evidence-based, digital therapeutic treatments that include gaming elements (11, 34, 58). These computerized treatments incrementally increase demands on their target processes (central executive working memory for CET, inhibitory control for ICT). In previous clinical trials, CET and ICT were both rated as acceptable and feasible by parents and children (11, 34). In terms of affecting their respective cognitive training targets, CET was superior to gold-standard behavioral parent training (34) and ICT (11) for improving working memory performance. Similarly, ICT was superior to CET for improving inhibitory control, albeit only on one of two outcome tests, demonstrating that both CET and ICT successfully improve their targeted executive function (11). In terms of clinical outcomes, CET produced improvements in parent-rated ADHD symptoms that were equivalent to those obtained from gold-standard behavioral parent training, and CET was superior to behavioral parent training for decreasing objectively assessed hyperactivity in a sequential recruitment controlled trial (34). In a double-blind randomized controlled trial comparing CET and ICT, CET was superior to ICT in reducing parent- and teacher-reported ADHD symptoms and objectively assessed hyperactivity; CET-related ADHD symptom improvement was also maintained at 2–4 months follow up (11).

Current study

Given that CET and ICT (a) effectively improve working memory and inhibitory control processes, respectively, that are linked with ADHD-related emotion dysregulation; and (b) have been shown previously to produce clinically meaningful improvements in other behavioral and functional domains for children with ADHD (11, 26, 59), we hypothesized that both CET and ICT would produce improvements in emotion regulation. In addition, given evidence that working memory is better than inhibitory control at predicting emotion regulation for children with ADHD (8), as well as prior evidence that CET produced superior improvements relative to ICT on other behavioral and functional outcomes (11, 59), we hypothesized that CET would be superior to ICT for reducing emotion dysregulation.

Methods

Study timeline, randomization, allocation concealment, and masking

The current study reports on secondary outcomes from a randomized clinical trial of CET vs. ICT for ADHD (11) (Table 1). The sample reflects consecutive referrals from March 2017 to April 2021. Prior to March 2020, children ($n = 73$; 77.7%) completed three visits during the pre-treatment evaluation, lasting approximately 3.5 h each. These children also completed testing sessions at mid-treatment, post-treatment, and 2–4 months follow up. Four treatment cases were lost to follow-up from March to June 2020 as the study was shut down due to COVID-19. Procedures were adjusted when the study resumed in June 2020 to minimize face-to-face contact, including reducing the pre-treatment battery to a single 4-h testing session that included the use of face masks and social distancing for participants and study team members ($n = 21$; 22.3%). Additionally, weekly treatment sessions that occurred in the clinic prior to the shutdown were conducted via telehealth for all children beginning in June 2020, and in-clinic mid/post/follow-up child testing was discontinued. Treatment delivery format (pre-COVID face-to-face vs. peri-COVID telehealth) was probed as a covariate as described below. Parents (pre/mid/post/2–4 months follow-up) and teachers (pre/post) completed measures at each time point according to the original protocol. Teacher questionnaires were sent during the post-treatment session and were completed by teachers approximately 1–2 months post-treatment (Figure 1).

Randomization was conducted by the study methodologist using unpredictable allocation stratified by medication status according to CONSORT guidelines. Study evaluators were masked to treatment group. Data screening, cleaning, and analyses were conducted masked to treatment group. Best practice guidelines for cognitive training studies were closely followed as described in Table 2.

TABLE 1 Sample demographic and pre-treatment characteristics.

Variable	ICT		CET		Cohen's *d*	BF_{01}	*p*
	M	*SD*	*M*	*SD*			
N (boys/girls)	50 (34/16)		44 (31/13)		–	4.15	0.80, *ns*
Age	10.44	1.53	9.96	1.29	0.34	1.43	0.11, *ns*
SES	48.88	9.82	47.06	11.70	0.17	3.42	0.42, *ns*
WISC-V VCI	105.9	1.52	100.82	13.60	0.36	0.86	0.053, *ns*
Race/Ethnicity (W, B, H, MR)	36, 6, 4, 4		35, 5, 3, 1		–	34.48	0.64, *ns*
Medication (no/yes)	34/16		28/16		–	3.77	0.66, *ns*
ADHD presentation (I, H/I, C)	16, 1, 33		14, 1, 29		–	66.67	0.99, *ns*
Comorbidity (no/yes)	18/32		16/28		–	31.25	0.29, *ns*
BASC-3 emotional self-control (total raw score)							
Parent	11.86	6.44	11.34	5.40	0.09	4.26	0.68, *ns*
Teacher	9.44	5.46	11.15	7.40	0.27	2.22	0.21, *ns*
BASC-3 negative emotionality (total raw score)							
Parent	8.60	4.54	8.66	4.03	0.01	4.61	0.95, *ns*
Teacher	4.96	3.46	5.98	4.85	0.25	2.48	0.25, *ns*

B, black; BASC-3, behavior assessment scale for children; C, combined presentation; CET, central executive training; H, Hispanic/Latino; H/I, predominantly hyperactive/impulsive presentation; I, predominantly inattentive presentation; ICT, inhibitory control training; MR, multiracial; SES, Hollingshead SES total score; W, white/non-Hispanic; WISC-V VCI, WISC-V verbal comprehension index standard score; BF, Bayes factor, BF_{01} is the odds ratio of the evidence favoring the null to the evidence favoring the alternative hypothesis. A value of 1 indicates that the data are equally likely under the null and alternative hypotheses, values >1 favor the null hypothesis that the groups are equivalent, and values >3 are considered statistically significant evidence of equivalence.

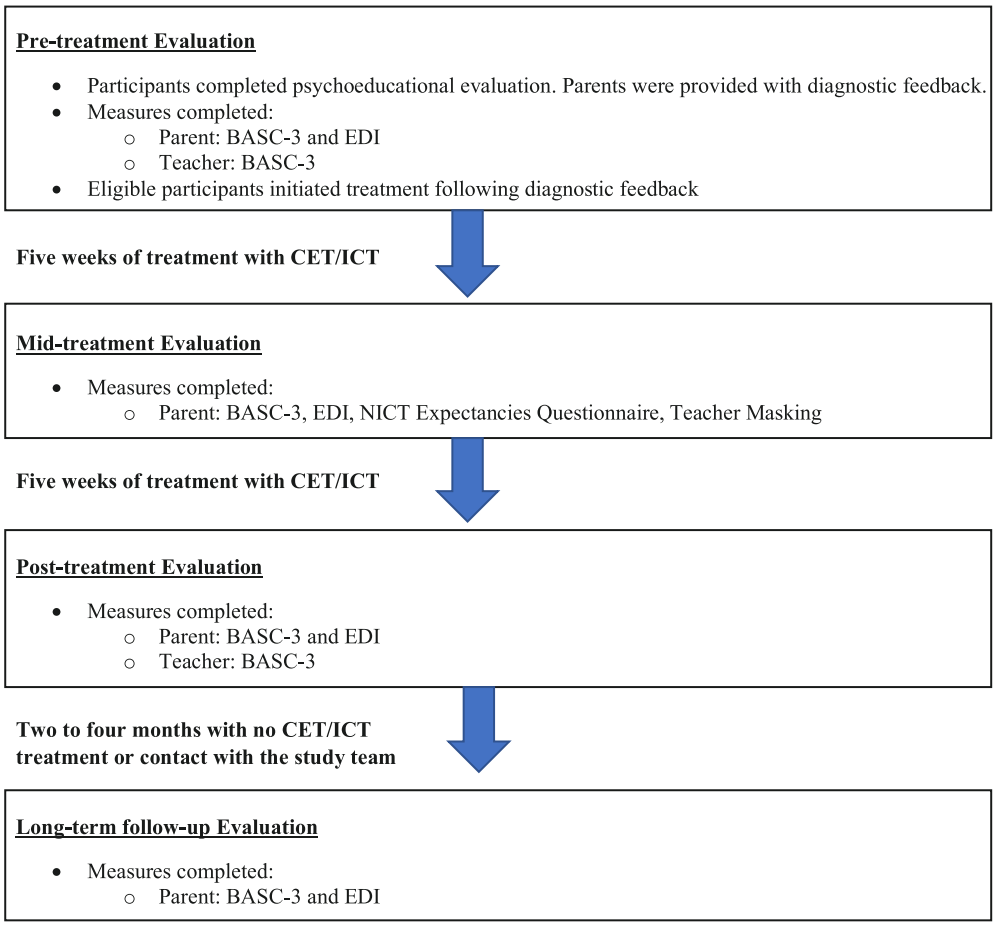


FIGURE 1
Study timeline. BASC-3, behavior assessment scale for children; CET, central executive training; EDI, emotion dysregulation inventory; ICT, inhibitory control training.

Participants

As shown in Table 1, the treated sample comprised 94 children with ADHD aged 8–13 years ($M = 10.22$, $SD = 1.43$; 29 girls) from the Southeastern US, consecutively referred to a university-based research clinic through community resources. Psychoeducational evaluations were provided to caregivers. IRB approval was obtained/maintained; all parents/children gave informed consent/assent. Child race/ethnicity was mixed, with 71 (75.5%) White/Non-Hispanic, 11 (11.7%) Black/African American, 7 (7.4%) Hispanic/Latino, and 5 (5.3%) multi-racial/ethnic children. All participants spoke English.

Inclusion/exclusion criteria

All families completed a comprehensive evaluation that included detailed semi-structured clinical interviewing [K-SADS; (60)] and age/sex norm-referenced parent and teacher ADHD ratings [ADHD-Rating Scale-5 and Behavior Assessment Scale for Children-3; (61, 62)]. Study eligibility required: (1) DSM-5 diagnosis of ADHD (any presentation) by the directing clinical psychologist and multidisciplinary treatment team based on K-SADS (2013 update for DSM-5) and differential diagnosis considering all available clinical information indicating onset, course, duration, and severity of ADHD symptoms consistent with the ADHD neurodevelopmental syndrome; (2) clinical/borderline elevations on at least one parent and one teacher ADHD rating scale (i.e., >90th percentile), or previous psychoeducational evaluation documenting cross-informant symptoms (e.g., for children prescribed medication that reduces ADHD symptoms at school); and (3) current impairment per K-SADS. Diagnoses comorbid with ADHD in the current sample included anxiety (31.9%), specific learning (22.3%), autism spectrum (13.8%), oppositional defiant (6.4%), and depressive (2.1%) disorders. Additional details regarding the psychoeducational evaluation

TABLE 2 Critical evaluation of the current study relative to best practice guidelines for cognitive training methodology and reporting standards [adapted from (66) and (75)].

	Criterion/*commentary*
Best practice recommendations from Simons et al. (66)	
√	Assess pre-treatment baseline performance for all groups
	The current study used a pre/mid/post (parent) and pre/post (teacher) design in which outcomes were assessed at a priori specified time points that included pre-treatment. Pre-treatment performance was assessed and controlled when probing between-group differences at post-treatment.
√	Include an active, credible control group matched for expectancies
	Working memory and inhibitory control are both putative core mechanisms implicated in ADHD-related emotion dysregulation. The two versions of the intervention are identical in all aspects except the target mechanism, and served as active, credible controls for each other. The CET and ICT interventions have been shown previously to be identical or not differ significantly in terms of expectancies as well as caregiver- and child-reported feasibility and acceptability (11).
√	Include at least 20 participants in each treatment arm
	All analyses include ICT n = 50 and CET n = 44 participants.
√	Randomly assign children to condition
	Children were randomly assigned using unpredictable allocation concealment.
√	Pre-register the trial, and explicitly acknowledge departures from pre-registered plan
	The trial was preregistered [https://osf.io/abwms]. All collected emotion regulation outcomes measures were reported. Data analyses were conducted masked to treatment allocation.
√	Mask raters for all subjective outcome measures
	Teachers were masked to treatment status and allocation condition. Caregivers were masked to allocation condition. However, caregivers were not masked to the fact that their child was receiving an intervention because they are active participants in both treatments (66). *Meta-analytic evidence indicates that estimates of treatment effects are inflated for unmasked raters vs. masked raters by d = 0.36–0.40 for neurocognitive training studies* (49).
√	Label any analyses conducted after inspecting the data as "exploratory"
	The analyses reported herein did not depart from the a priori plan, with one clearly marked exception related to an administrative error during data collection for our secondary outcome measure (EDI). Analytic decisions regarding this measure were made based on missing data counts without knowledge of their effects on study results.
√	Avoid subgroup analyses unless preregistered
	No subgroup analyses were preregistered; therefore, none were conducted. Within-group analyses were limited to planned comparisons to characterize the pattern of change for each group across assessment points.
√	Identify all outcome data collected, including outcomes not reported herein
	A complete list of data collected for secondary research questions can be found on the study's OSF preregistration website.
Additional recommendations from Redick (75)	
√	Report full pre-test and post-test means and SDs for all groups
	Pre-treatment and post-treatment means and SDs *are shown in* ***Tables 1, 3****, respectively.*
√	Provide full, subject-level data as supplementary material
	JASP (.jasp) data files are posted for peer review on the study's OSF website [https://osf.io/86fwu/].
√	Use likelihood ratios, in particular Bayes Factors
	Traditional p*-values are supplemented with Bayes Factors to allow stronger conclusions regarding both between-group equivalence and emerging between-group differences.*
√	Examine outcomes graphically to ensure that the pattern of pre- to post-test change is theoretically consistent with the expected pattern of results
	Graphical representations of study outcomes are shown in ***Figure 3****.*

and differential diagnosis process can be found on our preregistration website: https://osf.io/abwms.

As shown in the CONSORT diagram (**Figure 2**), a total of 112 children with ADHD were evaluated; $n = 16$ were eligible but declined participation, and $n = 2$ were excluded due to average or better performance on all pretreatment working memory tests, resulting in a total treated sample of 94 (83.9% of eligible cases). No inhibitory control thresholds were set as specified in our NIMH grant. Children with ADHD that did vs. did not participate in the treatment phase of the study did not differ on age, sex, SES, race/ethnicity, IQ, medication status, ADHD presentation, and the presence of common comorbidities (all $p \geq 0.20$). Untreated children with ADHD did not differ from treated children with ADHD on parent-reported emotion dysregulation ($p = 0.21$–0.27), but they had moderately higher teacher-reported emotion dysregulation ($p = 0.02$–0.03; $d = 0.57$–0.61) than the children with ADHD who participated in the treatment phase of the study. Children that were not randomized to CET or ICT were not followed past the pre-treatment evaluation.

Children were excluded from the larger study for gross neurological, sensory, or motor impairment; seizure disorder, psychosis, or intellectual disability; or non-stimulant medications that could not be withheld for testing.

TABLE 3 Post-treatment outcome data and covariates for exploratory analyses.

Variable	ICT ($n = 50$)		CET ($n = 44$)		Cohen's d	BF_{01}	p
	M	*SD*	*M*	*SD*			
Primary Outcomes							
BASC-3 emotional self-control (total raw score)							
Parent	8.60	5.36	8.99	4.46	0.08	4.31	0.70, *ns*
Teacher	6.11	4.26	8.18	5.76	0.41	0.80	0.05, *ns*
BASC-3 negative emotionality (total raw score)							
Parent	6.31	3.48	7.12	3.27	0.24	2.57	0.25, *ns*
Teacher	3.67	3.25	4.38	4.08	0.19	3.13	0.35, *ns*
Other Treatment-Related Variables							
Medication changes (stop, no, add)	2, 33, 15		2, 29, 13		–	20.41	0.99, *ns*
Informant expectancies							
Parent NICT expectancy score (mean raw score)	4.53	1.01	4.67	0.74	0.15	3.65	0.47, *ns*
Teacher masked to study (yes/no)	30/20		27/17		–	4.02	0.89, *ns*
COVID-19 telehealth (yes/no)	11/39		10/34		–	4.69	0.93, *ns*
Time elapsed from treatment to post ratings (days)							
Parent	17.02	31.60	16.10	34.31	0.03	4.55	0.89, *ns*
Teacher	40.33	42.19	37.80	35.63	0.06	4.35	0.75, *ns*

CET, central executive training; ICT, inhibitory control training; BASC-3, behavior assessment scale for children; effect sizes and statistical tests for the BASC subscales reflect control for pre-treatment scores for the same subscale. BF, Bayes factor, BF_{01} is the odds ratio of the evidence favoring the null to the evidence favoring the alternative hypothesis. A value of 1 indicates that the data are equally likely under the null and alternative hypotheses, values >1 favor the null hypothesis that the groups are equivalent, and values >3 are considered statistically significant evidence of equivalence.

Procedures

As detailed in Kofler et al. (11), identical procedures were used for both treatment groups. Both CET and ICT are 10-week digital therapeutic treatments accessed via computer or mobile device. Once a week, children were monitored by study staff for a 1-h session while they completed their training exercises in-office (pre-COVID) or via telehealth according to identical, manualized procedures. Additional weekly training sessions were parent-supervised, in-home training (goal: 15-min/day, 2–3 days/week). Weekly in-office (pre-COVID) or telehealth parent check-ins were also included to promote adherence and troubleshoot difficulties with the at-home training. No active treatment components are included in the parent check-ins, which were identical across groups.

Treatments

Central executive training and ICT each contain nine games, with each game created to train various functions/modalities of their respective targeted executive function. Both treatments include an automated token economy in which children receive virtual "tickets" for successful responses during games, completing games, and completing the daily "mission mode" to facilitate increased engagement in the task and reinforcement of training targets. Tickets were exchanged for tangible prizes that children collected during weekly in-office sessions or intermittently throughout treatment if they participated via telehealth due to COVID-19. The daily "mission mode" consists of three games that the child has not recently played that are selected by the software and must be completed prior to having access to all nine games, and it is designed to ensure appropriate breadth of training. Please see Kofler et al. (34) and Kofler et al. (11) for a more detailed description and rationale of the treatments' active control, adaptive training, and methods for maximizing dosage. Both interventions have been shown to have high feasibility and acceptability in terms of high parent satisfaction, high child-reported ease of use, and total child training time (11, 34).

Central executive training

The computerized CET protocol focused on improving children's working memory (11, 34). CET contains nine games that train each of the three primary central executive processes–updating, dual-processing, and temporal/serial reordering (30)–using three different stimulus modalities—verbal/phonological, visual, and spatial. CET's algorithms facilitate continually adaptive training by dynamically adjusting various parameters depending on the training target to incrementally increase central executive demands. Such parameters include target density, categories: stimuli ratio, target: non-target stimuli ratio, visual discriminability, and search space size. For example, increasing the search space size produces greater visual saccades, which, in turn, increases central executive demands during spatial working memory tasks because these saccades interrupt spatial rehearsal (63, 64).

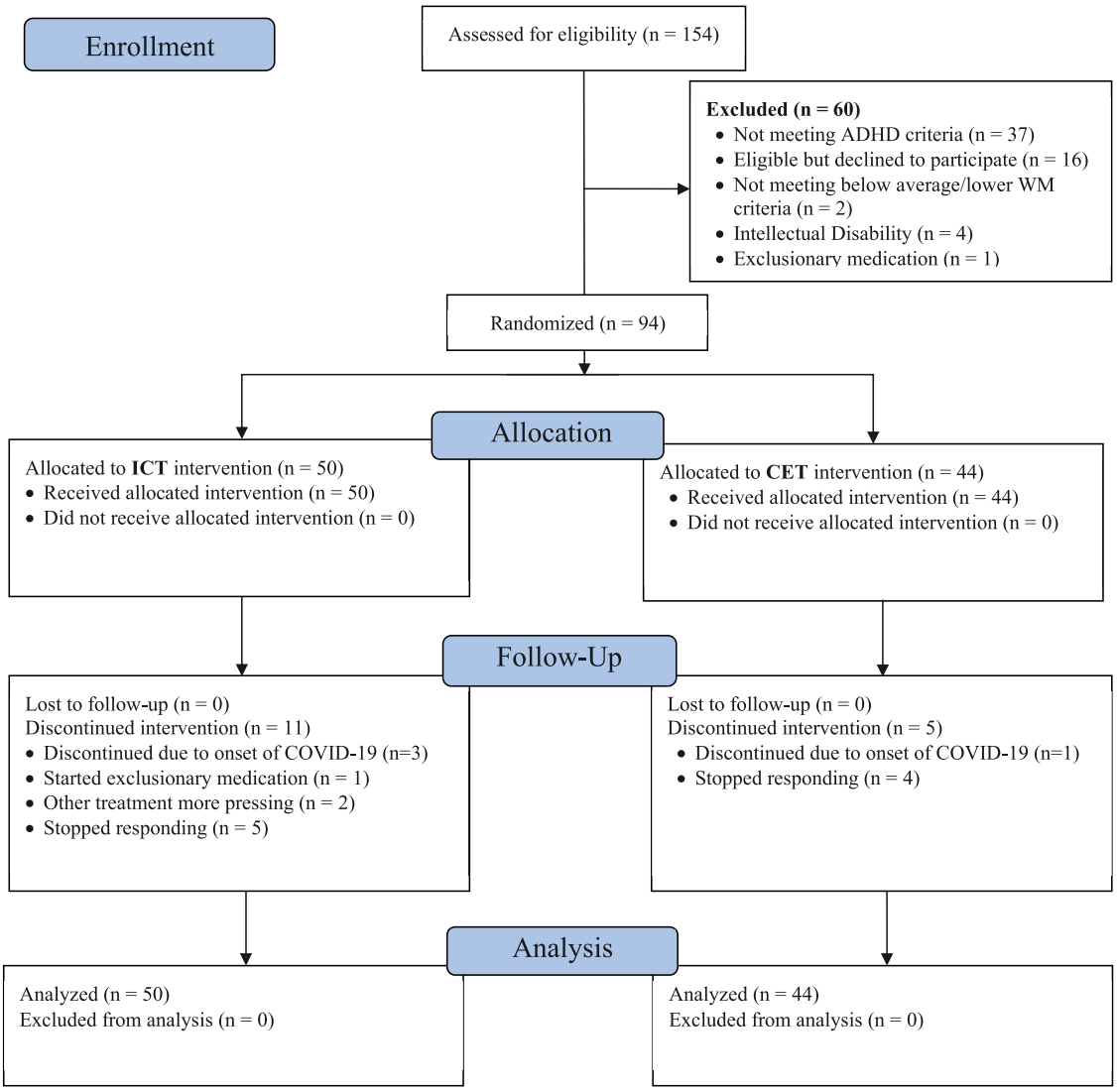


FIGURE 2
CONSORT diagram. The 154 children assessed for eligibility include all children recruited for evaluation in our research clinic during the study timespan, regardless of recruitment reason (because families would have been offered the intervention trial if their child was diagnosed with ADHD and otherwise eligible). The number of confirmed ADHD cases who were considered for eligibility is 112, of whom 94 (83.9%) were randomized and treated.

Inhibitory control training

The computerized ICT protocol focused on improving the "action restraint" and "action cancelation" components of inhibitory control (65). ICT was developed as an active, credible control comparison for CET based on best practice guidelines for rigorous digital therapeutic treatment trials (66). As such, each of the 9 ICT games contains an identical website address, name, art, animations, storyline, layout, interface, and use of adaptive training algorithms as its CET counterpart. Similarly, ICT dynamically adjusts parameters such as go:stop target ratio, presentation rate, response speed (timers), and number of stimuli (65) to ensure incremental increases in inhibitory control demands. For example, stretching the target density (i.e., increasing the proportion of "go" trials) increases inhibition demands by increasing prepotency, which makes it more difficult to inhibit during infrequently occurring "stop" trials (67).

Secondary intervention outcome assessment (emotion regulation)

Behavior assessment scale for children-3

The BASC-3 (62) contains two subscales that assess emotion regulation based on parent and teacher report: *Emotional Self-Control* and *Negative Emotionality*. The

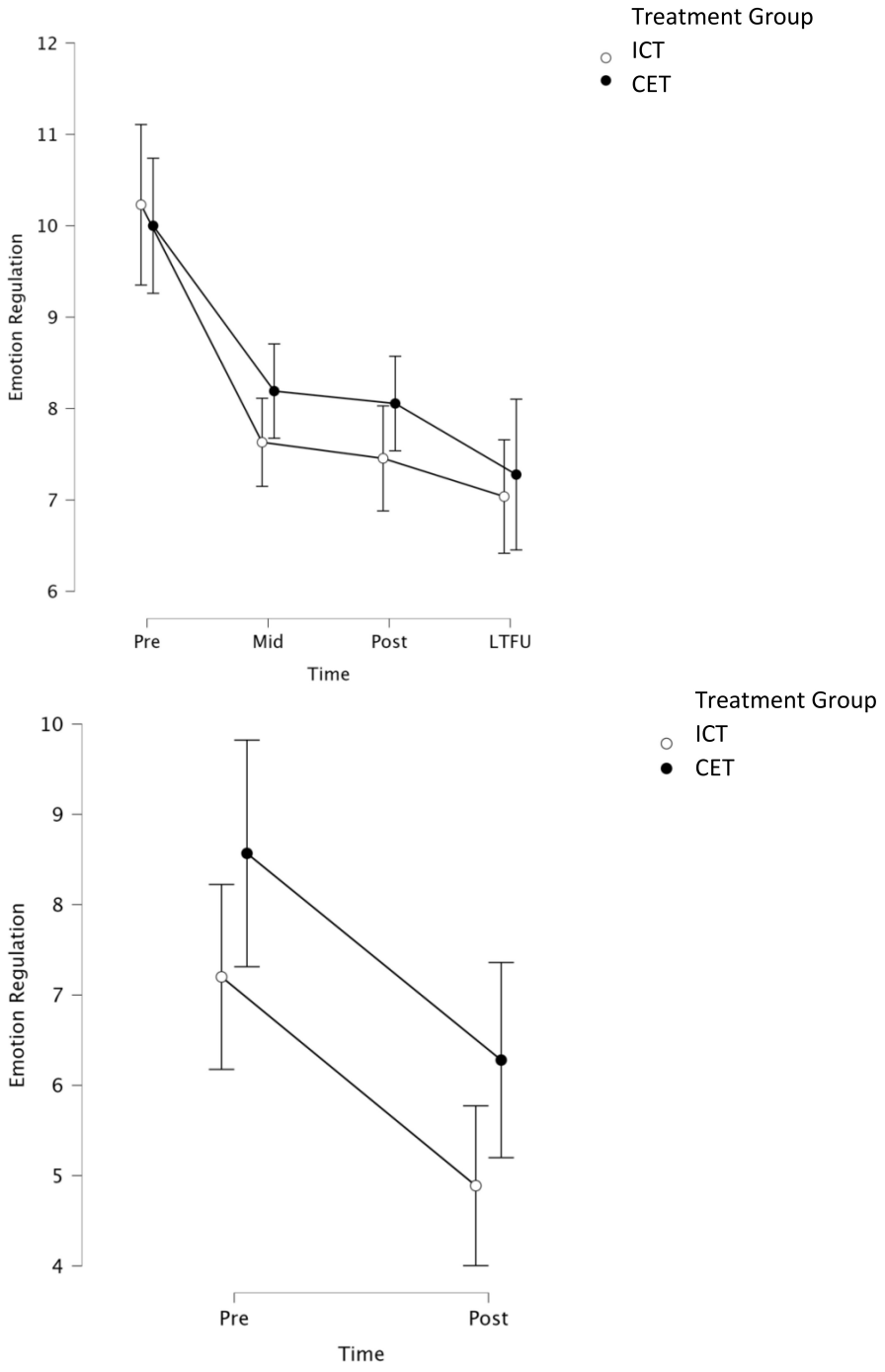


FIGURE 3
Graphical representation of parent-reported **(top)** and teacher-reported **(bottom)** models, with BASC-3 emotional self-control and negative emotionality subscales collapsed within each time point for each informant. Parent ratings were obtained at pre, mid, post, and LTFU. Teacher ratings were obtained at pre and 1–2 months post-treatment. LTFU, long-term follow-up (2–4 months after treatment concluded).

emotional self-control subscale assesses children's skill at regulating their emotions and affect in response to changes in the environment (e.g., "is overly emotional"), and the negative emotionality subscale assesses children's tendency to respond in an overly negative way to routine and novel environmental stimuli (e.g., "finds fault with everything"). These subscales were selected as the primary emotion regulation outcomes given their relative rigor according to an extensive review of all available emotion regulation measures (68). Psychometric support for the

emotional self-control and negative emotionality subscales includes high internal consistency (α = 0.87–0.91) and test–retest reliability [r = 0.86–0.88; (62)]. The emotional self-control subscale contains 12 items, and the negative emotionality subscale contains 9 items for parent report and 8 items for teacher report. All items are rated on a four-point Likert-type scale (*never*, *sometimes*, *often*, *and almost always*). Higher raw scores indicate more difficulties with emotion regulation.

Emotion dysregulation inventory

An additional measure of emotion regulation was added to the study protocol in March 2018 given emerging data linking emotion regulation with the executive functions targeted by our protocols as described above. The Emotion Dysregulation Inventory (EDI) (69) assesses children's emotion regulation based on parent report, and contains 13 items that are rated on a five-point Likert-type scale (e.g., "emotions go from 0 to 100 instantly"). Psychometric support for the EDI includes excellent internal consistency (α = 0.90–0.92), expected relations with other emotion regulation measures, and the ability to discriminate between children with known emotion regulation difficulties and their typically developing counterparts (70). Higher raw scores indicate more difficulties with emotion regulation.

Due to an administrative error, the parent *Emotion Regulation Checklist (ERC)* was administered instead of the EDI at pre-treatment for the first 62 participants. The EDI was administered at all other time points. The ERC (71) emotional lability subscale contains 15 items that are rated on a four-point Likert-type scale (e.g., "exhibits wide mood swings"). Psychometric support for the ERC includes high internal consistency (α = 0.98), discriminant validity relative to distinct constructs such as resilience, expected relations with other metrics of emotion regulation (r = 0.44–0.79), and the ability to differentiate between groups of children at-risk vs. not at-risk for emotional problems (71). Higher raw scores reflect more difficulties with emotion regulation. The ERC and EDI are strongly correlated [r = 0.53–0.64: (70)]; thus, we made the *a priori* decision to retain these participants and use their pre-treatment ERC data in the exploratory analyses. To equate the scaling across the ERC and EDI, we computed the proportion of the maximum possible score for each child for each measure at each time points.

Intellectual functioning and socioeconomic status at pre-treatment

Pre-treatment IQ was estimated using the WISC-V Verbal Comprehension Index (72). Hollingshead (73) SES was estimated based on caregiver(s)' education and occupation at pre-treatment.

Informant expectancy questionnaires

Parent expectancies

Parent treatment-related expectancies were assessed via the NICT Expectations of Cognitive Training scale (74) at mid-treatment. The scale contains seven items that assess the extent to which parents expect cognitive training to improve their child's functioning. Higher mean scores indicate higher expectancies (range = 1–7). The impact of parent expectancies on improvements in parent-reported emotion regulation during treatment was assessed via sensitivity analyses as described below.

Teacher expectancies

Teachers were not directly assessed for expectancies given our goal of obtaining ratings from teachers who were unaware that the children were receiving treatment. Instead, parents reported on the teachers' knowledge of treatment participation on a study-created post-treatment blinding questionnaire. Based on parent report, all teachers remained masked to treatment allocation/group, whereas 37 of 94 (39.4%) teachers were told that the child was participating in an intervention [i.e., masked to treatment allocation but unmasked to study participation, creating the opportunity for expectancy effects; (66)]. The potential impact of teacher expectancies was assessed via sensitivity analyses as described below.

Bayesian analyses

Traditional null hypothesis significance tests (p-values) were supplemented with Bayes Factors as recommended (75). Bayes Factors were added because they allow stronger conclusions by estimating the magnitude of support for both the alternative and null hypotheses (76). BF_{10} is the Bayes Factor (BF) indicating how much more likely the alternative hypothesis (H_1) is relative to the null hypothesis (H_0). Values >3.0 are considered moderate support for the alternative hypothesis (77). BF_{01} is the inverse of BF_{10} (i.e., $BF_{01} = 1/BF_{10}$), and is reported when the evidence favors the null hypothesis (76). BF_{01} is interpreted identically to BF_{10} (>3 = moderate, >10 = strong, >100 = decisive evidence that ICT and CET produce equivalent changes in an outcome). We refer to findings of BF_{10} >3 as significant evidence *for* an effect (i.e., support for the alternative hypothesis of an effect at/above pre-specified evidentiary thresholds), and findings of BF_{01} >3 as significant evidence *against* an effect (i.e., support for the null hypothesis of no effect at/above pre-specified evidentiary thresholds). Both p-values and Bayes Factors are reported. We refer to effects as "marginally significant" when results indicate $p < 0.05$ but $BF_{10} < 3.0$ (i.e., when the effect is supported by null hypothesis testing but the Bayes Factor suggests evidentiary value below our prespecified threshold).

Transparency and openness statement

Best practice guidelines for cognitive training studies were closely followed as detailed in **Table 2**. Trial outcomes and detailed data analytic plans for the CET vs. ICT randomized controlled trial were preregistered at https://osf.io/abwms. The analytic plan detailed in the preregistration was followed for the present study. All emotion regulation measures included in the study battery were analyzed and are reported here. Primary outcomes (effects on working memory, inhibitory control, and ADHD symptoms) and academic outcomes are reported in Kofler et al. (11) and Singh et al. (59), respectively, for subsets of the current sample. The deidentified raw data (.jasp) and results output (including analysis scripts and test statistics) are available for peer review as recommended (75): [https://osf.io/86fwu/]. We report how we determined our sample size, all data exclusions (if any), all manipulations, and all measures in the study.

Data analysis overview

Data analyses were conducted with default priors using JASP version 0.14.1 (78). Our analytic plan included a 2 (between-subjects factor Treatment Group: CET vs. ICT) × 2 (within-subject factor Subscale: emotional self-control, negative emotionality) × 3 (within-subject factor Time: pre-, mid-, post-treatment) repeated measures ANOVA to examine treatment-related changes in emotion regulation based on parent report, with *post-hocs* following significant interactions and *a priori* planned contrasts to characterize the pattern of change over time separately for each treatment group. Similarly, teacher data were analyzed using a 2 (between-subjects factor Treatment Group: CET vs. ICT) × 2 (within-subject factor Subscale: emotional self-control, negative emotionality) × 2 (within-subject Time: pre-, post-treatment) repeated measures/mixed model ANOVA with the same *post hoc*/planned contrast plan.

Results

Power analysis

Power analysis using G*Power 3.1 (79) indicated that our sample size ($N = 94$), with $\alpha = 0.05$, $\beta = 0.80$, and 3 time points (pre, mid, post), is powered to detect main effects of at least $d = 0.29$ and treatment group × time interaction effects of at least $d = 0.34$. Effects of these magnitudes were considered reasonable given evidence that (a) experimentally manipulating demands on working memory [$d = 0.95$; (10)] and inhibitory control [$d = 0.52$; (41)] both produce changes in emotion regulation that are at least moderate in magnitude; (b) CET produces large improvements in working memory [$d = 0.96$–1.20; (11, 34)]; and (c) ICT produces large improvements in stop-signal inhibitory control [$d = 1.12$; (11)]. Thus, the study is sufficiently powered to address its primary aims.

Study retention, outliers, and missing data handling

Study retention was high for both CET (89% completers) and ICT (78%); completion rates did not differ based on treatment allocation ($p = 0.33$). The treatment groups also did not differ on missing data rates ($p = 0.48$–0.73); complete data were available for 80.5% of post parent, 70.1% of post teacher, and 66.2% of follow-up parent ratings. Missing data were determined to be missing completely at random (Little's MCAR test: $p > 0.99$) and were imputed using expectation maximization based on all available data. This maximum likelihood-based approach has been shown to produce unbiased results for missingness rates at/above the current levels when data are missing at random (80), as was the case in the current study. Finally, all independent and dependent variables were screened for univariate outliers, defined as values greater than 3 *SD* outside the within-group mean. Outliers were corrected to the most extreme value within 3 *SD* of the mean; this process affected 2.3% of data points.

Pre-treatment characteristics

Children randomized to ICT ($n = 50$) vs. CET ($n = 44$) did not differ from each other in parent- or teacher-rated emotion regulation or any of the pre-treatment characteristics shown in **Table 1** (all $BF_{10} \leq 0.70$, $p > 0.05$). Additionally, the treatment groups did not differ regarding comorbid diagnoses, training duration, or proportio nof children prescribed psychostimulants.

Primary results

Tier 1: Parent-reported emotion regulation at immediate post-treatment

Behavior assessment scale for children-3 model

Consistent with our hypotheses, the 2 (between-subjects factor Treatment: CET, ICT) × 2 (within-subject factor Subscale: negative emotionality, emotional self-control) × 3 (within-subject factor Time: pre-, mid-, and post-treatment) repeated measures/mixed model ANOVA for parent-reported emotion regulation was significant for main effects of Time ($BF_{10} = 8.04 \times 10^{13}$, $p < 0.001$; $\eta^2_p = 0.28$, $d = 1.25$) and Subscale ($BF_{10} = 3.50 \times 10^{18}$, $p < 0.001$; $\eta^2_p = 0.44$, $d = 1.77$), and for the Time × Subscale interaction ($BF_{10} = 3.00 \times 10^{35}$, $p = 0.002$; $\eta^2_p = 0.06$, $d = 0.51$; **Figure 3**). Contrary to our expectations, there was significant evidence *against* the

Treatment × Time interaction (BF_{01} = 9.80, p = 0.32; $\eta^2_p = 0.01$, $d = 0.20$), indicating that the CET and ICT groups showed equivalent reductions in parent-reported emotion regulation. Similarly, there was no significant main effect of Treatment or a Treatment × Subscale × Time interaction (all $BF_{10} < 1$, $p \geq 0.70$). *A priori* planned contrasts indicated that both the CET and ICT groups demonstrated reductions in emotion dysregulation across both subscales from pre- to post-treatment (all $BF_{10} \geq 6.00$, all $p \leq 0.002$; $d_{ICT} = 0.55$ to 0.57; $d_{CET} = 0.42$ to 0.48). We also repeated the primary model using proportion of the total possible score for each subscale to account for the subscales containing different numbers of items. When using proportions, both the Subscale and Subscale × Time effects were no longer significant, suggesting that differences in subscales were due to scaling issues rather than differences in emotion regulation subcomponents. This interpretation is consistent with the planned contrasts, which indicated that both groups demonstrated improvement across both subscales from pre- to post-treatment.

Emotion dysregulation inventory model

Next, we examined the extent to which our primary results replicated using a narrowband measure of emotion regulation as described above. Results were consistent with the primary model, including a significant main effect of Time ($BF_{10} = 3.95 \times 10^{41}$, $p < 0.001$; η^2_p 0.62, $d = 1.28$) and evidence against a Treatment × Time interaction ($BF_{01} = 17.54$, $p = 0.70$; $\eta^2_p = 0.004$, $d = 0.13$).

Tier 2: Teacher-reported emotion regulation at 1–2 months post-treatment

Consistent with the parent-report model, the 2 (between-subjects factor Treatment: CET, ICT) × 2 (within-subject factor Subscale: negative emotionality, emotional self-control) × 2 (within subject factor Time: pre- and post-treatment) repeated measures/mixed model ANOVA was significant for the main effect of Time ($BF_{10} = 1.22 \times 10^6$, $p < 0.001$, $\eta^2_p = 0.19$, $d = 0.99$). Similar to the parent-report model, there was evidence against a Treatment × Time interaction ($BF_{01} = 5.08$, $p = 0.98$, $\eta^2_p = 0.000006$, $d = 0.005$), suggesting that children in both treatment conditions showed equivalent reductions in teacher-reported emotion regulation. There was a significant main effect of Subscale ($BF_{10} = 4.35 \times 10^{17}$, $p < 0.001$; $\eta^2_p = 0.61$, $d = 2.50$) and a Time × Subscale interaction ($BF_{10} = 5.03$, $p < 0.001$; $\eta^2_p = 0.17$, $d = 0.91$). However, in contrast to the parent model, both the Subscale and Time × Subscale effects remained significant when accounting for the different number of items per subscale using the proportion of total possible scores as described above, suggesting that patterns of improvement across the interventions were different for the distinct emotion regulation subcomponents. Indeed, *a priori* planned contrasts indicated that both treatment groups improved on the teacher-reported emotional self-control subscale from pre- to post-treatment (all $BF_{10} > 10$; all $p \leq 0.006$; $d_{ICT} = 0.68$; $d_{CET} = 0.35$), whereas neither group exhibited significant changes in negative emotionality ($BF_{10} \leq 2$; $p \geq 0.99$).

Tier 3: Parent-reported emotion regulation at 2–4 months post-treatment

Additional analyses were conducted to probe for maintenance of effects based on parent report. These analyses involved repeating the parent pre/mid/post-treatment model above, this time adding follow-up as a fourth time point. Of primary interest were planned contrasts assessing (a) whether emotion dysregulation remained significantly below pre-treatment levels at follow-up (pre vs. follow-up), and (b) whether post-treatment gains were lost across the no-contact follow-up duration (post vs. follow-up). Reporting is truncated for readability. Results were consistent with the primary parent-report model reported above, including a main effect of Time ($BF_{10} = 1.15 \times 10^{14}$, $p < 0.001$; $\eta^2_p = 0.27$, $d = 1.22$) and evidence against a Treatment × Time interaction ($BF_{01} = 45.45$, $p = 0.52$; $\eta^2_p = 0.008$, $d = 0.18$). Pre-planned contrasts indicated that parent-rated emotion dysregulation across both subscales remained significantly reduced at follow-up relative to pre-treatment for both treatment groups (all $BF_{10} > 10$, all $p \leq 0.01$; d_{ICT}: emotional self-control = 0.66, negative emotionality = 0.60; d_{CET}: emotional self-control = 0.67, negative emotionality = 0.50). Similarly, neither group demonstrated a significant loss in parent-rated emotional self-control or negative emotionality from post-treatment to follow-up, suggesting that the pre/post gains in emotion regulation were maintained at 2–4 months follow-up for both groups (all $BF_{10} < 3$, all $p > 0.99$).

Tier 4 sensitivity analyses: Expectancy effects, medication status, medication changes, maturation, and COVID protocol changes

Finally, we performed a series of exploratory analyses to examine the extent to which the significant reductions in emotion dysregulation during treatment may reflect an artifact of non-treatment processes (**Table 3**). The pattern, significance, magnitude, and interpretation of all results were unchanged when pre-treatment age, parent expectancies, teacher masking, medication status and changes, time between treatment completion and informants completing the post-treatment ratings, or COVID protocol status were added as covariates to the primary models. In all cases, the main effect of Time remained significant (all $BF_{10} > 10$, all $p < 0.01$; $d = 0.50$ to 1.15), suggesting that the improvements observed during treatment are unlikely to be due to these non-treatment factors. In no case did adding covariates result in a significant Treatment × Time interaction (all $BF_{01} > 3$; all $p \geq 0.52$). Finally, none of the covariates showed significant main effects of Time or interacted with Treatment, Time, or Subscale, with

one exception. In the teacher model, there was a significant main effect of age ($BF_{10} = 2.96$, $p = 0.02$; $\eta^2_p = 0.06$) and an age × Subscale interaction ($BF_{10} = 1.55 \times 10^{18}$, $p < 0.001$; $\eta^2_p = 0.17$), such that younger children exhibited greater improvements in teacher-reported emotional self-control (but not negative emotionality).

Discussion

The current study was the first randomized controlled trial to compare the effects of training central executive working memory vs. inhibitory control on emotion dysregulation for children with ADHD. We hypothesized that both treatment groups would demonstrate reductions in emotion dysregulation during treatment, and that CET would be superior to ICT. Results indicated that both treatment groups exhibited moderate improvements in emotional control from pre- to post-treatment per both parent and teacher report, and both groups experienced moderate reductions in negative emotionality according to parent, but not teacher, report. Contrary to our expectations, reductions in emotion dysregulation from pre- to- post treatment were equivalent across the CET and ICT groups, as demonstrated by consistent evidence against a treatment × time interaction across all tested models. The use of CET and ICT as active, credible controls for one another is a significant improvement on previous trials that are unable to account for expectancy/placebo effects (66) in cognitive training interventions, most of which have proven unsuccessful in ameliorating ADHD symptoms and impairments in well-controlled clinical trials (49). However, the lack of significant treatment group × time interactions precludes us from confidently attributing improvements in emotion regulation to active treatment components of CET and ICT despite sensitivity analyses suggesting that the improvements during treatment were robust to all assessed threats to validity.

The significant reductions in parent- and teacher-reported emotion dysregulation that occurred during treatment were robust to control for most extraneous variables. Our findings were consistent with extant studies documenting improvements in emotion regulation following working memory training in neurotypical and clinical samples that did not include ADHD (42–46, 48). Additionally, the current study contributes to a small, mixed literature, in which some studies find that training inhibitory control improves emotion regulation for neurotypical children and adults with emotion regulation difficulties (41, 48) but not for neurotypical adults (47). There is little research on emotion regulation outcomes when training executive functions in children with ADHD, but our findings conflict with Tamm et al. (50) and Tamm et al. (51) who found no impact of a play-based metacognitive attention training on emotion regulation. However, this discrepancy is unsurprising given the difference in training targets relative to the current study. That is, directly training a specific executive function provides a more potent treatment dose than attempting to target executive functioning more broadly (49), and CET and ICT target specific cognitive functions that are linked with ADHD-related emotion dysregulation (7–10). Thus, it appears possible that both working memory and inhibitory control may be causally linked with emotion regulation difficulties in ADHD. At the same time, clinical implications should be considered tentative because we did not include an untreated control group to conclusively rule out spontaneous recovery as an alternative explanation for the significant improvements associated with both active treatments.

Although the current study cannot conclusively rule out maturation/spontaneous recovery as an explanation for the significant improvements in emotion regulation that occurred during treatment for both groups, this explanation appears unlikely based on extant literature and when contextualized with other results from the RCT. For example, children often experience *more* difficulties with emotion regulation between the ages of 9 and 13 than they do in early childhood (81–83). In other words, maturation effects, if present, might be expected to produce *increases* in emotion regulation difficulties rather than the decreases observed in the current study. Thus, it seems unlikely that we would expect to see spontaneous improvements in emotion regulation over this time period in the absence of targeted intervention. Notably, however, to our knowledge, no study to date has reported on the spontaneous development of children's emotion regulation over the relatively short duration covered by our active treatment phase (i.e., 10 weeks).

Similarly, if the observed improvements in emotion regulation were artifacts of maturation, spontaneous recovery, or other factors unrelated to the tested interventions, it seems unlikely that we would have obtained the specific patterns of improvement that were observed across informants and time points. In particular, it seems reasonable to conclude that the pattern of significant pre/post improvement followed by the non-significant post/follow-up change makes these alternative explanations unlikely. Stated differently, it would seem to be a logical stretch to argue that children experienced an acute episode of maturation/spontaneous recovery that, coincidentally, temporally coincided with the active treatment component and then, coincidentally, abruptly stopped when treatment was completed. Similarly, the maintenance of gains at follow-up, combined with our sensitivity analyses, also appears to effectively rule out expectancy effects as an alternative explanation for the observed improvements. That is, our understanding is that expectancy effects are time limited rather than producing lasting change (66), in which case an expectancy hypothesis would not be able to account for the lasting improvements observed in the current study.

Alternatively, it is possible that the improvements observed during treatment were attributable to non-specific or shared components of the treatments rather than the treatment

targets specifically. For example, participants in both conditions participated in comprehensive psychoeducational evaluations, which included feedback sessions that provided psychoeducation about ADHD and associated difficulties as needed. However, psychoeducation alone has failed to improve ADHD-related emotion dysregulation and higher doses of psychoeducation may be iatrogenic for ADHD-related treatment outcomes (84–86). Additionally, both CET and ICT included routine contact with the study team. Extant literature documents the non-specific benefits of supportive clinician contact (87), which has also demonstrated incremental value in trials of internet-based psychosocial interventions (88, 89). However, it is unlikely that improvements are solely attributable to clinician contact given that significant treatment × time interaction effects have been found for most other studied outcomes from this trial, including ADHD symptoms, academic outcomes, and organizational skills (11, 59, 90). In essence, there does not appear to be a compelling argument that non-specific clinician contact would specifically impact some but not most studied outcomes—especially given that teachers also reported improvements despite no contact from the study team.

Beyond the potential but tentative clinical intervention implications, results of the current study add to research documenting cross-sectional links between inhibitory control and ADHD-related emotion regulation (9, 40), and extend these findings by suggesting that these relations may be causal. However, the equivalent reduction in emotion dysregulation for both treatment groups is somewhat inconsistent with cross-sectional evidence that working memory but not inhibition uniquely predicts ADHD-related emotion dysregulation when included in the same model (8). Future work specifically examining the extent to which (a) improvements in working memory or inhibitory control covary with improvements in emotion regulation; and (b) these performance improvements are reflected at the cortical level will be important for furthering our understanding of the role of these executive functions in children's emotion regulation skills.

Limitations

The present study demonstrates several strengths, including a carefully characterized sample of children with ADHD with and without comorbidities, outcome ratings from multiple informants masked to treatment allocation, and intervention groups that served as active, credible controls for one another (66). However, some limitations warrant consideration when interpreting results. First, ICT was developed as an ideal active, credible control for CET in consideration of expected effects on the trial's primary clinical outcomes (ADHD symptoms) given experimental evidence implicating working memory (37, 91), but not inhibitory control (92), as a causal mechanism underlying core ADHD symptoms. However, given experimental evidence for functional, if not causal, roles for both working memory and inhibitory control on emotion regulation (10, 41), it will be necessary for future trials to include a third treatment arm that targets processes(es) unrelated to children's emotion regulation skills. Given that previous studies suggest that executive functions, particularly working memory, exert direct effects on emotion regulation as well as indirect effects via ADHD inattentive and hyperactive/impulsive symptoms, future trials should consider the extent to which improvements in emotion regulation represent a direct outcome of executive function training vs. a downstream outcome of improved ADHD symptoms.

Additionally, the clinical diversity of the sample was useful given that comorbidity is the rule rather than the exception in individuals with ADHD [e.g., (93)], but the inclusion of comorbidities may limit the specificity of these findings regarding children with only ADHD. Unexpectedly, children with ADHD whose families self-selected out of the treatment phase had moderately higher emotion regulation difficulties based on teacher but not parent report. Despite finding medium to large improvements in emotion regulation for treated children, it is possible that larger effects would have been detected if more severely dysregulated children were retained in the trial and/or if we recruited specifically for children with emotion regulation difficulties. Finally, most participants in the current trial identified as White/non-Hispanic. Future studies should recruit samples with larger proportions of historically excluded racial/ethnic groups to ensure that results generalize to these groups.

Clinical and research implications

Taken together, results of this double-blind randomized controlled trial were consistent in documenting significant improvements in emotion regulation for children with ADHD that persist at least 2–4 months after treatment termination and are not likely artifacts of any assessed threats to validity. If results of the current study are consistent with future studies including an additional control treatment that targets a mechanism that would not be expected to affect emotion regulation, it would appear likely that working memory and inhibitory control are potentially functionally linked with emotion regulation difficulties in ADHD, consistent with prior experimental evidence in ADHD and non-ADHD samples (10, 41). At the same time, implications for clinical practice should be considered tentative because we did not include an additional control treatment that targets a mechanism that would not be expected to affect emotion regulation. The incremental value of adding CET and/or ICT to extant evidence-based treatments for emotion dysregulation should be examined in future work, as the combination of improving the underlying neurocognitive foundation and directly training emotion regulation skills may prove more beneficial for improving functioning for children

with ADHD than treating emotion dysregulation in isolation (54, 55).

Data availability statement

The datasets presented in this study can be found in online repositories. The names of the repository/repositories and accession number(s) can be found below: Open Science Framework (OSF) https://osf.io/86fwu/.

Ethics statement

The studies involving human participants were reviewed and approved by the Florida State University Human Subjects Committee. Written informed consent to participate in this study was provided by the participants' legal guardian/next of kin. Written informed consent was obtained from the minor(s)' legal guardian/next of kin for the publication of any potentially identifiable images or data included in this article.

Author contributions

NG was the lead investigator for analysis and interpretation of the data and drafting of the work. MK was the principal investigator for funding acquisition and had a supporting role in revision of the manuscript. All authors made substantial contributions to the conception of the work, revision of the manuscript, and the acquisition of data for the work, and provided approval for publication of the content.

Funding

This work was supported in part by the NIH grant (R01 MH115048, PI: MK). The sponsor had no role in the design and conduct of the study; collection, management, analysis, and interpretation of the data; or preparation, review, or approval of the manuscript.

Conflict of interest

The principal investigator MK/Florida State University (FSU) was awarded U.S. Patent 11,210,967 for the Neurocognitive Interventions described in the present study. Central Executive Training was recently licensed to Sky Therapeutics, where MK is in negotiations to serve as Chief Science Officer and consultant.

The remaining authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

Publisher's note

References

1. American Psychiatric Association. *Diagnostic and statistical manual of mental disorders*. 5th ed. Washington, DC: American Psychiatric Association (2013). doi: 10.1176/appi.books.9780890425596

2. Polanczyk GV, Willcutt EG, Salum GA, Kieling C, Rohde LA. ADHD prevalence estimates across three decades: an updated systematic review and meta-regression analysis. *Int J Epidemiol.* (2014) 43:434–42. doi: 10.1093/ije/dyt261

3. Graziano PA, Garcia A. ADHD and children's emotion dysregulation: a meta-analysis. *Clin Psychol Rev.* (2016) 46:106–23. doi: 10.1016/j.cpr.2016.04.011

4. Evans SW, Owens JS, Wymbs BT, Ray AR. Evidence-based psychosocial treatments for children and adolescents with attention deficit/hyperactivity disorder. *J Clin Child Adolesc Psychol.* (2018) 47:157–98. doi: 10.1080/15374416.2017.1390757

5. Galanter CA, Carlson GA, Jensen PS, Greenhill LL, Davies M, Li W, et al. Response to methylphenidate in children with attention deficit hyperactivity disorder and manic symptoms in the multimodal treatment study of children with attention deficit hyperactivity disorder titration trial. *J Child Adolesc Psychopharmacol.* (2003) 13:123–36. doi: 10.1089/104454603322163844

6. Waxmonsky J, Pelham WE, Gnagy E, Cummings MR, O'Connor B, Robb JA. The efficacy and tolerability of methylphenidate and behavior modification in children with ADHD and severe mood dysregulation. *J Child Adolesc Psychopharmacol.* (2008) 18:573–88. doi: 10.1089/cap.2008.065

7. Groves NB, Kofler MJ, Wells EL, Day TN, Chan ES. An examination of relations among working memory, ADHD symptoms, and emotion regulation. *J Abnorm Child Psychol.* (2020) 48:525–37. doi: 10.1007/s10802-019-00612-8

8. Groves NB, Wells EL, Soto EF, Marsh CL, Jaisle EM, Harvey TK, et al. Executive functioning and emotion regulation in children with and without ADHD. *Res Child Adolesc Psychopathol.* (2021) 50:721–35. doi: 10.1007/s10802-021-00883-0

9. Sjöwall D, Roth L, Lindqvist S, Thorell LB. Multiple deficits in ADHD: executive dysfunction, delay aversion, reaction time variability, and emotional deficits. *J Child Psychol Psychiatry.* (2013) 54:619–27. doi: 10.1111/jcpp.12006

10. Tarle SJ, Alderson RM, Arrington EF, Roberts DK. Emotion regulation and children with attention-deficit/hyperactivity disorder: the effect of varying phonological working memory demands. *J Attent Disord.* (2021) 25:851–64. doi: 10.1177/1087054719864636

11. Kofler MJ, Wells EL, Singh LJ, Soto EF, Irwin LN, Groves NB, et al. A randomized controlled trial of central executive training (CET) versus inhibitory control training (ICT) for ADHD. *J Consult Clin Psychol.* (2020) 88:738–56. doi: 10.1037/ccp0000550

12. Bunford N, Evans SW, Wymbs F. ADHD and emotion dysregulation among children and adolescents. *Clin Child Fam Psychol Rev.* (2015) 18:185–217. doi: 10.1007/s10567-015-0187-5

13. Zelkowitz RL, Cole DA. Measures of emotion reactivity and emotion regulation: convergent and discriminant validity. *Pers Individ Dif.* (2016) 102:123–32. doi: 10.1016/j.paid.2016.06.045

14. Zhao X, Page TF, Altszuler AR, Pelham WE, Kipp H, Gnagy EM, et al. Family burden of raising a child with ADHD. *J Abnorm Child Psychol.* (2019) 47:1327–38.

15. Bunford N, Evans SW, Langberg JM. Emotion dysregulation is associated with social impairment among young adolescents with ADHD. *J Attent Disord.* (2018) 22:66–82. doi: 10.1177/1087054714527793

16. Classi P, Milton D, Ward S, Sarsour K, Johnston J. Social and emotional difficulties in children with ADHD and the impact on school attendance and healthcare utilization. *Child Adolesc Psychiatry Ment Health.* (2012) 6:1–8. doi: 10.1186/1753-2000-6-33

17. Qian Y, Chang W, He X, Yang L, Liu L, Ma Q, et al. Emotional dysregulation of ADHD in childhood predicts poor early-adulthood outcomes: a prospective follow up study. *Res Dev Disabil.* (2016) 59:428–36. doi: 10.1016/j.ridd.2016.09.022

18. Walerius DM, Fogleman N, Rosen P. The role of ADHD and negative emotional lability in predicting changes in parenting daily hassles. *J Child Fam Stud.* (2016) 25:2279–91. doi: 10.1007/s10826-016-0381-1

19. Bunford N, Brandt NE, Golden C, Dykstra JB, Suhr JA, Owens JS. ADHD symptoms mediate the association between deficits in executive functioning and social impairment in children. *J Abnorm Child Psychol.* (2014) 43:133–47. doi: 10.1007/s10802-014-9902-9

20. Christiansen H, Hirsch O, Albrecht B, Chavanon ML. Attention-deficit/hyperactivity disorder (ADHD) and emotion regulation over the life span. *Curr Psychiatry Rep.* (2019) 21:1–11. doi: 10.1007/s11920-019-1003-6

21. Hirsch O, Chavanon M, Riechmann E, Christiansen H. Emotional dysregulation is a primary symptom in adult attention-deficit/hyperactivity disorder (ADHD). *J Affect Disord.* (2018) 232:41–7. doi: 10.1016/j.jad.2018.02.007

22. Richard-Lepouriel H, Etain B, Hasler R, Bellivier F, Gard S, Kahn J, et al. Similarities between emotional dysregulation in adults suffering from ADHD and bipolar patients. *J Affect Disord.* (2016) 198:230–6. doi: 10.1016/j.jad.2016.03.047

23. Steinberg E, Drabick D. A developmental psychopathology perspective on ADHD and comorbid conditions: the role of emotion regulation. *Child Psychiatry Hum Dev.* (2015) 46:951–66. doi: 10.1007/s10578-015-0534-2

24. Shaw P, Stringaris A, Nigg J, Leibenluft E. Emotion dysregulation in attention deficit hyperactivity disorder. *Am J Psychiatry.* (2014) 171:276–93. doi: 10.1176/appi.ajp.2013.13070966

25. Hinshaw SP. Attention deficit hyperactivity disorder (ADHD): controversy, developmental mechanisms, and multiple levels of analysis. *Annu Rev Clin Psychol.* (2018) 14:291–316. doi: 10.1146/annurev-clinpsy-050817-084917

26. Kofler MJ, Irwin LN, Soto EF, Groves NB, Harmon SL, Sarver DE. Executive functioning heterogeneity in pediatric ADHD. *J Abnorm Child Psychol.* (2018) 47:273–86.

27. Luo Y, Weibman D, Halperin JM, Li X. A review of heterogeneity in attention deficit/hyperactivity disorder (ADHD). *Front Hum Neurosci.* (2019) 13:42. doi: 10.3389/fnhum.2019.00042

28. Nigg JT, Willcutt EG, Doyle AE, Sonuga-Barke EJ. Causal heterogeneity in ADHD: do we need neuropsychologically impaired subtypes? *Biol Psychiatry.* (2005) 57:1224–30. doi: 10.1016/j.biopsych.2004.08.025

29. Baddeley A. *Working memory, thought, and action.* Oxford: Oxford Press (2007). doi: 10.1093/acprof:oso/9780198528012.001.0001

30. Fosco WD, Kofler MJ, Groves NB, Chan ES, Raiker JS. Which 'working' components of working memory aren't working in youth with ADHD? *J Abnorm Child Psychol.* (2019) 48:647–60. doi: 10.1007/s10802-020-00621-y

31. Karalunas SL, Gustafsson HC, Dieckmann NF, Tipsord J, Mitchell SH, Nigg JT. Heterogeneity in development of aspects of working memory predicts longitudinal attention deficit hyperactivity disorder symptom change. *J Abnorm Psychol.* (2017) 126:774–92. doi: 10.1037/abn0000292

32. Kasper LJ, Alderson RM, Hudec KL. Moderators of working memory deficits in children with ADHD: a meta-analytic review. *Clin Psychol Rev.* (2012) 32:605–17. doi: 10.1016/j.cpr.2012.07.001

33. Verbruggen F, Logan GD. Response inhibition in the stop-signal paradigm. *Trends Cogn Sci.* (2008) 12:418–24. doi: 10.1016/j.tics.2008.07.005

34. Kofler MJ, Sarver DE, Austin KE, Schaefer HS, Holland E, Aduen PA, et al. Can working memory training work for ADHD? Development of central executive training and comparison with behavioral parent training. *J Consult Clin Psychol.* (2018) 86:964–75. doi: 10.1037/ccp0000308

35. Sonuga-Barke E, Bitsakou P, Thompson M. Beyond the dual pathway model: evidence for the dissociation of timing, inhibitory, and delay-related impairments in attention-deficit/hyperactivity disorder. *J Am Acad Child Adolesc Psychiatry.* (2010) 49:345–55. doi: 10.1016/j.jaac.2009.12.018

36. Barkley RA. Behavioral inhibition, sustained attention, and executive functions: constructing a unifying theory of ADHD. *Psychol Bull.* (1997) 121:65–94. doi: 10.1037/0033-2909.121.1.65

37. Rapport MD, Bolden J, Kofler MJ, Sarver DE, Raiker JS, Alderson RM. Hyperactivity in boys with ADHD: a ubiquitous core symptom or manifestation of working memory deficits? *J Abnorm Child Psychol.* (2009) 37:521–34. doi: 10.1007/s10802-008-9287-8

38. McQuade JD, Breaux RP. Are elevations in ADHD symptoms associated with physiological reactivity and emotion dysregulation in children? *J Abnorm Child Psychol.* (2017) 45:1091–103. doi: 10.1007/s10802-016-0227-8

39. Gross JJ. The emerging field of emotion regulation: an integrative review. *Rev Gen Psychol.* (1998) 2:271–99.

40. Tenenbaum RB, Musser ED, Morris S, Ward AR, Raiker JS, Coles EK, et al. Response inhibition, response execution, and emotion regulation among children with attention-deficit/hyperactivity disorder. *J Abnorm Child Psychol.* (2019) 47:589–603.

41. Peckham AD, Johnson SL. Cognitive control training for emotion-related impulsivity. *Behav Res Ther.* (2018) 105:17–26. doi: 10.1016/j.brat.2018.03.009

42. Schweizer S, Hampshire A, Dalgleish T. Extending brain-training to the affective domain: increasing cognitive and affective executive control through emotional working memory training. *PLoS One.* (2011) 6:e24372. doi: 10.1371/journal.pone.0024372

43. Schweizer S, Grahn J, Hampshire A, Mobbs D, Dalgleish T. Training the emotional brain: improving affective control through emotional working memory training. *J Neurosci.* (2013) 33:5301–11. doi: 10.1523/JNEUROSCI.2593-12.2013

44. Xiu L, Zhou R, Jiang Y. Working memory training improves emotion regulation ability: evidence from HRV. *Physiol Behav.* (2016) 155:25–9. doi: 10.1016/j.physbeh.2015.12.004

45. Du Toit SA, Kade SA, Danielson CT, Schweizer S, Han J, Torok M, et al. The effect of emotional working memory training on emotional and cognitive outcomes in individuals with elevated social anxiety. *J Affect Disord.* (2020) 261:76–83. doi: 10.1016/j.jad.2019.09.085

46. Schweizer S, Samimi Z, Hasani J, Moradi A, Mirdoraghi F, Khaleghi M. Improving cognitive control in adolescents with PTSD. *Behav Res Ther.* (2017) 93:88–94. doi: 10.1016/j.brat.2017.03.017

47. Beauchamp KG, Kahn LE, Berkman ET. Does inhibitory control training transfer?: Behavioral and neural effects on an untrained emotion regulation task. *Soc Cogn Affect Neurosci.* (2016) 11:1374–82. doi: 10.1093/scan/nsw061

48. Xie J, Liu S, Fang P. Cognitive training improves emotion regulation in Chinese preschool children. *Pediatr Int.* (2021) 63:1303–10. doi: 10.1111/ped.14661

49. Rapport MD, Orban SA, Kofler MJ, Friedman LM. Do programs designed to train working memory, other executive functions, and attention benefit children with ADHD? A meta-analytic review of cognitive, academic, and behavioral outcomes. *Clin Psychol Rev.* (2013) 33:1237–52. doi: 10.1016/j.cpr.2013.08.005

50. Tamm L, Hughes C, Ames L, Pickering J, Silver CH, Emslie G. Attention training for school-aged children with ADHD: results of an open trial. *J Attent Disord.* (2010) 14:86–94.

51. Tamm L, Epstein JN, Peugh JL, Nakonezny PA, Hughes CW. Preliminary data suggesting the efficacy of attention training for school-aged children with ADHD. *Dev Cogn Neurosci.* (2013) 4:16–28. doi: 10.1016/j.dcn.2012.11.004

52. Tamm L, Nakonezny PA, Hughes CW. An open trial of a metacognitive executive function training for young children with ADHD. *J Attent Disord.* (2014) 18:551–9. doi: 10.1177/1087054712445782

53. Dovis S, Van der Oord S, Wiers RW, Prins PJ. Improving executive functioning in children with ADHD: training multiple executive functions within the context of a computer game. A randomized double-blind placebo controlled trial. *PLoS One.* (2015) 10:e0121651. doi: 10.1371/journal.pone.0121651

54. Pauli-Pott U, Mann C, Becker K. (2021). Do cognitive interventions for preschoolers improve executive functions and reduce ADHD and externalizing symptoms? A meta-analysis of randomized controlled trials. *Eur Child Adolesc Psychiatry*. 30:503–21.

55. Chacko A, Kofler M, Jarrett M. Improving outcomes for youth with ADHD: a conceptual framework for combined neurocognitive and skill-based treatment approaches. *Clin Child Fam Psychol Rev.* (2014) 17:368–84. doi: 10.1007/s10567-014-0171-5

56. Cortese S, Ferrin M, Brandeis D, Buitelaar J, Daley D, Dittmann RW, et al. Cognitive training for attention-deficit/hyperactivity disorder: meta-analysis of

clinical and neuropsychological outcomes from randomized controlled trials. *J Am Acad Child Adolesc Psychiatry.* (2015) 54:164–74.

57. Landis TD, Hart KC, Graziano PA. Targeting self-regulation and academic functioning among preschoolers with behavior problems: are there incremental benefits to including cognitive training as part of a classroom curriculum? *Child Neuropsychol.* (2019) 25:688–704. doi: 10.1080/09297049.2018.1526271

58. Prins PJM, Dovis S, Ponsioen A, ten Brink E, van der Oord S. Does computerized working memory training with game elements enhance motivation and training efficacy in children with ADHD? *Cyberpsychol Behav Soc Netw.* (2011) 14:115–22. doi: 10.1089/cyber.2009.0206

59. Singh L, Gaye F, Cole A, Chan E, Kofler MJ. Central executive training for ADHD: effects on academic achievement, productivity, and success in the classroom. *Neuropsychology.* (2022) 36:330–45. doi: 10.1037/neu0000798

60. Kaufman J, Birmaher B, Brent D, Rao UMA, Flynn C, Moreci P, et al. Schedule for affective disorders and schizophrenia for school-age children (K-SADS-PL): initial reliability and validity data. *J Am Acad Child Adolesc Psychiatry.* (1997) 36:980–8. doi: 10.1097/00004583-199707000-00021

61. DuPaul GJ, Power TJ, Anastopoulos AD, Reid R. *ADHD rating scale—5 for children and adolescents: checklists, norms, and clinical interpretation*. New York, NY: Guilford Press (2016). doi: 10.1037/t05638-000

62. Reynolds CR, Kamphaus RW. *Behavior assessment system for children (BASC-3)*. London: Pearson (2015). doi: 10.1002/9781118625392.wbecp447

63. Awh E, Jonides J. Overlapping mechanisms of attention and spatial working memory. *Trends Cogn Sci.* (2001) 5:119–26.

64. Postle BR, Awh E, Jonides J, Smith EE, D'Esposito M. The where and how of attention-based rehearsal in spatial working memory. *Cogn Brain Res.* (2004) 20:194–205.

65. Alderson RM, Rapport MD, Kofler MJ. Attention-deficit/hyperactivity disorder and behavioral inhibition: a meta-analytic review of the stop-signal paradigm. *J Abnorm Child Psychol.* (2007) 35:745–58.

66. Simons DJ, Boot WR, Charness N, Gathercole SE, Chabris CF, Stine-Morrow EA. Do "brain-training" programs work? *Psychol Sci Public Interest.* (2016) 17:103–86. doi: 10.1177/1529100616661983

67. Kane MJ, Engle RW. Working-memory capacity and the control of attention: the contributions of goal neglect, response competition, and task set to Stroop interference. *J Exp Psychol.* (2003) 132:47–70. doi: 10.1037/0096-3445.132.1.47

68. Mazefsky CA, Conner CM, Breitenfeldt K, Leezenbaum N, Chen Q, Bylsma LM, et al. Evidence base update for questionnaires of emotion regulation and reactivity for children and adolescents. *J Clin Child Adolesc Psychol.* (2021) 50:683–707. doi: 10.1080/15374416.2021.1955372

69. Mazefsky C, Day T, Siegel M, White S, Yu L, Pilkonis PA. Development of the emotion dysregulation inventory: a PROMIS® ing method for creating sensitive and unbiased questionnaires for autism spectrum disorder. *J Autism Dev Disord.* (2018) 48:3736–46. doi: 10.1007/s10803-016-2907-1

70. Mazefsky C, Yu L, Pilkonis P. Psychometric properties of the emotion dysregulation inventory in a nationally representative sample of youth. *J Clin Child Adolesc Psychol.* (2020) 50:1–13. doi: 10.1080/15374416.2019.1703710

71. Shields A, Cicchetti D. Emotion regulation among school-age children: the development and validation of a new criterion Q-sort scale. *Dev Psychol.* (1997) 33:906–16. doi: 10.1037//0012-1649.33.6.906

72. Wechsler D. *Wechsler intelligence scale for children*. 5th ed. San Antonio, TX: Pearson (2014).

73. Hollingshead AB. *Four factor index of social status*. New Haven, CT: Yale (1975).

74. Rabipour S, Davidson PS. Do you believe in brain training? A questionnaire about expectations of computerised cognitive training. *Behav Brain Res.* (2015) 295:64–70. doi: 10.1016/j.bbr.2015.01.002

75. Redick TS. Working memory training and interpreting interactions in intelligence interventions. *Intelligence.* (2015) 50:14–20.

76. Rouder JN, Morey RD. Default Bayes factors for model selection in regression. *Multivariate Behav Res.* (2012) 47:877–903.

77. Wagenmakers EJ, Love J, Marsman M, Jamil T, Ly A, Morey RD. Bayesian inference for psychology. Part II: example applications with JASP. *Psychon Bull Rev.* (2018) 25:58–76. doi: 10.3758/s13423-017-1323-7

78. JASP Team,. *JASP (version 0.14.1)*. (2020).

79. Faul F, Erdfelder E, Lang A-G, Buchner A. G*Power 3: a flexible statistical power analysis program for the social, behavioral, and biomedical sciences. *Behav Res Methods.* (2007) 39:175–91. doi: 10.3758/BF03193146

80. Kristman V, Manno M, Côté P. Loss to follow-up in cohort studies: how much is too much? *Eur J Epidemiol.* (2004) 19:751–60.

81. Cole PM. Moving ahead in the study of the development of emotion regulation. *Int J Behav Dev.* (2014) 38:203–7. doi: 10.1177/0165025414522170

82. Riediger M, Klipker K. Emotion regulation in adolescence. In: Gross JJ editor. *Handbook of emotion regulation*. New York, NY: The Guilford Press (2014). p. 187–202.

83. Zimmermann P, Iwanski A. Emotion regulation from early adolescence to emerging adulthood and middle adulthood: age differences, gender differences, and emotion-specific developmental variations. *Int J Behav Dev.* (2014) 38:182–94. doi: 10.1177/0165025413515405

84. Dekkers TJ, Hornstra R, Van der Oord S, Luman M, Hoekstra PJ, Groenman AP, et al. Meta-analysis: which components of parent training work for children with attention-deficit/hyperactivity disorder? *J Am Acad Child Adolesc Psychiatry.* (2022) 61:478–94. doi: 10.1016/j.jaac.2021.06.015

85. Ferrin M, Moreno-Granados JM, Salcedo-Marin MD, Ruiz-Veguilla M, Perez-Ayala V, Taylor E. Evaluation of a psychoeducation programme for parents of children and adolescents with ADHD: immediate and long-term effects using a blind randomized controlled trial. *Eur Child Adolesc Psychiatry.* (2014) 23:637–47. doi: 10.1007/s00787-013-0494-7

86. Hornstra, R, Groenman AP, Van der Oord S, Luman M, Dekkers TJ, van der Veen-Mulders L, et al. Which components of behavioral parent and teacher training work for children with ADHD?–a metaregression analysis on child behavioral outcomes. *Child Adolesc Ment Health.* (2022). doi: 10.1111/camh.12561

87. Cuijpers P, Driessen E, Hollon SD, van Oppen P, Barth J, Andersson G. The efficacy of non-directive supportive therapy for adult depression: a meta-analysis. *Clin Psychol Rev.* (2012) 32:280–91. doi: 10.1016/j.cpr.2012.01.003

88. Melville KM, Casey LM, Kavanagh DJ. Dropout from internet-based treatment for psychological disorders. *Br J Clin Psychol.* (2010) 49:455–71. doi: 10.1348/014466509X472138

89. Palmqvist B, Carlbring P, Andersson G. Internet-delivered treatments with or without therapist input: does the therapist factor have implications for efficacy and cost? *Expert Rev Pharmacoecon Outcomes Res.* (2007) 7:291–7. doi: 10.1586/14737167.7.3.291

90. Chan ESM, Gaye F, Cole AM, Singh LJ, Kofler MJ. Central executive and inhibitory control training effects for ADHD-related organizational impairments. *Poster presentation at the 56th association for behavioral and cognitive therapies (ABCT) conference*. New York, NY: (2022). .

91. Kofler MJ, Rapport MD, Bolden J, Sarver DE, Raiker JS. ADHD and working memory: the impact of central executive deficits and exceeding storage/rehearsal capacity on observed inattentive behavior. *J Abnorm Child Psychol.* (2010) 38:149–61. doi: 10.1007/s10802-009-9357-6

92. Alderson RM, Rapport MD, Kasper LJ, Sarver DE, Kofler MJ. Hyperactivity in boys with attention deficit/hyperactivity disorder (ADHD): the association between deficient behavioral inhibition, attentional processes, and objectively measured activity. *Child Neuropsychol.* (2012) 18:487–505. doi: 10.1080/09297049.2011.631905

93. Reale L, Bartoli B, Cartabia M, Zanetti M, Costantino MA, Canevini MP, et al. Comorbidity prevalence and treatment outcome in children and adolescents with ADHD. *Eur Child Adolesc Psychiatry.* (2017) 26:1443–57.

frontiers | Frontiers in Psychiatry

TYPE Original Research
PUBLISHED 21 December 2022
DOI 10.3389/fpsyt.2022.1038223

OPEN ACCESS

EDITED BY
Ru Ying Cai,
Autism Spectrum Australia, Australia

REVIEWED BY
Christiane E. Kehoe,
The University of Melbourne, Australia
Lauren Rice,
The University of Sydney, Australia
Elizabeth Roof,
Vanderbilt University, United States

*CORRESPONDENCE
Nawelle Famelart
nawelle.famelart@univ-tlse2.fr

†These authors have contributed equally to this work and share last authorship

SPECIALTY SECTION
This article was submitted to Autism,
a section of the journal Frontiers in Psychiatry

RECEIVED 06 September 2022
ACCEPTED 28 November 2022
PUBLISHED 21 December 2022

CITATION
Famelart N, Diene G, Çabal-Berthoumieu S, Glattard M, Molinas C, Tauber M and Guidetti M (2022) What underlies emotion regulation abilities? An innovative programme based on an integrative developmental approach to improve emotional competencies: Promising results in children with Prader–Willi syndrome.
Front. Psychiatry 13:1038223.
doi: 10.3389/fpsyt.2022.1038223

What underlies emotion regulation abilities? An innovative programme based on an integrative developmental approach to improve emotional competencies: Promising results in children with Prader–Willi syndrome

Nawelle Famelart[1]*, Gwenaelle Diene[2], Sophie Çabal-Berthoumieu[2], Mélanie Glattard[2], Catherine Molinas[2], Maithe Tauber[2,3†] and Michèle Guidetti[1†]

[1]Laboratory CLLE, University of Toulouse, CNRS, Toulouse, France, [2]Centre de Référence du Syndrome de Prader-Willi, CHU Toulouse, Toulouse, France, [3]Laboratory CPTP, University of Toulouse, CNRS, INSERM, Toulouse, France

Background: This study aimed to test the effect of a new training programme on emotional competencies, named EMO-T, and to show the value of an integrative developmental approach. This approach postulates that the emotion regulation disturbances commonly observed in neurodevelopmental disorders are the consequence of potential disruptions in the prerequisite emotion skills. This integrative approach is particularly suitable in the case of complex and multidimensional disorders such as Prader–Willi syndrome (PWS), a rare genetic disease.

Methods: We examined the emotion expression, recognition, comprehension, and regulation skills in 25 PWS children aged 5–10 and 50 typically developing children (TD) aged 3–10. After a pre-test session, half of the PWS children participated in the EMO-T programme with their regular therapist for 6 weeks, while the other half continued their usual rehabilitation programme. Two post-test sessions were conducted, one at the end of the programme and one 3 months later.

Results: At pre-test, PWS children displayed a deficit in the four emotional competencies (EC). PWS children who participated in the EMO-T programme showed a significant and sustainable post-test improvement regarding voluntary expression and emotion recognition abilities, such that the level reached was no longer different from the baseline level of TD children. They also tended to improve in their emotion regulation, although they received no specific training in this skill.

Discussion: These results support that emotion regulation abilities require prerequisite emotion skills, which should be more fully considered in current training programmes. Because emotion regulation disorders strongly impact all areas of life, an integrative developmental approach appears crucial especially in the case of neurodevelopmental disorders. Further studies should be conducted to explore this perspective.

KEYWORDS

emotion competencies, developmental model, Prader–Willi syndrome, training programme, integrative approach, children

Introduction

A developmental model of emotion competencies

In the last two decades, there has been growing interest in the study of emotional competencies (EC) for their key role in social adaptation. EC refer to the ability to use emotions daily, including their expression, recognition, comprehension, and regulation (1). Most studies tend to focus on these various aspects of EC independently, without considering what underlies them and how they interact during development. Disturbances in these EC are commonly observed in neurodevelopmental disorders. Notably, emotion regulation disturbances can strongly impact daily life (2). To improve the effectiveness of assessment tools and training programmes, it appears important to adopt an integrative developmental approach, i.e., an approach that tackles all areas of emotional competencies, considering their development timeline, and including the child's various caregivers.

From a developmental perspective, the different EC emerge in a hierarchical manner across childhood and adolescence (3–5). Expression and recognition abilities, which are the pillars on which interpersonal relationships are established, emerge very early during the first months of life (6). They can be considered as the basis of the development of emotion comprehension (7). This skill refers to comprehension of the causes and consequences of emotions according to the context and emerges later, at the end of the first year (8). Through the expression, recognition and comprehension skills, the individual gradually conceives of emotion as a concept (9). The construction of this "emotion theorizing" is closely linked to the emergence of the theory of mind for which a pivotal phase is typically observed from the age of 4. "Emotion theorizing" also appears necessary for the regulation of emotions and, by extension, adaptation in general (7, 8, 10–12). Emotion regulation skills develop more steadily across childhood and adolescence (4). In the early years, emotion is mostly co-regulated by parents or caregivers. Interpreting their child's expressions, parents implement strategies to help regulate them (e.g., trying to distract the child's attention to stop them crying). Over the course of development, a child broadens its repertoire, regulating emotions by increasingly sophisticated strategies. At first, this goes through a co-regulating process, but it also involves learning how to recognize and to emotionally master daily life situations. At the end of the second year, children begin to mobilize some regulation strategies on their own initiative (13–16). They are gradually able to manage their reactions through improved voluntary control of expression, an ability that starts to be particularly efficient from the age of 4 (17–20).

Given this developmental model, emotion regulation abilities seem to be strongly dependent on the expression, recognition, and comprehension skills, which can be considered as prerequisites (9). In this perspective, emotion regulation disorders–the most common reason for complaints by the entourage (2, 21)—might be the consequence of potential disruptions in the prerequisite skills. Thus, there is much to gain by basing EC training programmes on this developmental perspective.

Training programmes of emotion competencies

To date, various intervention programmes have been developed to improve EC in children. For the most part, they were developed to prevent (e.g., school context, at-risk populations) or to remedy (e.g., populations with neurodevelopmental disorders) behavioral and social adjustment problems.

Integrative programmes such as the *Social-Emotional Intervention* (13), the *Preschool PATHS* (22), the *SMILE* (23) or the *RULER* (24) programme showed the value of including parents or teachers in the promotion of emotion abilities in children. The *Emotion-Based Prevention* programme (25), which included school and family, showed an improvement in the emotion knowledge and the regulation abilities in children after training in the identification and comprehension of emotions. The study also revealed that the level of emotion knowledge moderated the effect of the programme on both

regulation abilities and on social skills. These results once again reinforce the developmental model of EC. The main challenge of training programmes is to promote the transfer of skills acquired during the training sessions into all areas of daily life. The integration of most of the child's caregivers (i.e., family, teachers, therapists) into the programme appears to be crucial, as highlighted in Hadwin, Howlin, and Baron–Cohen's programme (26).

Immersive programmes also showed their ability to remediate emotional difficulties. Computer programmes such as *Emotion Trainer* (27), *Mind Reading* (28), or *FaceSay* (29) were specifically developed to train emotion recognition and comprehension abilities in children with Autistic Spectrum Disorders (ASD). The main advantage of the computer format is the possibility of regular use at home. These training programmes are progressive and game-based, which adds an ecological value to the tool. Indeed, the game context (especially board games) seems to be relevant to promote the development of emotion regulation competencies in young children (30).

However, a developmental approach may be lacking in many of these programmes. Several of them focus only on one specific competence, and most of them do not include voluntary expressive skills, which are key for emotion regulation and social adjustment abilities. As a few studies have shown, it is advantageous to consider all the prerequisite skills required to develop a competence. For instance, the programme developed by Begeer et al. (31) aimed to improve social cognition in children with ASD, including therapists and families. The protocol started by a reinforcement of competencies considered as precursors of theory of mind (including imitation, perception, interactions with others). In the following steps, sessions focused on training the comprehension of social situations, the thoughts of others, and lastly the ability to reason from another person's perspective. Results showed a significant improvement in basic skills and the conceptual comprehension of theory of mind. The authors also observed a lack of transfer of the skills acquired into daily life, however, due to the generalization difficulties of individuals with ASD.

As mentioned in the review by Mazefsky et al. (21), a holistic approach to EC is crucial in the case of neurodevelopmental disorders. Emotion regulation difficulties should rather be considered as a symptom because they are part of a broader, multidimensional problem. An integrative programme based on a developmental model appeared particularly suitable for complex neurodevelopmental disorders such as Prader–Willi syndrome.

The case of Prader–Willi syndrome

Prader–Willi syndrome (PWS) is a rare genetic disease related to the loss of expression of paternally-inherited genes on chromosome 15 in the region q11–13. PWS is a complex neurodevelopmental disorder characterized by a significant dysfunction of the endocrine system, leading to neonatal hypotonia, growth retardation, eating disorders, and sleep disturbances (32). The phenotype also comprises learning difficulties and many psychological dysfunctions. People with PWS display a mild or moderate intellectual disability (average IQ of 60–70), memory, executive, and perceptive dysfunctions (33–35). They exhibit a language and a motor delay (36–38). In terms of social abilities, people with PWS show social maladjustment and many behavioral disorders, with several autistic features (39, 40). The literature describes a symptomatology such as tantrums, emotional lability, impulsive behavior, lack of empathy and of emotional regulation, anxiety and difficulties of social adaptation (35, 37, 40, 41), suggesting disturbances in social and emotional competencies. These problems strongly impact daily life, constituting the main reason besides hyperphagia for the family burden (42).

Regarding EC, the literature reports difficulties in the recognition and comprehension of basic emotions. Individuals with PWS make on average 10–20% more errors in identifying and assigning emotions than the typical population, even when matched for developmental age (35, 43). They take very little information into account to judge a situation, with difficulties accessing a global representation, focusing instead on details that are mostly irrelevant (39, 43, 44). The same difficulties are observed in face and voice processing (45, 46). This particularity is likely to compromise their capacity for emotional recognition and thus to place them at a disadvantage in everyday situations. In addition, the emotional expressions of PWS children are particularly poor and equivocal, making it difficult to interpret them (9).

Very few intervention programs aimed at directly improving EC in the PWS population exist, however. Most of the studies focus on hormonal treatments [Ghrelin, Oxytocin (47, 48)] or medical protocols [vagus nerve stimulation—tVNS (49)], showing some beneficial effects on social and emotional skills. Recently, two studies reported promising results on two online intervention programs aimed at developing social skills. The BOSS program (50) was offered to adolescents and young adults with PWS. The sessions were collective and took place by videoconference, 3 times a week for 10 weeks. The results showed an interesting effect of the program on socialization (friendly relations and quality of interactions with peers). The PRETEND (42) program was aimed at 3–5 year-old children with PWS and the play skills intervention work was done through the parents. The online coaching sessions took place weekly for 8 weeks. Again, the results were promising, showing interesting improvements in cognitive and affective play skills for some of the children with PWS.

To date, however, no intervention program specifically addressing EC in children with PWS has been tested, despite the need. Considering the complexity and the multidimensional disorders related to PWS, the extent of the EC difficulties

involved, and their consequences on daily life, the aim of the present study was to test the effect of a training programme on EC, named *EMO-T*. We focused on a relatively narrow age range, which is rare in studies of PWS. The period of school age is typically a pivotal phase in emotional development. During this period, the process of "emotion theorizing" is strongly accentuated in the light of the development of other skill areas [cognitive, socialization, learning, autonomy; (3, 7)]. We postulated that a programme based on an integrative developmental approach would enable children to make significant and lasting progress in EC.

Materials and methods

Population

The study population was composed of 25 children with PWS aged 5;5–10;5 years. The average IQ was 75.7 and the average intellectual developmental age was 5;7 years. The children were divided into two groups, one experimental group (PWS-EG) and one control group (PWS-CG) (see Table 1 for details). The distribution was quasi-randomized. It was carried out in such a way that at any time of inclusion (spanned 18 months), the two groups were comparable in terms of sex ratio, chronological age, and IQ. Group comparison analysis (Student *t*-test) showed that the two groups had equivalent IQ, but showed a slight difference—albeit not significant—in term of age (see Table 1). The parents were not informed which group their child was assigned to. The families socioeconomic status (SES) was measured by the Hollingshead scale (51), which is based on marital status, education level and current occupation. SES levels range between 1 and 5, 1 corresponding to a low SES and 5 to a high one. The families in the PWS sample had a median of 4 (middle-high SES). There was no significant difference between the PWS-EG and PWS-CG groups (see Table 1).

Fifty children with typical development (TD) also participated in the study, divided into two groups (see Table 1). The first group consisted of 25 children matched to PWS children by sex and chronological age (TD-CA group). The second group consisted of 25 other children matched to PWS children by sex and intellectual developmental age (TD-DA group). Group comparison analysis (Student t-test) confirmed that the mean age of the TD-CA group was equivalent to the mean chronological age of the PWS group [$t_{(48)} = 0.072$; *ns*; $d = 0.020$]. The mean age of the TD-DA was similar to the mean developmental age of the PWS group [$t_{(48)} = -0.025$; *ns*; $d = 0.007$]. None of the TD children had any academic or learning delays. The SES levels of families in the TD-DA group were higher (median = 5) than the TD-CA and PWS groups (median = 4) [Kruskal–Wallis test: $\chi^2_{(2,\ N\ =\ 75)} = 7.898$, $p = 0.019$; see Table 1]. However, this difference cannot explain the results obtained in experimental tasks, which varied in the other direction (see Section "Results").

A power analysis was conducted on emotion regulation rate. The estimation of the delta and the standard deviation was based on previous studies with individuals suffering from neurodevelopmental disorders on the one hand, and with typically developing individuals on the other hand. Setting the alpha at 0.05 and power at 0.80 yielded a total sample size of 32 (16 for each group). The recruitment of PWS children was carried out through the PWS French National Reference Centre and the French National Association of PWS. Initially, 29 families were enrolled in the study, corresponding to 12% of the target population. One family stopped its

TABLE 1 Descriptive characteristics for PWS and TD groups.

	All PWS	PWS-EG	PWS-CG	PWS groups equivalence	TD-CA	TD-DA
N	25	13	12	–	25	25
(girls/boys)	(14/11)	(7/6)	(7/5)		(14/11)	(14/11)
CA						
Mean (SD)	7;6 (1;6)	8;0 (1;8)	7;0 (1;1)	$t_{(23)} = -1.754$; *ns*; $d = 0.73$	7;6 (1;5)	5;7 (1;4)
Range	5;5–10;4	5;9–10;4	5;5–8;7	(Student *t*-test)	5;2–10;10	3;0–8;4
IQ						
Mean (SD)	75.7 (17.1)	75.9 (19.6)	75.4 (14.8)	$t_{(23)} = -0.072$; *ns*; $d = 0.03$		
Range	44–103	44–103	50–94	(Student *t*-test)		
DA						
Mean (SD)	5;7 (1;5)	5;11 (1;7)	5;2 (1;1)	$t_{(23)} = -1.273$; *ns*; $d = 0.53$		
Range	3;2–9;2	4;3–9;1	3;2–6;9	(Student *t*-test)		
SES						
Median	4	4	3.5	$W = 60$; *ns*	4	5
(Q1–Q3)	(3–4)	(4–4)	(3–4)	(Wilcoxon test)	(3–5)	(4–5)

Means, ranges, and SDs of chronological age (CA), intellectual developmental age (DA), and full-scale IQ (IQ); Medians, 1st and 3rd quartiles of socioeconomic status (SES); Results of PWS groups equivalence tests. *ns*, not significant; age, [years; months]; PWS-EG, PWS experimental group; PWS-CG, PWS control group; TD-CA, TD group with chronological age matched; TD-DA, TD group with development age matched.

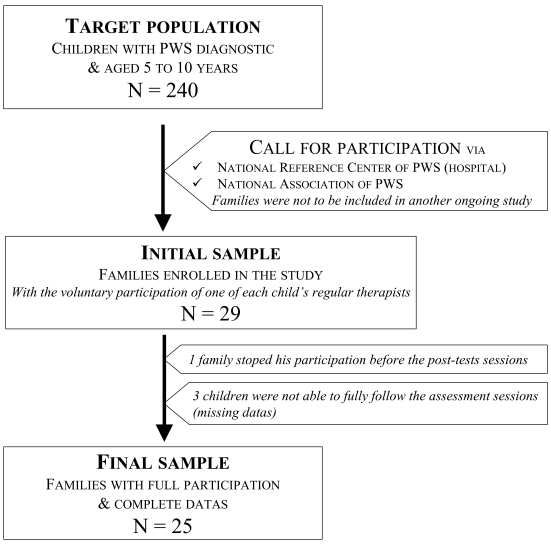


FIGURE 1
Prader–Willi syndrome (PWS) participants flow chart.

participation before the end of the protocol. Three children were not able to fully follow the assessment sessions and were removed from the analysis because of missing data. The participation of PWS families in the EMO-T programme was established in collaboration with one of each child's therapists (speech therapist, psychomotor therapist, or psychologist). The programme was included in the child's regular care programme (see the flowchart in **Figure 1**).

Materials

Given our objectives, the challenge was two-fold. The first task was to assess the children with respect to four emotional competencies: expression, recognition, comprehension, and regulation. The assessment tasks used were created or adapted from previous studies. The second challenge was to develop a training programme consisting of exercises aiming to improve emotional competencies. As the developmental model considers expression, recognition and comprehension as prerequisite skills for emotion regulation, the exercises of the EMO-T programme focused exclusively on these three competencies. The exercises used were created or adapted from previous studies in order to maintain consistency with the assessment tasks, without being identical.

The tasks and exercises are presented briefly below (for more details, see **Supplementary Appendix A**).

Assessment tasks

Expression

The two tasks used were created to assess spontaneous emotional reactions and voluntary production of emotional expressions [from Famelart et al.'s (9) study]. Scores are expressed as a proportion between 0 and 1, 1 meaning that the emotion is fully detectable and matches the theoretical pattern. The *EMOrea Task* consisted in recording the facial reactions of the child while he/she was watching a funny video clip (that was likely to induce the emotion of joy in children). The *EMOmim Task* consisted in asking the child to produce emotional facial expressions (voluntary expressions) of joy, sadness, fear, and anger.

Recognition

Three tasks with different levels of complexity were used to assess the ability to recognize and name the emotions of joy, sadness, fear, and anger. Scores are presented in percentage of correct responses. In the *Identification task*, the participant had to point out the picture of the person expressing the emotion specified in the verbal instruction. In the *Matching task*, the participant had to select the picture of the person expressing the same emotion as in the target picture presented at the top of the screen. In the *Naming task*, the participant had to say which emotion the person was feeling in a short video.

Comprehension

The *AJQ task* (Affective Judgment Questionnaire) was used to assess the ability to attribute an emotion according to the context of stories. The AJQ task consisted in presenting short illustrated stories and asking the child to say which emotion the character felt (emotions targeted: joy, sadness, fear, and anger) and to justify their response. Scores were based on emotion attribution and kind of justifications, and they are presented as the percentage of correct responses.

Regulation

To assess the emotion regulation skills, we used the *Emotion Regulation Checklist* (ERC—French version), which is a questionnaire completed by parents. Scores of the composite scale range between 0 and 4, and the higher the score, the better the regulation abilities.

EMO-T programme

The EMO-T programme was conducted by one of the child's therapists (speech therapist, psychomotor therapist, or psychologist) and was included in the child's regular care programme. The therapist received a half-day training session on the programme proposed by the researchers. The programme was applied weekly over six 30-min sessions. Each session consisted of the same exercises; however, the supports and stimuli evolved so that the task became more complex as the sessions progressed.

Each session included exercises to train the abilities of expression, recognition, and comprehension of emotions. All the exercises were based on the child's justifications and

arguments and the discussion with the therapist. The sessions followed the same sequence of five exercises:

(1) Sorting of static emotional facial expressions–*Recognition*;
(2) Naming of dynamic emotional expressions (vocal and facial) –*Recognition*;
(3) Mimicking emotions and recognizing them–*Expression*;
(4) Emotion attribution–*Comprehension*;
(5) Narration of emotional experience–*Comprehension*.

Experimental design

For each test session, the children were individually interviewed in a quiet room at home. All the children were met by the same experimenter, a researcher trained as a psychologist. The tasks were always presented in the same order: EMOrea, EMOmim, Matching, Identification, Naming, and AJQ. During this phase, the parents completed the ERC questionnaire.

All the children–PWS and TD children–were first assessed in a pre-test session. The PWS children included in the experimental group (PWS-EG) then received the training programme conducted by the previously trained therapist (see above), over 6 weeks (weekly 30-min sessions). During this phase, PWS children from the control group (PWS-CG) continued their usual rehabilitation with their therapist. In the third step, all the PWS children were retested, immediately after the end of the programme (post-test 1) and again 3 months later (post-test 2). Children in the PWS-CG group followed the training programme at the end of the protocol and their therapist was trained only at this time.

Results

Design of analyses

To test the efficiency of the EMO-T programme on the children's EC, analyses were conducted in three steps for each task, considering the PWS Experimental Group as a reference for analyses: (1) comparison of PWS groups with the TD groups at the pre-test session (group effect); (2) comparison of the trajectory evolution of the two PWS groups between the pre- and the two post-test sessions (interaction effect); (3) comparison of the PWS groups at the second post-test session with the TD groups considered as a baseline (from the pre-test session). We used a Generalised Linear Mixed Model (GLMM) to adapt the analytical model to the specificity of the variable distribution and to integrate random effects (repeated measures). Preliminary analyses showed that age, IQ, and sex had no significant interaction or influence on any of the main effect shown.

Analysis results

Figure 2 and **Table 2** summarize the results and statistical analyses for each task. All the details are presented in the **Supplementary Appendix B**.

Regarding the first step of analyses, results from the pre-test session indicated a strong group effect for all tasks, except for the EMOrea task. The scores of the two PWS groups were significantly lower than those of the two TD groups. In addition, analyses showed that the scores of the two PWS groups were statistically similar. However, for the AJQ task, the scores of the PWS-EG group tended to be higher than the scores of the PWS-CG group.

In the second step, results revealed a strong interaction between the PWS groups and the test sessions for the EMOmim task and the three recognition tasks. The scores of the PWS-EG group increased significantly between the pre-test session and the first post-test session, in which they achieved significantly higher scores than the PWS-CG group. The scores of the PWS-EG group between the two post-test sessions remained statistically unchanged in the EMOmim task and the Naming task, while the group showed a significant increase in the Matching and Identification tasks.

No significant interaction was observed for the EMOmim task and the AJQ task. Nevertheless, results from the AJQ task indicated a group effect, with the PWS-EG group mainly displaying higher scores than the PWS-CG group. Results also highlighted an effect of the test session independently of the group. Specifically in the PWS-EG group, analyses showed a significant increase in scores between the pre-test session and the first post-test session, and a strong tendency to increase between the first and the second post-test session.

In the ERC questionnaire, analyses showed a strong tendency toward interaction. Scores in the PWS-EG group increased between the pre-test session and the first post-test session, without any significant differences between the two post-test sessions.

Lastly, in the third step of the analyses, we observed a significant group effect for the EMOmim task (Facial modality only), the three recognition tasks and the AJQ tasks. Scores in the PWS-EG group were significantly higher than those in the PWS-CG group at the second post-test session. The scores of the PWS-EG group no longer statistically differed from those of the two TD groups in the EMOmim task (Facial modality only) and in the AJQ task. In the three recognition tasks, the scores of the PWS-EG group no longer statistically differed from those of the TD-DA group, but they remained significantly lower than those of the TD-CA group. In the ERC questionnaire, scores in the PWS-EG group remained significantly lower than those of the two TD groups and statistically similar to those of the PWS-CG group. In the EMOrea task, the four groups remained statistically similar.

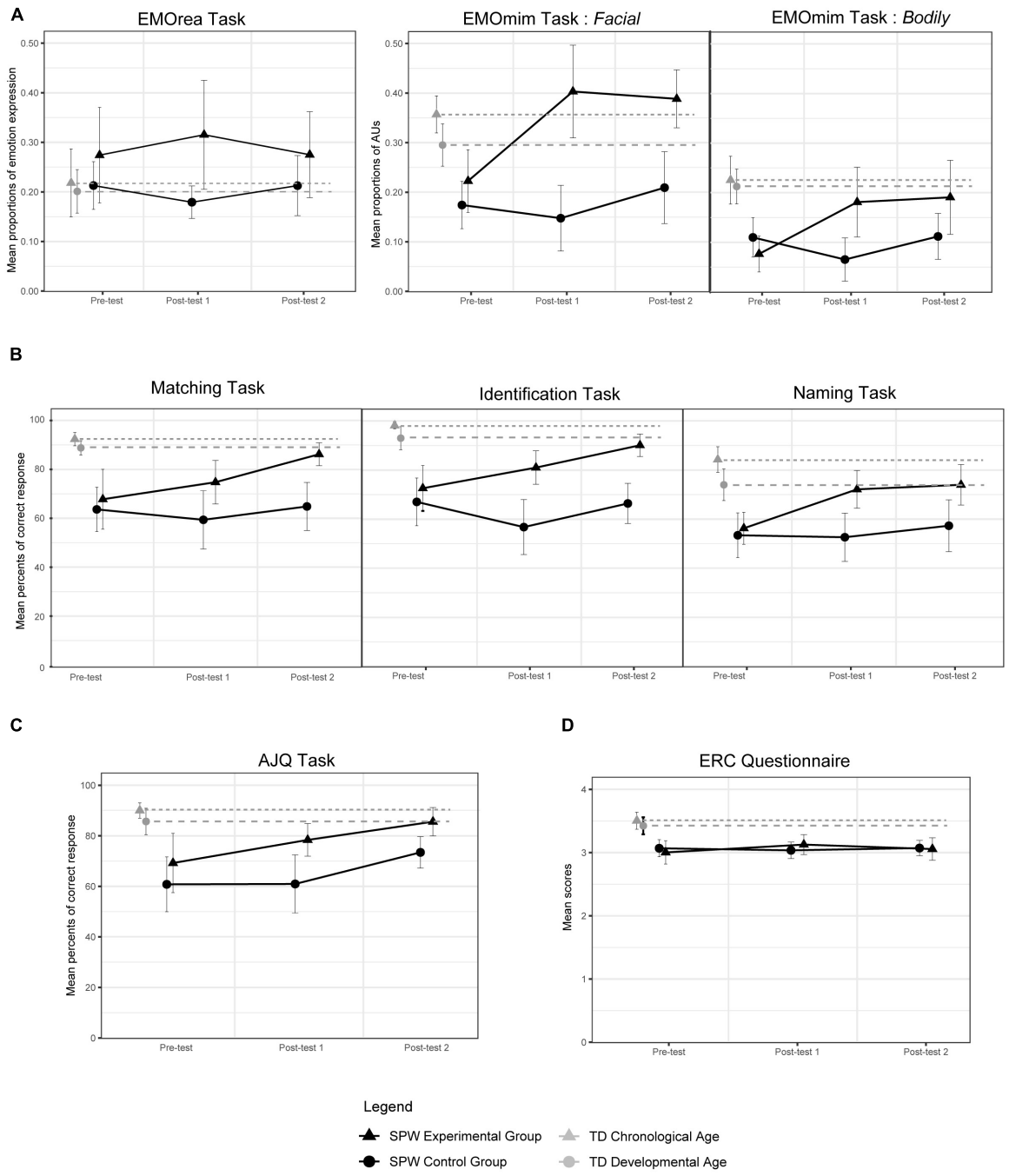


FIGURE 2
Graphical results of the three test sessions for each task. **(A)** Expression. **(B)** Recognition. **(C)** Comprehension. **(D)** Regulation.

Discussion

This study aimed to test the effect of the EMO-T programme on EC, based on an integrative developmental approach. The programme was applied to children with PWS as they display specific patterns of emotion regulation which are part of a broader and multidimensional problem.

Analyses showed four main results. First, PWS children displayed a specific deficit in the four EC (i.e., expression, recognition, comprehension, and regulation of emotions). Second, at post-test 2, PWS children who benefited from the EMO-T programme showed a significant improvement in their voluntary expression and recognition abilities. Third, this level reached was maintained over time, and even continued to

10.3389/fpsyt.2022.1038223

TABLE 2 Summary of results of the three steps of analyses for each task.

Task	Base level	Sign improvement		Level reached
	Comparison group pre-test	EG: Pre≠post1	EG: Post1≠2	Comparison group post-test 2
Expression				
EMOrea	**EG**–CG–DA–CA	→	→	**EG**–CG–DA–CA
	ns	*Ns*		*ns*
EMOmim				
Facial	**EG**–CG < DA–CA	↗°	→°	CG < DA–CA–**EG**
Bodily	**EG**–CG < DA–CA	↗°	→	**EG**–CG < DA–CA
	***	***		***
Recognition				
Matching	**EG**–CG < DA < CA	↗°	↗°	CG < DA–**EG** < CA
Identification	**EG**–CG < DA < CA	↗°	↗°	CG < DA–**EG** < CA
Naming	**EG**–CG < DA < CA	↗°	→°	CG < DA–**EG** < CA
	***	***		***
Comprehension				
AJQ	CG ≤ **EG** < DA–CA	(↗°)	(↗°)	CG < DA–CA–**EG**
	***	*Ns*		***
Regulation				
ERC	**EG**–CG < DA–CA	(↗°)	(→)	**EG**–CG < DA–CA
	***	¤		***

EG, PWS experimental group; CG, PWS control group; CA, TD group with chronological age matched; DA, TD group with development age matched. Comparisons group (X-Y: no difference score between X and Y; X < Y: X's score is lower than Y's score; X ≤ Y: X's score tends to be lower than Y's score). The improvement sign shows the difference between pre-test and post-test 1/between post-test 1 and 2 for the PWS-EG group (→: no difference; ↗: increase; °: higher score than PWS-CG group). *P*-values (*ns*: not significant; ***$p < 0.001$; ¤: tendency $0.05 < p < 0.08$). *P*-values refer to group effect analyses (comparison group at pre-test and at post-test 2) and to interaction effect analyses (two PWS groups × three test sessions).

increase beyond the programme (recognition). Fourth, the post-programme level of PWS children was no longer different from the baseline level of TD children matched by intellectual developmental age, nor even of TD children matched by chronological age (voluntary expression and comprehension).

Lastly, PWS children in the experimental group tended to improve in their emotion regulation, although they received no specific training in this skill. These results support the developmental model of EC, showing that emotion regulation abilities require expression, recognition, and comprehension skills. Such prerequisite skills should be more fully considered in current training programmes.

Nevertheless, the improvement in emotion regulation was limited and was not sustained over time, which can be attributed to several reasons. First and foremost, the mixed results regarding emotion comprehension could be a cause of this. Since comprehension abilities require several skills in many areas, especially cognitive skills, they are considered as the pillar of emotional development (7, 8, 11). In this light, the programme should include the remediation of cognitive and executive abilities, which are also heavily involved in EC, as highlighted in the study by Li et al. (52) or the study by Weiss et al. (53).

The limited sample size could also have made the results less consistent. The fact that the theoretical sample determined from the power analysis could not be reached may also explain the trend results in the emotion regulation scale. In addition, the great variability of profiles existing in PWS (35) made it more complex to set up suitable tasks. This can be illustrated by the three children who were withdrawn from the analyses because they showed too low a level of understanding and were not able to follow the instructions.

The unsustainable improvement in emotion regulation could also be due to the programme format. The EMO-T programme was applied over 6 weeks, a choice that was determined by organizational constraints, but this can be considered as a minimum duration. From the other programmes, a longer period of 3 months seems to offer a better opportunity of stabilizing the progression (25, 26, 31). Additionally, the EMO-T programme could be made more progressive, focusing initially on the ability to express and recognize emotions, since they are prerequisites for social perception skills (43). From this basis, the sessions could then include exercises on social cognition (emotion comprehension, ToM, etc.). The work on emotion regulation could then be initiated in a final stage. Feedback from therapists supports this proposition, although they also highlighted that a certain degree of redundancy may be relevant for some children who need a routine.

Lastly, the issue of how to measure emotional regulation in children remains unresolved. It is still difficult to develop a standardized methodology to carry out direct observations to measure how and when regulatory strategies are used, and to assess their effectiveness (11). It would be worthwhile further investigating board games in this respect because they create a specific context that requires regulation skills in children (30). The coding system used in Hagstrøm et al.'s (54) study or in Penela et al. (55) also shows interesting possibilities.

To conclude, this study underlined the value of considering a developmental model to promote an EC training programme. This appears crucial especially in the case of neurodevelopmental disorders with multidimensional dysfunctions. The study also showed the importance of adopting an integrative approach, by including the training programme in regular care. The EMO-T programme delivered in a therapeutic context showed an improvement through assessment sessions conducted in the family context. Interestingly, the results on the emotion regulation questionnaire completed by parents showed a potential impact of the programme on daily life. The transfer of acquisitions could be even stronger, however, by considering the "caregiver triad": family, school, and therapists. Further studies should be conducted to explore this perspective.

Data availability statement

The raw data supporting the conclusions of this article are available from the corresponding author on reasonable request.

Ethics statement

Ethical review and approval was not required for the current study in accordance with the local legislation and institutional requirements. Only voluntary children with informed parental consent participated in the study. In line with the latest Declaration of Helsinki (2013), all the children, parents and therapists were fully informed of the nature and characteristics of the study. Written informed consent to participate in this study was provided by the participants' legal guardian/next of kin.

Author contributions

NF: elaboration of the study and the method, data collection, data analysis, and writing the manuscript. MT and MGu: study supervision and writing the manuscript. GD, SÇ-B, MGl, and CM: help with the recruitment of participants and the data collection. All authors contributed to the article and approved the submitted version.

Funding

This study was supported by grants from the University of Toulouse and the Occitanie Region (France). This work was also supported by funding from the Prader–Willi Syndrome French Reference Center and the French Prader–Willi Association.

Acknowledgments

We thank all the participants and their families included in this study, as well as the therapists who agreed to collaborate on this project. We are deeply grateful to the National Reference Center and the French Prader-Willi Association for financial support and for their help in recruiting the participants. We also thank the CLLE Laboratory for assistance with the statistical analysis and equipment.

Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

Publisher's note

All claims expressed in this article are solely those of the authors and do not necessarily represent those of their affiliated organizations, or those of the publisher, the editors and the reviewers. Any product that may be evaluated in this article, or claim that may be made by its manufacturer, is not guaranteed or endorsed by the publisher.

Supplementary material

The Supplementary Material for this article can be found online at: https://www.frontiersin.org/articles/10.3389/fpsyt.2022.1038223/full#supplementary-material

References

1. Mikolajczak M. Emotional Competence. In: Carducci B, Nave C editors. *The wiley encyclopedia of personality and individual differences*. New Haven, CT: Wiley (2020). p. 137–41. doi: 10.1002/9781118970843.ch200

2. Hsiao Y. Parental stress in families of children with disabilities. *Interv Sch Clin.* (2018) 53:201–5. doi: 10.1177/1053451217712956

3. Decety J. The neurodevelopment of empathy in humans. *Dev Neurosci.* (2010) 32:257–67. doi: 10.1159/000317771

4. Denham S. Emotional Competence During Childhood and Adolescence. In: LoBue V, Pérez-Edgar K, Buss K editors. *Handbook of emotional development*. Cham: Springer International Publishing (2019). p. 493–541. doi: 10.1007/978-3-030-17332-6_20

5. Pollak S, Camras L, Cole P. Progress in understanding the emergence of human emotion. *Dev Psychol.* (2019) 55:1801–11. doi: 10.1037/dev0000789

6. Walle E, Reschke P, Camras L, Campos J. Infant differential behavioral responding to discrete emotions. *Emotion.* (2017) 17:1078–91. doi: 10.1037/emo0000307

7. Denham S, Blair K, DeMulder E, Levitas J, Sawyer K, Auerbach-Major S, et al. Preschool emotional competence: pathway to social competence? *Child Dev.* (2003) 74:238–56. doi: 10.1111/1467-8624.00533

8. Camras L, Halberstadt A. Emotional development through the lens of affective social competence. *Curr Opin Psychol.* (2017) 17:113–7. doi: 10.1016/j.copsyc.2017.07.003

9. Famelart N, Diene G, Çabal-Berthoumieu S, Glattard M, Molinas C, Guidetti M, et al. Equivocal expression of emotions in children with prader-willi syndrome: what are the consequences for emotional abilities and social adjustment? *Orphanet J Rare Dis.* (2020) 15:55. doi: 10.1186/s13023-020-1333-9

10. Flournoy J, Pfeifer J, Moore W, Tackman A, Masten C, Mazziotta J, et al. Neural reactivity to emotional faces may mediate the relationship between childhood empathy and adolescent prosocial behavior. *Child Dev.* (2016) 87:1691–702. doi: 10.1111/cdev.12630

11. Nader-Grosbois N, Mazzone S. Validation of the french version of the emotion regulation checklist (ERC-vf). *Eur Rev Appl Psychol.* (2015) 65:29–41. doi: 10.1016/j.erap.2014.10.002

12. Parsafar P, Davis E. Intrapersonal emotion regulation processes influence what children remember about their emotional experiences. *Child Dev.* (2019) 90:1935–51. doi: 10.1111/cdev.13070

13. Denham S, Burton R. *Social and emotional prevention and intervention programming for preschoolers*. Boston, MA: Springer (2003). doi: 10.1007/978-1-4615-0055-1

14. Izard C, Stark K, Trentacosta C, Schultz D. Beyond emotion regulation: emotion utilization and adaptive functioning. *Child Dev Perspect.* (2008) 2:156–63. doi: 10.1111/j.1750-8606.2008.00058.x

15. Stegge H, Meerum Terwogt M. Awareness and regulation of emotion in typical and atypical development. In: Gross J editor. *Handbook of emotion regulation*. New York, NY: Guilford Press (2007). p. 288–305.

16. Thompson R. Emotion and emotion regulation: two sides of the developing coin. *Emot Rev.* (2011) 3:53–61. doi: 10.1177/1754073910380969

17. Cole P, Jacobs A. From children's expressive control to emotion regulation: looking back, looking ahead. *Eur J Dev Psychol.* (2018) 15:658–77. doi: 10.1080/17405629.2018.1438888

18. Famelart N, Guidetti M. The effect of laughter expression modulation on emotional experience in 4 to 10 year-old children. *Eur J Dev Psychol.* (2017) 14:311–23. doi: 10.1080/17405629.2016.1201474

19. Gross J, Thompson R. Emotion Regulation: Conceptual Foundations. In: Gross J editor. *Handbook of emotion regulation*. New York, NY: Guilford Press (2007). p. 3–24.

20. Gross J, Cassidy J. Expressive suppression of negative emotions in children and adolescents: theory, data, and a guide for future research. *Dev Psychol.* (2019) 55:1938–50. doi: 10.1037/dev0000722

21. Mazefsky C, Pelphrey K, Dahl R. The need for a broader approach to emotion regulation research in autism: applying an emotion regulation framework in ASD. *Child Dev Perspect.* (2012) 6:92–7. doi: 10.1111/j.1750-8606.2011.00229.x

22. Domitrovich C, Cortes R, Greenberg M. Improving young children's social and emotional competence: a randomized trial of the preschool "PATHS" curriculum. *J Primary Prevent.* (2007) 28:67–91. doi: 10.1007/s10935-007-0081-0

23. Pons F, Harris P, Doudin P. Teaching emotion understanding. *Eur J Psychol Educ.* (2002) 17:293–304. doi: 10.1007/BF03173538

24. Brackett M, Bailey C, Hoffmann J, Simmons D. RULER: a theory-driven, systemic approach to social, emotional, and academic learning. *Educ Psychol.* (2019) 54:144–61. doi: 10.1080/00461520.2019.1614447

25. Izard C, King K, Trentacosta C, Morgan J, Laurenceau J, Krauthamer-Ewing E, et al. Accelerating the development of emotion competence in head start children: effects on adaptive and maladaptive behavior. *Dev Psychopathol.* (2008) 20:369–97. doi: 10.1017/S0954579408000175

26. Hadwin J, Howlin P, Baron-Cohen S. *Teaching children with autism to mind-read: the workbook*. Hoboken, NJ: John Wiley & Sons (2015). p. 156.

27. Silver M, Oakes P. Evaluation of a new computer intervention to teach people with autism or asperger syndrome to recognize and predict emotions in others. *Autism.* (2001) 5:299–316. doi: 10.1177/1362361301005003007

28. Baron-Cohen S, Golan O, Wheelwright S, Hill J. *Mind reading: the interactive guide to emotions*. London: Jessica Kingsley Publishers (2004).

29. Hopkins I, Gower M, Perez T, Smith D, Amthor F, Casey Wimsatt F, et al. Avatar assistant: improving social skills in students with an ASD through a computer-based intervention. *J Autism Dev Disord.* (2011) 41:1543–55. doi: 10.1007/s10803-011-1179-z

30. Dell'Angela L, Zaharia A, Lobel A, Vico Begara O, Sander D, Samson A. Board games on emotional competences for school-age children. *Games Health J.* (2020) 9:187–96. doi: 10.1089/g4h.2019.0050

31. Begeer S, Gevers C, Clifford P, Verhoeve M, Kat K, Hoddenbach E, et al. Theory of mind training in children with autism: a randomized controlled trial. *J Autism Dev Disord.* (2011) 41:997–1006. doi: 10.1007/s10803-010-1121-9

32. Cassidy S, Schwartz S, Miller J, Driscoll D. Prader-willi syndrome. *Genet Med.* (2012) 14:10–26. doi: 10.1038/gim.0b013e31822bead0

33. Chevalère J, Jauregi J, Copet P, Laurier V, Thuilleaux D, Postal V. Investigation of the relationship between electrodermal and behavioural responses to executive tasks in prader-willi syndrome: an event-related experiment. *Res Dev Disabil.* (2019) 85:229–42. doi: 10.1016/j.ridd.2018.11.013

34. Copet P, Jauregi J, Laurier V, Ehlinger V, Arnaud C, Cobo A, et al. Cognitive profile in a large french cohort of adults with prader-willi syndrome: differences between genotypes. *J Intellect Disabil Res.* (2010) 54:204–15. doi: 10.1111/j.1365-2788.2010.01251.x

35. Whittington J, Holland A. Cognition in people with prader-willi syndrome: insights into genetic influences on cognitive and social development. *Neurosci Biobehav Rev.* (2017) 72:153–67. doi: 10.1016/j.neubiorev.2016.09.013

36. Akefeldt A, Åkefeldt B, Gillberg C. Voice, speech and language characteristics of children with prader-willi syndrome. *J Intellect Disabil Res.* (2007) 41:302–11. doi: 10.1111/j.1365-2788.1997.tb00713.x

37. Dimitropoulos A, Ferranti A, Lemler M. Expressive and receptive language in prader–willi syndrome: report on genetic subtype differences. *J Commun Disord.* (2013) 46:193–201. doi: 10.1016/j.jcomdis.2012.12.001

38. Pansy J, Barones C, Urlesberger B, Pokorny F, Bartl-Pokorny K, Verheyen S, et al. Early motor and pre-linguistic verbal development in prader-willi syndrome – A case report. *Res Dev Disabil.* (2019) 88:16–21. doi: 10.1016/j.ridd.2019.01.012

39. Koenig K, Klin A, Schultz R. Deficits in social attribution ability in prader-willi syndrome. *J Autism Dev Disord.* (2004) 34:573–82. doi: 10.1007/s10803-004-2551-z

40. Reddy L, Pfeiffer S. Behavioral and emotional symptoms of children and adolescents with prader-willi syndrome. *J Autism Dev Disord.* (2007) 37:830–9. doi: 10.1007/s10803-006-0210-2

41. Dykens E, Roof E. Behavior in prader-willi syndrome: relationship to genetic subtypes and age. *J Child Psychol Psychiatry.* (2008) 49:1001–8. doi: 10.1111/j.1469-7610.2008.01913.x

42. Dimitropoulos A, Doernberg E, Russ S, Zyga O. Intervention response by genetic subtype: pretend-preschool program for children with prader-willi syndrome via remote parent training. *J Autism Dev Disord.* (2022) 52:5191–206. doi: 10.1007/s10803-022-05695-9

43. Dykens E, Roof E, Hunt-Hawkins H, Daniell C, Jurgensmeyer S. Profiles and trajectories of impaired social cognition in people with prader-willi syndrome. *PLoS One.* (2019) 14:e0223162. doi: 10.1371/journal.pone.0223162

44. Feldman B, Dimitropoulos A. Face discrimination skills in prader-willi syndrome and autism spectrum disorder. *J Mental Health Res Intellect Disabil.* (2014) 7:264–85. doi: 10.1080/19315864.2013.857744

45. Debladis J, Valette M, Strenilkov K, Mantoulan C, Thuilleaux D, Laurier V, et al. Face processing and exploration of social signals in prader-willi syndrome: a genetic signature. *Orphanet J Rare Dis.* (2019) 14:262. doi: 10.1186/s13023-019-1221-3

46. Strenilkov K, Debladis J, Salles J, Valette M, Mantoulan C, Thuilleaux D, et al. A study of voice and non-voice processing in prader-willi syndrome. *Orphanet J Rare Dis.* (2020) 15:22. doi: 10.1186/s13023-020-1298-8

47. Mahmoud R, Swanson H, Butler M, Flodman P, Gold J, Miller J, et al. Molecular classes and growth hormone treatment effects on behavior and emotion in patients with prader–willi syndrome. *JCM.* (2022) 11:2572. doi: 10.3390/jcm11092572

48. Tauber, M, Boulanouar K, Diene G, Çabal-Berthoumieu S, Ehlinger V, Fichaux-Bourin P, et al. The use of oxytocin to improve feeding and social skills in infants with prader–willi syndrome. *Pediatrics.* (2017) 139:1–11. doi: 10.1542/peds.2016-2976

49. Holland A, Manning K. t-VNS to treat disorders of behaviour in prader-willi syndrome and in people with other neurodevelopmental conditions. *Auto Neurosci.* (2022) 239:102955. doi: 10.1016/j.autneu.2022.102955

50. Dykens E, Roof E, Hunt-Hawkins H, McDonald C. The feasibility and effectiveness of a novel, on-line social skills intervention for individuals with prader-willi syndrome. *Front Psychiatry.* (2022) 13:863999. doi: 10.3389/fpsyt.2022.863999

51. Hollingshead A. *Four factor index of social status. Unpublished working paper*. New Haven, CT: Yale University (1975).

52. Li Q, Liu P, Yan N, Feng T. Executive function training improves emotional competence for preschool children: the roles of inhibition control and working memory. *Front Psychol.* (2020) 11:347. doi: 10.3389/fpsyg.2020.00347

53. Weiss J, Thomson K, Burnham Riosa P, Albaum C, Chan V, Maughan A, et al. A randomized waitlist-controlled trial of cognitive behavior therapy to improve emotion regulation in children with autism. *J Child Psychol Psychiatr.* (2018) 59:1180–91. doi: 10.1111/jcpp.12915

54. Hagstrøm J, Spang K, Vangkilde S, Maigaard K, Skov L, Pagsberg A, et al. An observational study of emotion regulation in children with tourette syndrome. *J Child Psychol Psychiatr.* (2021) 62:790–7. doi: 10.1111/jcpp.13375

55. Penela E, Walker O, Degnan K, Fox N, Henderson H. Early behavioral inhibition and emotion regulation: pathways toward social competence in middle childhood. *Child Dev.* (2015) 86:1227–40. doi: 10.1111/cdev.12384

frontiers | Frontiers in Psychiatry

TYPE Brief Research Report
PUBLISHED 10 January 2023
DOI 10.3389/fpsyt.2022.1078211

OPEN ACCESS

EDITED BY
Jeffrey C. Glennon,
University College Dublin, Ireland

REVIEWED BY
Wolfgang Rauch,
Ludwigsburg University, Germany
Maria Cristina Porfirio,
Policlinico Tor Vergata, Italy

*CORRESPONDENCE
Mirko Uljarević
✉ mirkoulj@stanford.edu

SPECIALTY SECTION
This article was submitted to Autism, a section of the journal Frontiers in Psychiatry

RECEIVED 24 October 2022
ACCEPTED 16 December 2022
PUBLISHED 10 January 2023

CITATION
Uljarević M, Cai RY, Hardan AY and Frazier TW (2023) Development and validation of the Executive Functioning Scale. *Front. Psychiatry* 13:1078211. doi: 10.3389/fpsyt.2022.1078211

Development and validation of the Executive Functioning Scale

Mirko Uljarević[1]*, Ru Ying Cai[2], Antonio Y. Hardan[1] and Thomas W. Frazier[3]

[1]Department of Psychiatry and Behavioral Sciences, School of Medicine, Stanford University, Stanford, CA, United States, [2]Aspect Research Centre for Autism Practice, Frenchs Forest, NSW, Australia, [3]Department of Psychology, John Carroll University, University Heights, OH, United States

Executive functioning (EF) processes are essential for adaptive and flexible responding to the demands and complexities of everyday life. Conversely, if impaired, these processes are a key transdiagnostic risk factor that cuts across autism and a range of other neurodevelopmental (NDD) and neuropsychiatric (NPD) conditions. However, there are currently no freely available informant-report measures that comprehensively characterize non-affective (e.g., working memory, response inhibition, and set shifting) and affective (e.g., emotion regulation) EF subdomains. This study describes the development, refinement, and initial psychometric evaluation of a new 52-item Executive Functioning Scale (EFS). Two independent data collections yielded exploratory (n = 2004, 169 with autism, ages 2–17) and confirmatory (n = 954, 74 with autism, ages 2–17) samples. Exploratory Structural Equation Modeling (ESEM) model with six specific factors that closely matched hypothesized executive functioning subdomains of working memory and sequencing, response inhibition, set-shifting, processing speed, emotion regulation, and risk avoidance, and one general factor, showed the best fit to the data and invariance across age, sex, race, and ethnicity groups. Model reliability and internal consistency were excellent for the general factor (ω = 0.98; α = 0.97) and specific factors ($\omega \geq 0.89–0.96$; $\alpha \geq 0.84–0.94$). Conditional reliability estimates indicated excellent reliability ($\geq$0.90) for the total EF scale and adequate or better reliability ($\geq$0.70) for subscale scores. With further replication, the EFS has excellent potential for wide adoption across research and clinical contexts.

KEYWORDS

emotion regulation, assessment, autism, executive functioning, working memory, response inhibition, self-regulation, neurodevelopmental

10.3389/fpsyt.2022.1078211

1. Introduction

Executive functioning (EF), emotion regulation (ER), and valuation of risk and reward are essential processes for adaptive and flexible responding to continuously shifting tasks and demands, and complexities of everyday life (1–3). Indeed, these processes underpin healthy social and emotional development (4–6) and have been associated with a range of outcomes, including academic performance (7), healthy habits (8), and different aspects of quality of life (9). Conversely, EF and ER impairments have been suggested as critical transdiagnostic risk factors that cut across a range of neurodevelopmental (NDD) and neuropsychiatric (NPD) disorders (10–14). For instance, impairments in these processes are frequent in autism spectrum disorder [ASD; (15, 16)], attention-deficit/hyperactivity disorder [ADHD; (17, 18)], schizophrenia (19, 20), depression (21, 22), obsessive-compulsive disorder [OCD; (23, 24)] and posttraumatic stress disorder (25). Further, EF and ER difficulties are associated with specific symptom domains commonly observed across NDD/NPD, including anxiety (16, 26), sameness/rituals (27, 28), social functioning difficulties (29, 30), positive and negative symptoms of psychoses (31–33) and externalizing problems (34). Thus, the existence of measures that can comprehensively evaluate noted processes that represent vital risk factors for developing and maintaining a range of clinically impactful symptoms seen across NDD and NPD is crucial for advancing etiological research and identifying treatment targets.

Although it is widely accepted that EF is best understood as a complex and multifaceted domain comprising several distinct, yet related subdomains mediated by fronto-striatal circuits (1, 35), the consensus in terms of the specific components is still lacking. More specifically, certain frameworks have focused on decontextualized and non-affective processes, emphasizing working memory, response inhibition, and set shifting, and in some instances, sequencing and planning as core EF components [e.g., (2, 36)]. Others have emphasized the need for broader conceptualization that in addition to noted "cool" EF components, also includes affective-related (or "hot") processes, in particular, monitoring and modifying emotional responses, or ER (3, 37, 38), and risk aversion/risk-taking that encompasses evaluation of reward and punishment probability (39–41). In addition, although processing speed has not been consistently included in the definitions of EF, it has been noted that it is crucial for EF models and assessments to consider and capture processing speed given that it can underly distinct EF subdomains and has been demonstrated to show additional predictive validity regarding a range of psychopathology manifestations (42, 43). Given the noted complexity and lack of universally agreed on EF taxonomy, it is necessary for instruments to enable fine-grained capture of individual differences in a range of distinct "hot" and "cold" EF subdomains.

Several questionnaire instruments were specifically designed for assessing EF deficits in NDD and NPD. These include the Behavior Rating Inventory of Executive Functioning, second edition [BRIEF-2; (44)], the Comprehensive Executive Function Inventory [CEFI; (45)], and the Barkley Deficits in Executive Functioning Scale [BDEFS; (46, 47, 48)]. These measures have been shown to have better ecological validity compared to performance-based and experimental batteries, including, but not limited to, Delis-Kaplan Executive Function System [D-KEFS; (49)], Cambridge Neuropsychological Test Automated Battery [CANTAB; (50)], and a Developmental Neuropsychological Assessment, Second Edition [NEPSY-2; (51)], NIH Toolbox Cognition Battery (52), or the Computerized Battery for Neuropsychological Evaluation of Children [BENCI; (53)]. Further, the BRIEF-2, CEFI, and BDEFS have been extensively used across normative and a range of clinical populations, generally showing good validity and reliability. However, the above-noted instruments present a range of significant limitations. Firstly, BRIEF-2, CEFI, and BDEFS are all commercial instruments which significantly limits access and use in large-scale clinical and research collection efforts. Secondly, these measures have poor coverage and representation of specific domains. For instance, the BDEFS does not capture set-shifting. Importantly, none of the instruments capture the upregulation of positive emotions, which is a facet of ER that is just as important as the down-regulation of negative emotions (54–56) and when excessive, may also be associated with reward sensitivity and difficulties avoiding risk (57). Thirdly, there is limited evidence for the construct validity, measurement invariance, and conditional reliability of existing instruments (58). Measurement invariance is particularly important for ensuring that the measure is applicable across a broad demographic spectrum. Demonstrating good conditional reliability across a wide range of score levels is crucial for accurate assessment across neurotypical and pathological EF levels and essential for tracking change across development and due to interventions. Finally, rather than assessing everyday, developmentally appropriate behavioral instances related to specific facets of executive functioning, most available instruments assess symptoms/behavioral psychopathology thought to result from EF deficits. This focus on symptoms significantly limits the ability to capture subtle variations in functional abilities and to understand associations between these processes and specific symptom domains.

1.2. The present study

The present paper describes the development and preliminary psychometric evaluation of the Executive Functioning Scale (EFS)—a brief, freely available, informant-report measure specifically designed to address limitations

of existing instruments and comprehensively characterize individual variation in specific, well-defined facets of EF across the normative-pathological continuum. The EFS was developed based on the recommendations for item generation and refinement outlined by the National Institute of Health's Scientific Standards of the Patient-Reported Outcomes Measurement Information System (PROMIS) and in conjunction with NDD and NPD individuals and their parents. Detailed psychometric evaluation was conducted in two independent, large, representative samples spanning normative and atypical development and included evaluation of factor structure, measurement invariance, classical test theory and item response theory-derived reliability, and testing of convergent and discriminant validity.

2. Materials and methods

2.1. Participants

Parent informants were recruited using the Prolific online data collection service,[1] and interested participants were directed *via* a link to the Qualtrics survey. Two separate data collections were conducted to establish exploratory and confirmatory samples. For the exploratory sample, data were collected from 03/04/2022 to 04/17/2022. A total of 2,486 informants consented and responded to the survey, with the final sample comprising 2,004 valid responses (124 respondents were excluded due to not completing the survey, 72 were excluded due to completing too rapidly to produce valid results, and 286 were excluded due to failing at least one of the four attention checks from the modified Conscientious Responders Scale [CRS; (59)]. According to informant reports, 169 children have received a diagnosis of autism spectrum disorder, 541 had other NDD/NPD, and 1,294 were neurotypical. For the confirmatory sample, data were collected from 05/03/2022 to 07/20/2022. A total of 1,361 informants consented and responded to the survey; however, given the considerable length of the survey, EFS data was collected only from 954 responders to reduce the participant burden (407 participants who did not complete the EFS completed additional instruments). Thus, the final confirmatory sample comprised 954 responses. Based on informant reports, 74 children have received a diagnosis of autism spectrum disorder, 249 had other NDD/NPD, and 631 were neurotypical. Inclusion criteria for both exploratory and confirmatory samples included: residence in the US, having a dependent child aged 2–17, and informant proficiency in English. Detailed characteristics across exploratory and confirmatory samples are presented in **Table 1**.

1 https://prolific.co/

TABLE 1 Demographic and clinical characteristics across autism spectrum disorder (ASD), developmental disability (DD), and neurotypical (NT) controls across exploratory and confirmatory samples.

	NT	DD	ASD	X^2/F (p)
	n (%)	*n* (%)	*n* (%)	
N	1925	790	243	
Informant (n,%)				74.86 (<0.001)
Biological mother	1119 (58.1%)	513 (64.9%)	167 (68.7%)	
Biological father	686 (35.6%)	184 (23.3%)	53 (21.8%)	
Other/Not reported	120 (6.3%)	93 (11.8%)	23 (9.5%)	
Highest parental education (n,%)				26.8 (0.003)
Less than HS	6 (0.6%)	2 (0.6%)	1 (1.0%)	
High school or GED	90 (8.9%)	38 (10.8%)	11 (10.6%)	
Some college	178 (17.6%)	94 (26.8%)	32 (30.8%)	
College graduate	427 (42.2%)	132 (37.6%)	40 (38.5%)	
Graduate degree or higher	295 (29.2%)	80 (22.8%)	18 (17.3%)	
Unknown	16 (1.6%)	5 (1.4%)	2 (1.9%)	
US region				10.9 (0.205)
Northeast	188 (18.6%)	51 (14.5%)	16 (15.4%)	
Midwest	215 (21.3%)	69 (19.7%)	23 (22.1%)	
South	402 (39.8%)	168 (47.9%)	50 (48.1%)	
West	203 (20.1%)	62 (17.7%)	15 (14.4%)	
Other/Chose not to respond	4 (0.4%)	1 (0.3%)	0 (0.0%)	
Household income (n,%)				59.2 (<0.001)
<$25,000	281 (9.5%)	92 (11.6%)	38 (15.6%)	
$25,000–$34,999	288 (9.7%)	87 (11.0%)	29 (11.9%)	
$35,000–$49,999	348 (11.8%)	96 (12.2%)	43 (17.7%)	
$50,000–$74,999	641 (21.7%)	176 (22.3%)	50 (20.6%)	
$75,000–$99,999	496 (16.8%)	140 (17.7%)	34 (14.0%)	
$100,000–$149,999	563 (19.0%)	129 (16.3%)	26 (10.7%)	
$150,000–$199,999	176 (5.9%)	36 (4.6%)	13 (5.3%)	
$200,000 and above	138 (4.7%)	28 (3.5%)	8 (3.3%)	
Unknown	27 (0.9%)	6 (0.8%)	2 (0.8%)	
Child age (M, SD)	8.58 (4.70)	11.46 (4.50)	10.31 (4.74)	111.8 (<0.001)
Child biological sex (n,% male)	915 (47.7%)	431 (54.6%)	181 (74.8%)	69.9 (<0.001)
Race				
White/Caucasian (n,%)	1578 (82.0%)	658 (83.3%)	200 (82.3%)	0.67 (0.716)
Black/African American (n,%)	182 (9.5%)	71 (9.0%)	29 (11.9%)	1.9 (0.385)
Middle Eastern (n,%)	5 (0.3%)	4 (0.5%)	2 (0.8%)	2.4 (0.305)

(Continued)

TABLE 1 (Continued)

	NT	DD	ASD	X^2/F (p)
	n (%)	*n* (%)	*n* (%)	
East Asian (n,%)	66 (3.4%)	11 (1.4%)	4 (1.6%)	9.9 (0.007)
South Asian (n,%)	33 (1.1%)	2 (0.1%)	2 (0.1%)	10.1 (0.006)
Pacific Islander (n,%)	10 (0.5%)	4 (0.5%)	1 (0.1%)	0.5 (0.975)
Native American (n,%)	22 (1.1%)	20 (2.5%)	6 (2.5%)	7.9 (0.019)
Multiracial (n,%)	151 (5.1%)	74 (2.5%)	27 (0.0%)	3.9 (0.139)
Unknown race (n,%)	3 (0.2%)	2 (0.3%)	0 (0.0%)	0.7 (0.683)
Chose not to respond (n,%)	15 (0.8%)	3 (0.4%)	1 (0.4%)	1.6 (0.445)
Hispanic or Latino (n,%)	101 (11.4%)	43 (12.1%)	26 (22.4%)	12.4 (0.015)
Non-ASD diagnoses (n,%)				
ID/GDD	–	10 (2.8%)	6 (5.8%)	2.1 (0.150)
Speech/Language disorder	–	75 (21.4%)	16 (15.5%)	1.7 (0.193)
ADHD	–	146 (41.6%)	29 (27.9%)	6.1 (0.014)
ODD/CD	–	25 (7.1%)	5 (4.9%)	0.7 (0.415)
Anxiety disorder	–	111 (31.6%)	19 (18.4%)	6.8 (0.009)
Specific learning disorder	–	33 (9.4%)	3 (2.9%)	4.6 (0.032)
Motor/Coordination disorder	–	16 (4.6%)	2 (1.9%)	1.4 (0.231)
Depressive disorder	–	50 (14.2%)	8 (1.8%)	3.0 (0.083)
Bipolar disorder/Mania	–	7 (2.0%)	1 (1.0%)	0.5 (0.488)
Obsessive compulsive disorder	–	11 (3.1%)	5 (4.9%)	0.7 (0.405)
Tic disorder	–	6 (1.7%)	1 (1.0%)	0.3 (0.593)
Feeding/Eating disorder	–	16 (4.6%)	0 (0.0%)	4.9 (0.029)

NT, neurotypical controls; DD, non-ASD developmental disability; ASD, autism spectrum disorder; ID/GDD, intellectual disability/global developmental delay; ADHD, attention-deficit/hyperactivity disorder; ODD/CD, oppositional defiant disorder/conduct disorder. Non-ASD diagnoses do not sum to 100% because children could be diagnosed with more than one condition. Cognitive level information was completed for $n = 886$.

2.2. Measures

2.2.1. Exploratory sample

2.2.1.1. Demographic and diagnostic information

Informants completed a background survey indicating informant and child age, informant and child gender, child race/ethnicity, informant relationship status, household income, and estimates of autism spectrum disorder (ASD) symptom severity and cognitive level. Informants also indicated whether the child had a clinical diagnosis of neurodevelopmental or neuropsychiatric disorder, including intellectual disability/global developmental delay, speech/language disorder, attention-deficit/hyperactivity disorder, oppositional defiant disorder/conduct disorder, anxiety disorder, specific learning disorder, motor/coordination disorder, depressive disorder, bipolar disorder/mania, OCD, tic disorder, and feeding/eating disorder. All children without ASD but with other diagnoses were recoded to a developmental disability (DD) category. Participants with no developmental or neuropsychiatric diagnosis were assigned to the neurotypical (NT) group.

2.2.1.2. Executive Functioning Scale (EFS)

EFS items were developed and refined through the iterative steps described by the PROMIS framework described below.

2.2.1.2.1. Conceptual model generation

Systematic literature search was performed to identify existing EF instruments and conceptual models. As noted, there is a lack of universally agreed-on EF taxonomy. Given different definitions and that a wide range of potential EF subdomains put forward across different conceptualizations is strongly related to clinical symptoms observed across NDD and NPD, we took the position that it is important to capture a broad set of distinct "hot" and "cold" EF subdomains. Identified subdomains included: working memory and sequencing, response inhibition, set-shifting, processing speed, emotion regulation, and risk avoidance.

2.2.1.2.2. Item writing

Systematic review of the literature was conducted to identify existing scales relevant to each domain and content area. Scales reviewed included: (i) dedicated questionnaire measures of executive functioning such as the BRIEF, ECI, and BDEFS, (ii) dedicated experimental measures of distinct executive functioning domains such as the Dimensional Card Sorting Task, Stroop Task, as well as comprehensive testing batteries including D-KEFS (49), CANTAB (50), and a NEPSY-2 (51); (iii) general psychopathology and development instruments including the Behavior Assessment System for Children [BASC; (60)] and Infant–Toddler Social and Emotional Assessment [ITSEA; (61)], (iv) temperament and personality measures [e.g., the Infant Behavior Questionnaire (62)], and (v) measures of emotion regulation and self-regulation such as the Emotion Regulation Checklist (63) and Difficulties in Emotion Regulation Scale (64). The first and senior authors reviewed instruments, and items across identified measures were organized into specific latent constructs, then pruned and adjusted for consistency. At least three items were written to ensure that the content area is adequately assessed and that future analyses on these items could identify any sub-factors within each domain. As much as possible, items were developed not to probe more than one construct, or the endorsement of an item is not a consequence of distinct processes.

2.2.1.2.3. Preliminary item evaluation and refinement

Fifty-two items were developed by the research team to evaluate each of the above executive functioning subdomains. The preliminary item bank was evaluated by ten neurodevelopmental disability clinician-scientist experts and ten neurodevelopmental disability caregiver/patient informants with regards to whether each item: (i) effectively evaluated the specific executive functioning subdomain (experts and informants), (ii) was relevant to patients (experts) or child (informants), (iii) was relevant to the full age and functional range of patients (experts), and (iv) was easy/difficult to understand (experts and informants). Neither parents nor experts indicated the need to remove any items; no additional behaviors/skills were identified as missing. Minor wording changes were made to several items following parental feedback.

2.2.1.2.4. Final scale

The final scale consisted of 52 items that were rated on a 5-point Likert scale (0 = Never, 1 = Rarely 2 = Sometimes 3 = Often 4 = Very Often). Parents were instructed to "for each item, please indicate how often over the last week the person shows this behavior, skill, or ability using the response options below."

2.2.2. Confirmatory sample

In addition to the demographic and diagnostic information questionnaire and the EFS described above, informants completed a comprehensive set of questionnaire measures to evaluate the convergent and discriminant validity of the EFS. These measures included:

Abbreviated version of the Behavior Rating Inventory of Executive Function [BRIEF-sf; (44, 65)]. BRIEF-sf is a 24-item abbreviated version of the standard BRIEF that has demonstrated good reliability and validity across three independent youth samples (65). BRIEF is an informant-report scale designed to capture different aspects of executive functioning. Each item is rated on a 3-point Likert scale (1 = Never, 2 = Sometimes, 3 = Often). The total raw score was used (higher scores mean more severe impairments).

Attention-deficit/hyperactivity disorder assessment [ADHD-ASSESS; (66)]. ADHD-ASSESS is an 18-item informant-report scale designed to capture ADHD symptoms in children aged 2 to 17, including inattention, hyperactivity, and impulsivity. Each item is rated on a 5-point Likert scale A (0 = Never, 1 = Rarely, 2 = Sometimes, 3 = Often, 4 = Very Often). The total raw score was used (higher scores mean more severe impairments).

Comprehensive Anxiety Scale [CAS; (66)]. CAS is a 35-item informant-report scale designed to capture anxiety symptoms in children aged 2 to 17. The instrument provides a total score and six subscale scores covering generalized anxiety, social anxiety, separation anxiety, panic/physiological anxiety symptoms, obsessive/compulsive symptoms, and specific fears. Each item is rated on a 5-point Likert scale (0 = Never, 1 = Rarely, 2 = Sometimes, 3 = Often, 4 = Very Often). This study focused on the total raw score (higher scores mean more severe impairments).

Daily Living Skills Scale [DLSS; (67)]. DLSS is a 53-item informant-report scale designed to capture daily living skills in children aged 2 to 17. The instrument provides a total and three content subscale scores for enhanced interpretation across self-care, home care, and community participation. Each item is rated on a 4-point Likert Scale [0 = Not able to complete (total assistance needed), 1 = Requires significant prompting or assistance, 2 = Requires minimal prompting or assistance, 3 = Completely independent (does not require any assistance or prompting)]. For the current study, we focused on the total raw score (higher scores mean better ability).

2.3. Procedure

In Qualtrics, prospective participants reviewed an electronic consent form. Participants who decided to continue with participation indicated consent electronically and began the survey. All participants were paid US$10 for survey completion based on the expected survey completion time (35 min). All data were collected anonymously.

2.4. Statistical analyses

Descriptive statistics for demographic and clinical factors were computed to characterize the sample.

2.4.1. Factor structure

Exploratory structural equation models (ESEM) were estimated (68) in the exploratory sample to identify the factor structure of the EFS. These models used weighted least squares mean and variance adjusted estimation, specified four to seven specific factors with an additional general bifactor that included estimation of loadings from all items, and were estimated using geomin rotation. Models were re-estimated in the confirmatory subsample, and the best-fitting model was chosen using a combination of fit statistics and interpretability. Once the best-fitting ESEM model was identified, this model was re-estimated in the total sample, and an equivalent confirmatory ESEM model with all standardized loadings <0.20 set to 0 was estimated. Model fit was evaluated using the comparative fit index (CFI), Tucker–Lewis index (TLI), root mean square error of approximation (RMSEA), and the 95% confidence interval of RMSEA were used to examine model fit (69, 70).

2.4.2. Measurement invariance

The optimal model derived from the factor analyses described above was used as the basis for the evaluation of measurement invariance (71) across age groups (ages 2–4, 5–11, and 12–17 years), sex (male and female), race (Caucasian

10.3389/fpsyt.2022.1078211

and other), and ethnicity (Hispanic and non-Hispanic). To examine measurement invariance (equivalence), a series of multi-group confirmatory factor analyses were computed using the theta parameterization and weighted least square mean and variance adjusted (WLSMV) estimation for categorical indicators, following recommended conventions (72) and prior work (73). Model comparisons for measurement invariance analyses were based on empirical work indicating that a drop in CFI or TLI > 0.01 or an increase in RMSEA > 0.01 implies measurement non-equivalence (74, 75).

2.4.3. Reliability

Using the optimal factor model, items with substantive loadings were assigned to scales and classical test theory (CTT) reliability (internal consistency and correct item-total correlations) (76), and item response theory (IRT) analyses were conducted (77) in the entire sample (n = 2,004). IRT analyses were conducted separately for each scale as the multi-dimensional bifactor IRT model was not possible to estimate. Analyses used maximum likelihood estimation with robust standard errors, and a logit link with the single factor mean and variance fixed to 0 and 1, respectively. Reliability estimates falling in the ranges 0.70 to 0.79, 0.80 to 0.89, and >0.90 were considered fair, good, and excellent (78). Average corrected item-total correlations ≥0.30 were considered adequate or better (76). Differential item and test functioning were evaluated by examining differences in item characteristic curves and test information curves across age groups, sex, race, and ethnicity.

2.4.4. Convergent and discriminant validity

Convergent and discriminant validity were computed using bivariate correlations (Pearson or Spearman's non-parametric, where applicable).

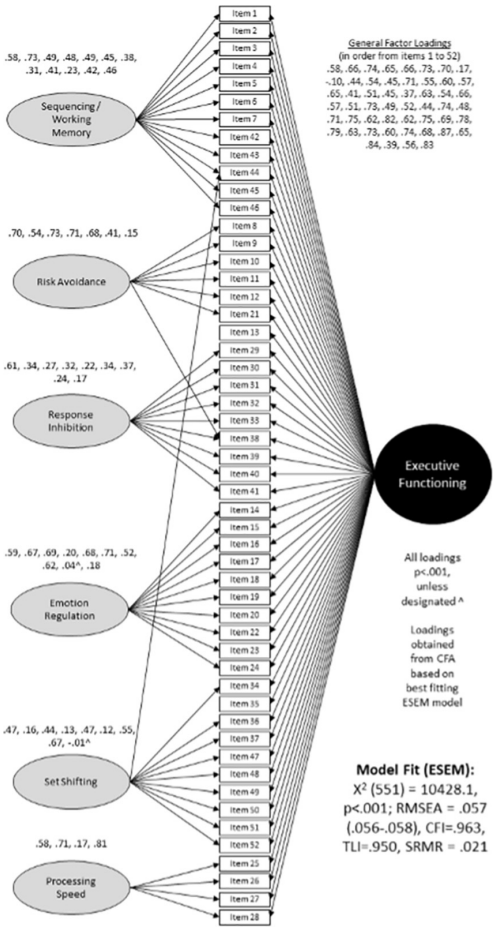


FIGURE 1
Factor structure of the Executive Functioning Scale (EFS).

3. Results

3.1. Participant characteristics

The exploratory sample included 2,004 children and adolescents, and the confirmatory sample included 954 children and adolescents. See **Table 1** for detailed characteristics across exploratory and confirmatory samples.

3.2. Factor structure

In the exploratory and confirmatory samples, ESEM suggested improvements in fit through the six specific factors with a general bifactor ESEM solution. Increases in CFI and TLI and decreases in RMSEA beyond this solution tended to be modest (≤ | 0.006|). The ESEM with seven specific factors and a general executive functioning bifactor had inconsistent difficult to interpret loading patterns across the exploratory and confirmatory subsamples. Thus, the ESEM model with six specific and one general factor was considered the optimal model for additional consideration (**Figure 1**) (Exploratory sample fit indices: CFI = 0.967, TLI = 0.956, RMSEA = 0.054 [95% CI:0.052,0.055], SRMR = 0.021; Confirmatory sample fit indices: CFI = 0.966, TLI = 0.955, RMSEA = 0.055 [95% CI:0.053,0.056], SRMR = 0.022). A final model was estimated in the total sample (N = 2,958) using this model. Fit indices for this model were: CFI = 0.963, TLI = 0.950, RMSEA = 0.057 [95% CI:0.056,0.058], SRMR = 0.021. Given the observation of substantive cross-loadings from items with primary loadings on other factors, a CFA model based on the ESEM model was not estimated. The final scoring was based on the ESEM model, with the subscale choice based on the highest loading for each item. **Figure 1** presents the ESEM factor structure of the EFS in the total sample. Identified factors strongly resembled six conceptually based EF constructs of working memory and sequencing (example items: Follows a complete sequence of steps or actions; Can hold several pieces of information in

mind at once; Is good at remembering the exact way something happened), response inhibition (example items: Stops what they are doing when told to stop; Focuses on finishing important tasks without being distracted by more interesting activities; Can resist immediate desires because they are not good over the long-term), set-shifting (example items: Can transition from one activity to another without problems; Misses important information because they are engrossed in what they are doing; Has trouble with mentally juggling multiple things) processing speed (example items: Seems to process information slowly; Responds slowly, even when asked to do something they enjoy; Works quickly and accurately on an activity), emotion regulation (example items: Has trouble soothing themselves; Remains upset or emotional longer than others; If they are sad, they seem to have difficulty lifting their mood), and risk avoidance (example items: Does not consider possible danger when doing something; Considers consequences before acting; Seems to crave excitement and new experiences).

3.3. Measurement invariance

Estimating measurement invariance for ESEM bifactor models often results in convergence problems. Therefore, a simple confirmatory model without the EF bifactor was used to estimate measurement invariance. This permits examination of measurement equivalence for the subscales with the assumption that if these show scalar invariance, then invariance of the general EF factor is likely. As can be seen from Table 2, results indicated strong (scalar) invariance across age, sex, race, and ethnicity groups.

3.4. Reliability

Table 3 shows detailed reliability indices. As can be seen, model reliability was excellent for the general factor ($\omega = 0.98$) and specific factors ($\omega \geq 0.89$–0.96). Using item scores, internal consistency reliability was excellent for the total scale ($\alpha = 0.97$) and very good to excellent for all subscale scores ($\alpha \geq 0.84$–0.94). Conditional reliability estimates indicated excellent reliability ($\geq$0.90) for the total EF scale from extremely low ($\theta \sim -4.2$) to very high ($\theta \sim +2.6$) scores. Adequate or better reliability ($\geq$0.70) was present for subscale scores in the range from very low ($\theta \sim -3.0$) to high scores ($\sim +1.8$), except processing speed which showed a drop off in measurement precision beyond high average scores ($\theta \sim +0.8$) (Figure 2).

3.5. Convergent and discriminant validity

The EFS showed strong convergent validity with the 24-item BRIEF-sf ($r = 0.85$) and with the ADHD-ASSESS total score ($r = -0.76$); the latter is relevant because ADHD symptoms include several aspects of cognitive functioning that overlap or are closely related to executive functions,

TABLE 2 Measurement invariance analyses for the executive functioning (EF) specific factor model across sex, age, race, and ethnicity.

Sex (M, F)												
		Fit					Difference testing					
Model	Par	X^2	DF	RMSEA	CFI	TLI	X^2	DF	*p*	ΔRMSEA	ΔCFI	ΔTLI
Configural	607	26721.6	2461	0.082	0.904	0.896	–	–	–	–	–	–
Metric	511	9453.5	2557	0.043	0.973	0.972	85.1	96	0.779	−0.039	0.069	0.076
Scalar	331	11630.5	2737	0.047	0.965	0.966	1269.4	180	<0.0001	0.004	−0.008	−0.006
Age (2–4, 5–11, and 12–17)												
Configural	899	25120.7	3703	0.077	0.917	0.911	–	–	–	–	–	–
Metric	707	15692.6	3895	0.055	0.954	0.953	1219.8	192	<0.0001	−0.022	0.037	0.042
Scalar	347	18757.4	4255	0.059	0.944	0.947	4100.1	360	<0.0001	0.004	−0.010	−0.006
Race (Caucasian, other races)												
Configural	607	23891.5	2461	0.077	0.915	0.908	–	–	–	–	–	–
Metric	511	9484.4	2557	0.043	0.973	0.971	112.6	96	0.118	−0.034	0.058	0.063
Scalar	331	9453.0	2737	0.041	0.973	0.974	279.3	180	<0.0001	−0.002	0.000	0.003
Ethnicity (Hispanic, non-Hispanic)												
Configural	607	21860.2	2461	0.073	0.918	0.911	–	–	–	–	–	–
Metric	511	8447.5	2557	0.039	0.975	0.974	69.5	96	0.981	−0.034	0.057	0.063
Scalar	331	8422.9	2737	0.037	0.976	0.977	217.1	180	0.031	−0.002	0.001	0.003

The observed X^2 from WLSMV estimated measurement invariance models cannot be directly compared but rather must be compared using difference testing in MPlus. Thus, the apparent reduction from configural to metric models is not an accurate representation of model fit. Instead, the positive X^2 values from difference testing reflects worse fit of metric relative to configural models.

particularly impulsivity (response inhibition). Evidence of discriminant validity with measures of other aspects of functioning and psychopathological symptoms was also good, including associations with DLS ($r = 0.59$) and CAS ($r = -0.49$). Analysis of the pattern of associations with EFS subscales provided further evidence for convergent and discriminant validity. For instance, the EFS ER subscale was significantly more strongly associated with anxiety (measured by CAS; $r = -0.60$) than with daily living skills (measured by DLSS; $r = 0.35$). Table 4 shows the full list of correlations between EFS total and subscale scores with relevant measures used to establish convergent and discriminant validity.

4. Discussion

The EFS is an informant-report measure developed to comprehensively capture a range of crucial executive functioning subdomains relevant across normative and atypical development, including individuals with ASD, other NDD, and NPD. Findings presented in this initial validation demonstrated that the EFS is a psychometrically sound suggesting that it might be a promising instrument for assessing executive functioning across research and clinical contexts. Indeed, the EFS had a clear and replicable factor structure across two independent samples showing strong reliability, good measurement equivalence across age, sex, race, and ethnicity, and preliminary evidence for good convergent and discriminant validity. Crucially, EFS is considerably briefer (52 items) than other dedicated EF measures, such as the BRIEF (86 items).

The EFS was found to have a well-differentiated factor structure that replicated well across exploratory and confirmatory sub-samples. The final model included six specific factors matching the originally hypothesized EF subdomains of working memory and sequencing, response inhibition, set-shifting, processing speed, ER, and risk avoidance, as well as a general EF factor. As noted, there is a wide range of definitions and conceptualizations of EF, some of which emphasize a narrower range of non-affective ("cool") processes [e.g., (2, 36, 79, 80)] and others arguing for a broader conceptualization that also includes affective ("hot") constructs (37, 39, 41). Although most disorders are associated with relatively uniform impairments across specific constructs of "cool" EF, there are pronounced variations in effect sizes of specific EF deficits in certain conditions (14). Similarly, different NDD and NPD have been suggested to show distinct profiles of "hot" EF subdomains, including ER and risk avoidance (41). Thus, it is essential to fully capture different EF subdomains to understand whether specific subdomains might be more strongly associated with particular aspects of psychopathology.

TABLE 3 Reliability statistics for Executive Functioning Scale (EFS) general (total scores) and specific factors (subscale scores).

	Internal consistency	Model reliability
	α	ω
EF total	0.97	0.98
Sequencing/Working memory	0.94	0.96
Risk avoidance	0.82	0.89
Response inhibition	0.89	0.92
Emotion regulation	0.90	0.93
Set shifting	0.91	0.95
Processing speed	0.84	0.90

Model reliability is McDonald's omega coefficient derived from bifactor modeling.

Several existing instruments, including the BRIEF, CEFI, and BDEFS, have been designed explicitly for assessing deficits in different aspects of EF in NDD and NPD. However, as noted, certain domain coverage and representation limitations, such as lack of coverage of risk avoidance/taking and upregulation of positive emotions, restrict their utility for comprehensive characterization of EF. Further, a key assumption that must be met for widespread measure adoption across diverse sex, age, race, ethnicity, and clinical groups is demonstrated invariance; however, there is little evidence for the invariance of the existing scales. Moreover, the majority of existing EF instruments lacks evidence for conditional reliability, a key feature necessary for capturing and tracking very high and very low levels of a particular trait with good precision. Conversely, the best-fitting EFS model was consistent across sex, age, race, and ethnicity groups, indicating that it can be interpreted consistently when implemented across diverse demographics. Further, conditional reliability estimates showed excellent reliability ($\geq$0.90) for the total EFS from extremely low to very high scores and at least adequate reliability ($\geq$0.70) for subscales from very low to high scores.

Despite the strengths of this two-sample development and validation approach, several limitations are important to note. The main limitation of this study was a reliance on informant reports, including diagnoses, cognitive levels, and symptom severity estimates. Given the online nature of the research and the need to collect large sample sizes across both exploratory and confirmatory samples, it was not possible to independently confirm the diagnostic status of participants and administer gold-standard diagnostic assessments, including the Autism Diagnostic Observation Schedule and the Autism Diagnostic Interview-Revised and dedicated cognitive assessments. However, it is essential to note that high rates of verification of ASD from clinical reports (81, 82) and high concordance (>97%) with clinician best estimate diagnoses and with standardized instruments (83) have been shown across prior online studies collecting parent-reported diagnoses. It has also been demonstrated that parent-report of children's IQ strongly correlates with standardized clinical IQ testing [e.g., (84)]. In addition, given that the current study relied on parent-reported clinical information only to

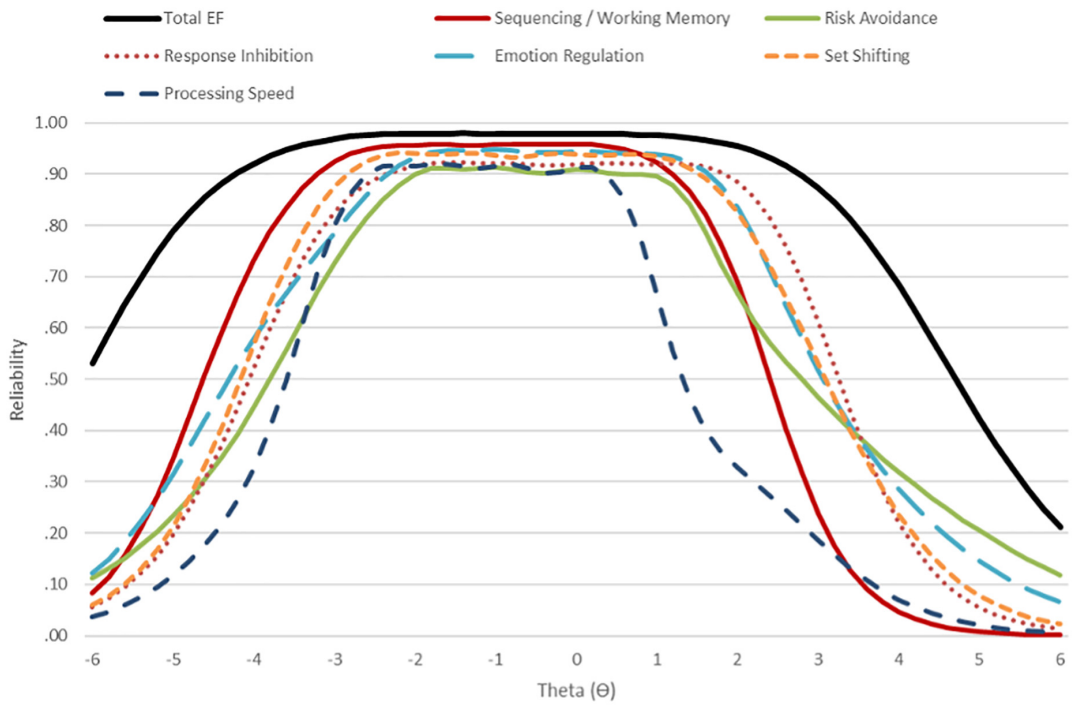


FIGURE 2
Conditional reliability for the Executive Functioning Scale (EFS) total scale and subscales.

TABLE 4 Executive Functioning Scale (EFS) convergent and discriminant validity.

	ADHD-ASSESS	DLS	CAS	BRIEF-sf
EFS total score	−0.76**	0.59**	−0.49**	−0.85**
EFS sequencing/Working memory	−0.61**	0.63**	−0.34**	−0.66**
EFS risk avoidance	−0.54**	0.38**	−0.20**	−0.52**
EFS response inhibition	−0.63**	0.61**	−0.29**	−0.66**
EFS emotion regulation	−0.64**	0.35**	−0.60**	−0.80**
EFS set shifting	−0.74**	0.47**	−0.51**	−0.82**
EFS processing speed	−0.52**	0.28**	−0.42**	−0.79**

*$p < 0.01$; **$p < 0.001$; ADHS-ASSESS, attention-deficit/hyperactivity disorder assessment; BRIEF-sf, Behavior Rating Inventory of Executive Functioning, second edition short form; CAS, Comprehensive Anxiety Scale; DLSS, Daily Living Skills Scale; EFS, Executive Functioning Scale.

describe the sample, we believe that these variables are an adequate proxy at the psychometric evaluation stage. However, it will be important for future studies to further evaluate the factor structure and psychometric properties of the EFS in large samples of individuals with discrete categorically defined neurodevelopmental and neuropsychiatric diagnoses established based on the gold-standard diagnostic instruments and clinical consensus. The present study was further limited by the lack of a more comprehensive set of questionnaires and performance-based EF assessments and by the relatively small sample of individuals with ASD. Thus, given the described limitations, it will be crucial for future studies to further validate the EFS in clinical settings conducive to detailed in-person observational and performance-based assessments and, ideally, utilize longitudinal designs to explore the predictive validity of the EFS. Finally, given dynamic and non-linear changes in the manifestation and complexity of specific facets of EF across different stages of development, it will be important for future studies to provide more detailed testing of the EFS performance across different periods of development, in particular during first 5 years of life, and, where relevant, develop further items to capture developmentally sensitive and specific instances and manifestations of the EF. Although not a limitation *per se*, it is important to highlight the fact that EFS captures several constructs, including emotion regulation and processing speed, that are not included in all of the existing EF models. However, given that there is no universally accepted EF taxonomy and

given the high clinical relevance of noted constructs across a range of neurodevelopmental and neuropsychiatric conditions, EFS was designed to provide a comprehensive capture of a broader range of EF-related constructs.

In summary, despite the noted limitations, the present data provide preliminary evidence that the EFS is a free, relatively brief, open-source, valid, and reliable measure for the comprehensive characterization of distinct subdomains of executive functioning that are relevant for the understanding of individual differences in clinical outcomes across a range of NDD and NPD. Further, EFS shows excellent measurement precision for capturing a wide range of abilities, which suggests a tremendous potential for its use for treatment tracking. Thus, with further replication, the EFS has excellent potential for wide adoption across research and clinical contexts.

Data availability statement

The raw data supporting the conclusions of this article will be made available by the authors, without undue reservation.

Ethics statement

This studies involving human participants were reviewed and approved by John Carroll University Institutional Review Board. Written informed consent to participate in this study was provided by the participants' legal guardian/next of kin.

Author contributions

TF, MU, and AH designed the study. TF and MU collected the data, had full access to the data, and conducted the analyses. MU, TF, RC, and AH drafted the initial manuscript. All authors critically reviewed and provided the feedback on the initial version of manuscript and approved the final version of the manuscript.

Funding

The study was supported by Simons Foundation Autism Research Initiative–Director's Award #831500 (PI: TF) and PTEN Research Foundation Grants #JCU-20-001 (PI: TF) and #UOM-20-001 (PI: MU).

Acknowledgments

We are sincerely indebted to the generosity of the participants who contributed their time and effort to this study. We would also like to thank the PTEN Research Foundation for their continued support.

Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

Publisher's note

References

1. Barkley RA. The assessment of executive functioning using the Barkley deficits in executive functioning scales. In: Goldstein S, Naglieri JA editors. *Handbook of Executive Functioning*. Berlin: Springer Science + Business Media (2014). p. 245–63. doi: 10.1007/978-1-4614-8106-5_15

2. Diamond A. Executive functions. *Annu Rev Psychol.* (2013) 64:135–68. doi: 10.1146/annurev-psych-113011-143750

3. Gross JJ. Emotion regulation: current status and future prospects. *Psychol Inquiry.* (2015) 26:1–26. doi: 10.1080/1047840X.2014.940781

4. Kochanska G, Murray K, Coy KC. Inhibitory control as a contributor to conscience in childhood: from toddler to early school age. *Child Dev.* (1997) 68:263–77. doi: 10.1111/j.1467-8624.1997.tb01939.x

5. Rothbart MK, Ellis LK, Posner MI. Temperament and self-regulation. In: Vohs KD, Baumeister RF editors. *Handbook of Self-Regulation: Research, Theory, and Applications*. New York, NY: The Guilford Press (2011). p. 441–60.

6. Tangney JP, Baumeister RF, Boone AL. High self-control predicts good adjustment, less pathology, better grades, and interpersonal success. *J Pers.* (2004) 72:271–324. doi: 10.1111/j.0022-3506.2004.00263.x

7. Duncan GJ, Dowsett CJ, Claessens A, Magnuson K, Huston AC, Klebanov P, et al. School readiness and later achievement. *Dev Psychol.* (2007) 43:1428–46. doi: 10.1037/0012-1649.43.6.1428

8. Baker S, Morawska A, Mitchell A. Promoting children's healthy habits through self-regulation via parenting. *Clin Child Family Psychol Rev.* (2019) 22:52–62. doi: 10.1007/s10567-019-00280-6

9. Davis JC, Marra CA, Najafzadeh M, Liu-Ambrose T. The independent contribution of executive functions to health related quality of life in older women. *BMC Geriatr.* (2010) 10:16. doi: 10.1186/1471-2318-10-16

10. Cai RY, Hardan AY, Phillips JM, Frazier TW, Uljarević M. Brief report: emotion regulation influences on internalizing and externalizing symptoms across

the normative-clinical continuum. *Front Psychiatry*. (2021) 12:693570. doi: 10.3389/fpsyt.2021.693570

11. Etkin A, Gyurak A, O'Hara R. A neurobiological approach to the cognitive deficits of psychiatric disorders. *Dialog Clin Neurosci.* (2013) 15:419–29. doi: 10.31887/DCNS.2013.15.4/aetkin

12. Goschke T. Dysfunctions of decision-making and cognitive control as transdiagnostic mechanisms of mental disorders: advances, gaps, and needs in current research. *Int J Methods Psychiatr Res.* (2014) 23(Suppl. 1):41–57. doi: 10.1002/mpr.1410

13. McTeague LM, Goodkind MS, Etkin A. Transdiagnostic impairment in cognitive control neurocircuits: behaviour, structure, and function. In: Egner T editor. *The Wiley Handbook of Cognitive Control*. New York, NY: Wiley Blackwell (2017). p. 539–55. doi: 10.1002/9781118920497.ch30

14. Snyder HR, Miyake A, Hankin BL. Advancing understanding of executive function impairments and psychopathology: bridging the gap between clinical and cognitive approaches. *Front. Psychol.* (2015) 6:328. doi: 10.3389/fpsyg.2015.00328

15. Cai RY, Richdale AL, Uljarević M, Dissanayake C, Samson AC. Emotion regulation in autism spectrum disorder: where we are and where we need to go. *Autism Res.* (2018) 11:962–78. doi: 10.1002/aur.1968

16. Wallace GL, Yerys BE, Peng C, Dlugi C, Anthony LG, Kenworthy L. Chapter three – assessment and treatment of executive function impairments in autism spectrum disorder: an update. *Int Rev Res Dev Disabil.* (2016) 51:85–122. doi: 10.1016/bs.irrdd.2016.07.004

17. Alderson RM, Kasper LJ, Hudec KL, Patros CH. Attention-deficit/hyperactivity disorder (ADHD) and working memory in adults: a meta-analytic review. *Neuropsychology.* (2013) 27:287–302. doi: 10.1037/a0032371

18. Faraone SV, Rostain AL, Blader J, Busch B, Childress AC, Connor DF, et al. Practitioner review: emotional dysregulation in attention-deficit/hyperactivity disorder – implications for clinical recognition and intervention. *J Child Psychol Psychiatry Allied Discip.* (2019) 60:133–50. doi: 10.1111/jcpp.12899

19. Forbes NF, Carrick LA, McIntosh AM, Lawrie SM. Working memory in schizophrenia: a meta-analysis. *Psychol Med.* (2009) 39:889–905. doi: 10.1017/S0033291708004558

20. Stefanopoulou E, Manoharan A, Landau S, Geddes JR, Goodwin G, Frangou S. Cognitive functioning in patients with affective disorders and schizophrenia: a meta-analysis. *Int Rev Psychiatry.* (2009) 21:336–56. doi: 10.1080/09540260902962149

21. Rock PL, Roiser JP, Riedel WJ, Blackwell AD. Cognitive impairment in depression: a systematic review and meta-analysis. *Psychol Med.* (2014) 44:2029–40. doi: 10.1017/S0033291713002535

22. Snyder HR. Major depressive disorder is associated with broad impairments on neuropsychological measures of executive function: a meta-analysis and review. *Psychol Bull.* (2013) 139:81–132. doi: 10.1037/a0028727

23. Abramovitch A, Abramowitz JS, Mittelman A. The neuropsychology of adult obsessive-compulsive disorder: a meta-analysis. *Clin Psychol Rev.* (2013) 33:1163–71. doi: 10.1016/j.cpr.2013.09.004

24. Shin NY, Lee TY, Kim E, Kwon JS. Cognitive functioning in obsessive-compulsive disorder: a meta-analysis. *Psychol Med.* (2014) 44:1121–30. doi: 10.1017/S0033291713001803

25. Polak AR, Witteveen AB, Reitsma JB, Olff M. The role of executive function in posttraumatic stress disorder: a systematic review. *J Affect Disord.* (2012) 141:11–21. doi: 10.1016/j.jad.2012.01.001

26. Hollocks MJ, Jones CR, Pickles A, Baird G, Happé F, Charman T, et al. The association between social cognition and executive functioning and symptoms of anxiety and depression in adolescents with autism spectrum disorders. *Autism Res.* (2014) 7:216–28. doi: 10.1002/aur.1361

27. Uljarević M, Richdale AL, Evans DW, Cai RY, Leekam SR. Interrelationship between insistence on sameness, effortful control and anxiety in adolescents and young adults with autism spectrum disorder (ASD). *Mol Autism.* (2017) 8:36. doi: 10.1186/s13229-017-0158-4

28. Uljarević M, Frazier TW, Rached G, Busch RM, Klaas P, Srivastava S, et al. Brief report: role of parent-reported executive functioning and anxiety in insistence on sameness in individuals with Germline PTEN mutations. *J Autism Dev Disord.* (2022) 52:414–22. doi: 10.1007/s10803-021-04881-5

29. Leung RC, Vogan VM, Powell TL, Anagnostou E, Taylor MJ. The role of executive functions in social impairment in autism spectrum disorder. *Child Neuropsychol.* (2016) 22:336–44. doi: 10.1080/09297049.2015.1005066

30. Lieb RW, Bohnert AM. Relations between executive functions, social impairment, and friendship quality on adjustment among high functioning youth with autism spectrum disorder. *J Autism Dev Disord.* (2017) 47:2861–72. doi: 10.1007/s10803-017-3205-2

31. Horne CM, Sahni A, Pang SW, Vanes LD, Szentgyorgyi T, Averbeck B, et al. The role of cognitive control in the positive symptoms of psychosis. *Neuroimage Clin.* (2022) 34:103004. doi: 10.1016/j.nicl.2022.103004

32. Orellana G, Slachevsky A. Executive functioning in schizophrenia. *Front Psychiatry.* (2013) 4:35. doi: 10.3389/fpsyt.2013.00035

33. Ruiz-Castañeda P, Santiago-Molina E, Aguirre-Loaiza H, Daza González MT. "Cool" and "Hot" executive functions in patients with a predominance of negative schizophrenic symptoms. *Front Psychol.* (2020) 11:571271. doi: 10.3389/fpsyg.2020.571271

34. Maddox BB, Cleary P, Kuschner ES, Miller JS, Armour AC, Guy L, et al. Lagging skills contribute to challenging behaviors in children with autism spectrum disorder without intellectual disability. *Autism.* (2018) 22:898–906. doi: 10.1177/1362361317712651

35. Miyake A, Friedman NP. The nature and organization of individual differences in executive functions: four general conclusions. *Curr Dir Psychol Sci.* (2012) 21:8–14. doi: 10.1177/0963721411429458

36. Goldstein S, Naglieri JA, Princiotta D, Otero TM. *Introduction: A History of Executive Functioning as a Theoretical and Clinical Construct. In: Handbook of Executive Functioning*. New York, NY: Springer (2014). p. 3–12. doi: 10.1007/978-1-4614-8106-5_1

37. Zelazo PD, Carlson SM. Hot and cool executive function in childhood and adolescence: development and plasticity. *Child Dev Perspect.* (2012) 6:354–60. doi: 10.1111/j.1750-8606.2012.00246.x

38. Gross JJ, Jazaieri H. Emotion, emotion regulation, and psychopathology: an affective science perspective. *Clin Psychol Sci.* (2014) 2:387–401. doi: 10.1177/2167702614536164

39. Casey BJ. Beyond simple models of self-control to circuit-based accounts of adolescent behavior. *Annu Rev Psychol.* (2015) 66:295–319. doi: 10.1146/annurev-psych-010814-015156

40. Ernst M. The triadic model perspective for the study of adolescent motivated behavior. *Brain Cogn.* (2014) 89:104–11. doi: 10.1016/j.bandc.2014.01.006

41. Nigg JT. 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. *J Child Psychol Psychiatry Allied Discip.* (2017) 58:361–83. doi: 10.1111/jcpp.12675

42. McGrath LM, Braaten EB, Doty ND, Willoughby BL, Wilson HK, O'Donnell EH, et al. Extending the 'cross-disorder' relevance of executive functions to dimensional neuropsychiatric traits in youth. *J Child Psychol Psychiatry.* (2015) 57:462–71. doi: 10.1111/jcpp.12463

43. Nigg JT, Jester JM, Stavro GM, Ip KI, Puttler LI, Zucker RA. Specificity of executive functioning and processing speed problems in common psychopathology. *Neuropsychology.* (2017) 31:448–66. doi: 10.1037/neu0000343

44. Gioia GA, Isquith PK, Guy SC, Kenworthy L. *Behavior Rating Inventory of Executive Function®, Second Edition (BRIEF® 2)*. Lutz, FL: PAR Inc (2015).

45. Naglieri JA, Goldstein S. Using the comprehensive executive function inventory (CEFI) to assess executive function: from theory to application. In: Goldstein S, Naglieri J editors. *Handbook of Executive Functioning*. New York, NY: Springer (2014). doi: 10.1007/978-1-4614-8106-5_14

46. Barkley RA. *Barkley Deficits in Executive Functioning Scale (BDEFS for Adults)*. New York, NY: Guilford Press (2011).

47. Barkley RA, Murphy KR. The nature of executive function (EF) deficits in daily life activities in adults with ADHD and their relationship to performance on EF tests. *J Psychopathol Behav Assess.* (2011) 33:137–58. doi: 10.1007/s10862-011-9217-x

48. Barkley RA. *Barkley Deficits in Executive Functioning Scale–Children and Adolescents (BDEFS-CA)*. New York, NY: Guilford Press (2012).

49. Delis DC, Kaplan E, Kramer JH. *Delis-Kaplan Executive Function System (D–KEFS) [Database Record]*. San Antonio, TX: APA PsycTests. (2001). doi: 10.1037/t15082-000

50. Cambridge Cognition. *Neuropsychological Test Automated Battery (CANTABeclipse) Manual*. Cambridge: Cambridge Cognition Limited (2006).

51. Korkman M, Kirk U, Kemp S. *NEPSY-II*. 2nd ed. San Antonio, TX: Harcourt Assessment (2007).

52. Zelazo PD, Anderson JE, Richler J, Wallner-Allen K, Beaumont JL, Conway KP, et al. NIH toolbox cognition battery (CB): validation of executive function measures in adults. *J Int Neuropsychol Soc.* (2014) 20:620–9. doi: 10.1017/S1355617714000472

53. Fernández-Alcántara M, Albaladejo-Blázquez N, Fernández-Ávalos MI, Sánchez-SanSegundo M, Cruz-Quintana F, Pérez-Martínez V, et al. Validity of the computerized battery for neuropsychological evaluation of children (BENCI) in

Spanish children: preliminary results. *Eur J Investig Health Psychol Educ.* (2022) 12:893–903. doi: 10.3390/ejihpe12080065

54. Eisenberg N, Spinrad TL, Eggum ND. Emotion-related self-regulation and its relation to children's maladjustment. *Annu Rev Clin Psychol.* (2010) 6:495–525. doi: 10.1146/annurev.clinpsy.121208.131208

55. Carl JR, Soskin DP, Kerns C, Barlow DH. Positive emotion regulation in emotional disorders: a theoretical review. *Clin Psychol Rev.* (2013) 33:343–60.

56. Gross JJ, John OP. Individual differences in two emotion regulation processes: implications for affect, relationships, and well-being. *J Pers Soc Psychol.* (2003) 85:348–62. doi: 10.1037/0022-3514.85.2.348

57. Ng TH, Stange JP, Black CL, Titone MK, Weiss RB, Abramson LY, et al. Impulsivity predicts the onset of DSM-IV-TR or RDC hypomanic and manic episodes in adolescents and young adults with high or moderate reward sensitivity. *J Affect Disord.* (2016) 198:88–95. doi: 10.1016/j.jad.2016.03.045

58. Jimenez A, Lucas-Molina B. Dimensional structure and measurement invariance of the BRIEF-2 across gender in a socially vulnerable sample of primary school-aged children. *Child Neuropsychol.* (2019) 25:636–47. doi: 10.1080/09297049.2018.1512962

59. Marjanovic Z, Bajkov L, MacDonald J. The conscientious responders scale helps researchers verify the integrity of personality questionnaire data. *Psychol Rep.* (2019) 122:1529–49. doi: 10.1177/0033294118783917

60. Reynolds CR, Kamphaus RW. *Behavior Assessment System for Children*. 3rd ed. London: Pearson (2015). doi: 10.1002/9781118625392.wbecp447

61. Briggs-Gowan MJ, Carter AS, Irwin JR, Wachtel K, Cicchetti DV. The brief infant-toddler social and emotional assessment: screening for social-emotional problems and delays in competence. *J Pediatr Psychol.* (2004) 29:143–55. doi: 10.1093/jpepsy/jsh017

62. Gartstein MA, Rothbart MK. Studying infant temperament via the revised infant behavior questionnaire. *Infant Behav Dev.* (2003) 26:64–86. doi: 10.1016/S0163-6383(02)00169-8

63. Shields A, Cicchetti D. *Emotion Regulation Checklist*. Indianapolis, IN: APA PsycTests (1995). doi: 10.1037/t08132-000

64. Gratz KL, Roemer L. Multidimensional assessment of emotion regulation and dysregulation: development, factor structure, and initial validation of the difficulties in emotion regulation scale. *J Psychopathol Behav Assess.* (2004) 26:41–54. doi: 10.1023/B:JOBA.0000007455.08539.94

65. LeJeune B, Beebe D, Noll J, Kenealy L, Isquith P, Gioia G. Psychometric support for an abbreviated version of the behavior rating inventory of executive function (BRIEF) parent form. *Child Neuropsychol.* (2010) 16:182–201. doi: 10.1080/09297040903352556

66. Spackman E, Uljarević M, Hardan AY, Frazier TW. Characterizing attention-deficit/hyperactivity disorder, anxiety and mood symptoms in youth with autism and other neurodevelopmental and neuropsychiatric conditions: development and validation of new assessment battery. In: *Poster Presented at the San Francisco Bay Area Conference*. San Francisco, CA (2022).

67. Frazier TW, Spackman E, Hardan AY, Uljarević M. Development and validation of a new open source measure of daily living skills. In: *Poster Presented at the San Francisco Bay Area Conference*. San Francisco, CA (2022).

68. Marsh HW, Morin AJ, Parker PD, Kaur G. Exploratory structural equation modeling: an integration of the best features of exploratory and confirmatory factor analysis. *Annu Rev Clin Psychol.* (2014) 10:85–110. doi: 10.1146/annurev-clinpsy-032813-153700

69. Hu L, Bentler PM. Cutoff criteria for fit indexes in covariance structure analysis: conventional criteria versus new alternatives. *Struct Equ Model.* (1999) 6:1–55. doi: 10.1080/10705519909540118

70. Marsh HW, Hau K-T, Wen Z. In search of golden rules: comment on hypothesis-testing approaches to setting cutoff values for fit indexes and dangers in overgeneralizing Hu and Bentler's (1999) findings. *Struct Equ Model.* (2004) 11:320–41. doi: 10.1207/s15328007sem1103_2

71. Chiorri C, Marsh HW, Ubbiali A, Donati D. Testing the factor structure and measurement invariance across gender of the big five inventory through exploratory structural equation modeling. *J Pers Assess.* (2016) 98:88–99. doi: 10.1080/00223891.2015.1035381

72. Putnick DL, Bornstein MH. Measurement invariance conventions and reporting: the state of the art and future directions for psychological research. *Dev Rev.* (2016) 41:71–90. doi: 10.1016/j.dr.2016.06.004

73. Frazier TW, Hardan AY. Equivalence of symptom dimensions in females and males with autism. *Autism.* (2017) 21:749–59. doi: 10.1177/1362361316660066

74. Chen FF. Sensitivity of goodness of fit indexes to lack of measurement invariance. *Struct Equ Model.* (2007) 14:464–504. doi: 10.1080/10705510701301834

75. Cheung GW, Rensvold RB. Evaluating goodness-of-fit indexes for testing measurement invariance. *Struct Equ Model.* (2002) 9:233–55. doi: 10.1207/S15328007SEM0902_5

76. Streiner D, Norman G. *Health Measurement Scales: a Practical Guide to Their Development and Use*. 2nd ed. Oxford: Oxford University Press (1995).

77. Embretson SE, Reise SP. *Item Response Theory for Psychologists*. Mahwah, NJ: Lawrence Erlbaum Associates Publishers (2000).

78. Nunnally JC, Bernstein IH. The assessment of reliability. *Psychometr Theor.* (1994) 3:248–92.

79. Collins A, Koechlin E. Reasoning, learning, and creativity: frontal lobe function and human decision-making. *PLoS Biol.* (2012) 10:e1001293. doi: 10.1371/journal.pbio.1001293

80. Lunt L, Bramham J, Morris RG, Bullock PR, Selway RP, Xenitidis K, et al. Prefrontal cortex dysfunction and 'Jumping to Conclusions': bias or deficit? *J Neuropsychol.* (2012) 6:65–78. doi: 10.1111/j.1748-6653.2011.02005.x

81. Daniels AM, Rosenberg RE, Anderson C, Law JK, Marvin AR, Law PA. Verification of parent-report of child autism spectrum disorder diagnosis to a web-based autism registry. *J Autism Dev Disord.* (2012) 42:257–65. doi: 10.1007/s10803-011-1236-7

82. Feliciano P, Zhou X, Astrovskaya I, Turner TN, Wang T, Brueggeman L, et al. Exome sequencing of 457 autism families recruited online provides evidence for autism risk genes. *NPJ Genom Med.* (2019) 4:19. doi: 10.1038/s41525-019-0093-8

83. Lee H, Marvin AR, Watson T, Piggot J, Law JK, Law PA, et al. Accuracy of phenotyping of autistic children based on Internet implemented parent report. *Am J Med Genet B Neuropsychiatr Genet.* (2010) 153B:1119–26. doi: 10.1002/ajmg.b.31103

84. Shu C, Green Snyder L, Shen Y, Chung WK, SPARK Consortium. Imputing cognitive impairment in SPARK, a large autism cohort. *Autism Res.* (2022) 15:156–70. doi: 10.1002/aur.2622

frontiers | Frontiers in Psychiatry

TYPE Original Research
PUBLISHED 01 February 2023
DOI 10.3389/fpsyt.2023.1098210

OPEN ACCESS

EDITED BY
Andrea C. Samson,
UniDistance Suisse, Switzerland

REVIEWED BY
Yael Enav,
University of Haifa, Israel
Andreia P. Costa,
University of Luxembourg, Luxembourg

*CORRESPONDENCE
Amélia Walter
✉ amelia.walter@etu.unistra.fr

SPECIALTY SECTION
This article was submitted to Autism, a section of the journal Frontiers in Psychiatry

RECEIVED 14 November 2022
ACCEPTED 09 January 2023
PUBLISHED 01 February 2023

CITATION
Walter A, Martz E, Weibel S and Weiner L (2023) Tackling emotional processing in adults with attention deficit hyperactivity disorder and attention deficit hyperactivity disorder + autism spectrum disorder using emotional and action verbal fluency tasks. *Front. Psychiatry* 14:1098210. doi: 10.3389/fpsyt.2023.1098210

Tackling emotional processing in adults with attention deficit hyperactivity disorder and attention deficit hyperactivity disorder + autism spectrum disorder using emotional and action verbal fluency tasks

Amélia Walter[1]*, Emilie Martz[2], Sébastien Weibel[2,3] and Luisa Weiner[3,4]

[1]Institut des Neurosciences Cellulaires et Intégratives, Centre National de la Recherche Scientifique (UPR 3212), Strasbourg University, Strasbourg, France, [2]Institut National de la Santé et de la Recherche Médicale U1114, Strasbourg, France, [3]Department of Psychiatry, University Hospital of Strasbourg, Strasbourg, France, [4]Laboratoire de Psychologie des Cognitions, University of Strasbourg, Strasbourg, France

Introduction: Attention Deficit Hyperactivity Disorder (ADHD) and Autism Spectrum Disorder (ASD) are two neurodevelopmental conditions with neuropsychological, social, emotional, and psychopathological similarities. Both are characterized by executive dysfunction, emotion dysregulation (ED), and psychiatric comorbidities. By focusing on emotions and embodied cognition, this study aims to improve the understanding of overlapping symptoms between ADHD and ASD through the use of verbal fluency tasks.

Methods: Fifty-two adults with ADHD, 13 adults with ADHD + ASD and 24 neurotypical (NT) participants were recruited in this study. A neuropsychological evaluation, including different verbal fluency conditions (e.g. emotional and action), was proposed. Subjects also completed several self-report questionnaires, such as scales measuring symptoms of ED.

Results: Compared to NT controls, adults with ADHD + ASD produced fewer anger-related emotions. Symptoms of emotion dysregulation were associated with an increased number of actions verbs and emotions produced in ADHD.

Discussion: The association between affective language of adults with ADHD and symptoms of emotion dysregulation may reflect their social maladjustment. Moreover, the addition of ADHD + ASD conditions may reflect more severe affective dysfunction.

KEYWORDS

emotion dysregulation, verbal fluency, ADHD, ASD, neurodevelopmental disorders, executive functions

1. Introduction

Neurodevelopmental disorders affect 5–15% of children (1, 2) and often persist over the lifetime with significant impact on adaptive, academic, and social functioning (3). Attention Deficit Hyperactivity Disorder (ADHD) and Autism Spectrum Disorder (ASD) in particular are among the most common neurodevelopmental disorders (4).

Attention deficit hyperactivity disorder is characterized by a triad of symptoms: i.e., inattention, hyperactivity, and impulsivity. In children, the prevalence is estimated between 7.2 and 9.4% (5). Moreover, ADHD symptoms persist in adulthood in two-thirds of them (6, 7), as 4% of adults have ADHD (8–10). Nevertheless, the clinical presentation of ADHD in adults is different: i.e., motor hyperactivity symptoms are internalized in favor of mental hyperactivity, and attentional difficulties persist or even increase (11–14). In addition to these cognitive symptoms, adults with ADHD often report emotional difficulties. Indeed, emotion dysregulation (ED) concerns 30–70% of adults with ADHD (15). Emotion dysregulation can be defined by three dimensions: affect control, affective lability, and emotional over-reactivity (16–18). These symptoms can, respectively be reflected by irritability, frequent emotional fluctuations and greater emotional sensitivity. Importantly, neurodevelopmental and psychiatric comorbidities associated with ADHD are in part related to emotion dysregulation and its functional impact (19). Indeed, up to 75% of adults with ADHD have an associated psychiatric disorder (20), including depression in half the cases, anxiety disorders, bipolar disorder, and personality disorders (21). ADHD is also often associated with learning disabilities (22) and one in eight children have a co-occurring ASD (23).

Autism spectrum disorder is characterized by social communication peculiarities and the presence of restricted, repetitive behaviors (3). ASD affects approximately 1% of the world's population (24). Despite obvious differences between diagnostic criteria of ADHD and ASD, the differential diagnosis can be challenging. Indeed, both conditions are characterized by attention difficulties (25, 26), disturbances in social interactions (27, 28), and abnormal sensorial sensitivity (29). Furthermore, emotion dysregulation is also common among autistic individuals, affecting up to 80% of them (30). No study to date has accurately described the facets of emotion dysregulation in adults with autism. That said, several studies highlight irritability, aggression, self-injury, impulsivity (31, 32) as manifestations of emotion dysregulation in children. Other studies have pointed out strong links between emotion dysregulation and other ASD symptoms such as anxiety (33), cognitive rigidity (34), and repetitive behaviors (35).

From a neuropsychological standpoint, emotional regulation is defined as the process of initiating, avoiding, inhibiting, maintaining, or modulating emotional states to accomplish individual goals or social adaptation (36). In neurodevelopmental conditions, emotion dysregulation is mainly explained by executive dysfunction (37, 38). Executive functions refer to a set of high-level cognitive processes that enable goal-directed behavior and problem solving (39). Executive functions can be classified into two categories: "cold" executive functions, such as mental flexibility, inhibition, and working memory, and "hot" executive functions, associated with the processing of social-emotional components (40, 41). These components include emotion regulation, empathy, rewards processing, and social adaptation (36, 37). Most studies focusing on the neuropsychological mechanisms of emotion dysregulation in neurodevelopmental disorders emphasize the involvement of cold executive functions. As an example, Barkley's ADHD model (37) postulates that inhibition impairments cause emotional impulsivity (37, 42). In ADHD, emotion dysregulation also results from an inability to engage in self-regulatory actions, such as the ability to refocus attention (42). In ASD, inhibition and mental flexibility deficits are thought to result in perseverative behaviors, which in turn are related to maladaptive emotion regulation strategies, such as excessive rumination (43–46). When ADHD co-occurs with ASD, studies in children show that maladaptive emotion regulation strategies are exacerbated by ADHD symptoms (46). Therefore, the combination of these two conditions is characterized by more severe social and adaptive dysfunction compared to ADHD or ASD alone (47–49).

One way to study cold and hot executive functions is to use verbal fluency tasks (VFT). VFT, particularly letter and semantic conditions (i.e., generation of a maximum of words based on a letter or a semantic cue during a given time), have long been used to assess the executive and language functioning of patients (e.g., 50). VFT tap on lexical-semantic language processes (51), mental flexibility, inhibition, and information processing speed (52). These same processes are involved in the generation of emotional words (53). While the letter and semantic conditions preferentially target cold executive functions and semantic memory, respectively, the Emotional Word Fluency Test (EWFT), developed by Abeare et al. (54), assess the affective component of word production, i.e., affective language. Affective language corresponds to the production and comprehension of words or sentences with affective valence, but also emotional intonation and prosody (54). The EWFT consists of producing the maximum of emotion nouns during an allotted time (54). Disturbances in affective language have been found in several clinical populations, including individuals with right hemisphere brain damage (55, 56), depressive disorder (57), schizophrenia (58), or ASD (59). Nevertheless, in clinical settings, the EWFT has only been used in studies with individuals with traumatic brain injury (60). Using the EWFT with healthy subjects, Abeare et al. (61) found that trait anxiety was positively correlated with the number of emotions with negative valence produced. Furthermore, they showed that the physiological response of the sympathetic nervous system, e.g., skin conductance, was higher during emotional word generation compared to a control task, i.e., semantic VFT (i.e., "animal names").

Typically, performances on VFT are assessed by the number of words generated, rule breaks, and repetitions (62). However, these quantitative measures provide little information on the cognitive mechanisms (i.e., language or executive) involved in word output. Several qualitative methods have therefore been developed to identify the cognitive processes involved in spontaneous word generation. For example, Troyer et al. (63) investigated the involvement of lexical-semantic processing and executive functions in VFT, *via* clustering and switching. Clustering corresponds to the production of words within a semantic or phonemic category, while switching reflects the ability to shift efficiently to a new category. Moreover, VFT are also influenced by parameters of imaginability, and concreteness, as well as linguistic parameters (e.g., lexical frequency). Indeed, access to the lexical stock is facilitated for more frequent and concrete words (64, 65). This facilitation allows a faster speed of word production, and an increased number of words produced for a specific time duration (66).

Few studies have investigated VFT performance in people with ADHD and autistic adults, and none have focused on affective language. For instance, adults with ADHD have been shown to produce fewer words than NT controls in semantic VFT (67, 68), and especially in letter conditions (69, 70). Since the letter conditions rely more on executive functions (71), these findings have been explained by the executive dysfunction usually associated with ADHD. In contrast, other studies found no deficits in VFT (72, 73). Therefore, the results are overall inconsistent in ADHD, probably due to the fact

that only the number of words and errors were measured in most studies. In studies focusing on process-oriented measures of VFT (i.e., clustering and switching), some authors reported a decrease in the number of switches in the ADHD + ASD group population (68), while others have found the opposite pattern (73).

In ASD, studies have found reduced (74–76) or identical word count compared to control subjects in letter and semantic VFT (77–79). In contrast, the process-oriented measures of VFT, including clustering, switching, imaginability, and concreteness, did not differ from the NT controls (75, 77). These discrepant findings may be due to the heterogeneity of ASD presentations (80, 81). Indeed, autistic adults without intellectual disability have been found to produce fewer words during the first thirty seconds of letter and semantic conditions of VFT, which could be explained by an initiation deficit (82). Autistic children, on the other hand, generate less prototypical exemplars of semantic categories than NT controls (83), which can be explained by semantic deficits involved in comprehension and expression difficulties (84–86).

Regarding the assessment of hot executive functions using VFT, in ASD, most studies have focused on emotion identification, but not on affective word generation. Yet, emotional language disturbances involve both language comprehension and production (59). Autistic individuals produce sentences of greater duration, amplitude, and intensity than NT subjects (87). In addition, difficulties in identifying and describing one's own feelings, i.e., alexithymia, are common in ASD (88). These difficulties are associated with the production of fewer emotional words when subjects are talking about negative events (89). Moreover, autistic adults assign less nuanced intensity and valence to stimuli compared to NT subjects (90, 91). For example, positive and negative images or words are evaluated less positively and negatively, respectively, than control subjects (92).

In ADHD, reduced abilities to recognize facial, vocal emotions (93, 94), and contextual information (95) have been reported (27, 96). In a study comparing children with ASD, ADHD, and ASD + ADHD, Tye et al. (97) found atypical processing of facial emotions for all the three groups. Specifically, autistic children displayed reduced neurophysiological response to angry faces in early stages of information processing. In contrast, children with ADHD displayed electrophysiological abnormality at the contextual processing stage when they were confronted with the emotions of fear and joy. The ADHD + ASD group presented an additive effect of the unique deficits of both conditions. Therefore, emotion processing seems to differ between the two groups, with autistic individuals presenting more difficulties at the early stages of processing of anger, whereas children with ADHD have difficulties at later stages of processing of fear and joy.

In addition to EWFT, other VFT involving embodied cognition may provide additional information about emotional processing in ADHD and ASD. Embodied cognition refers to the relationship between cognitive and bodily experience, which can be tackled *via* the VFT condition of action words generation (98, 99). In this task condition, subjects have to produce as many names of action verbs as possible during an allotted time. Whereas other VFT (i.e., semantic and letter) involve mainly temporo-parietal regions (100), Action Verb Fluency Test (AVFT) is sensitive to the activation of left fronto-striatal circuits (101). Fronto-striatal circuits are involved in "hot" executive functions such as rewards processing, emotion regulation and motivational states (102), and dysfunction of these circuits is involved in the cognitive and motivational control impairment found in ADHD (103). Relatedly, the same regions are thought to be involved in the difficulties in predicting emotions and actions of others in ASD (104). When using AVFT with autistic adults, Inokuchi and Kamio (105) found a reduced production of semantic clusters and an increased production of phonological clusters compared to control subjects, which they attributed to initiation difficulties due to executive dysfunction (106). However, AVFT performance has not been investigated in ADHD.

To our knowledge, no study has compared the performance on the EWFT and the AVFT in adults with ADHD and ADHD + ASD. Yet, it is of the upmost importance to better understand the overlap between these two conditions, especially regarding emotional processing, given its impact on quality of life and social functioning. The present study aims to evaluate several types of VFT, including EWFT and AVFT, in adults with ADHD, ASD + ADHD, and NT controls. We are particularly interested in the relationship between emotion dysregulation and verbal fluency tasks performance on EWFT and AVFT in ADHD. To this end, the number of words and errors produced, but also the type, the valence, the arousal, and the dominance of words generated in the EWFT and the AVFT will be analyzed. Given that emotion dysregulation is found in ADHD and ASD, we hypothesize that affective language particularities, i.e., a reduction in the number of emotions produced on the EWFT, will be found both in ADHD, and ADHD + ASD compared to NT controls. However, since impairments at earlier stages of emotion identification are found in ASD (97) compared to ADHD (89, 93), we expect adults with ADHD + ASD to produce fewer emotions on the EWFT compared to adults with ADHD. Due to abnormalities in the contextual processing stages of specific emotions (97), we also hypothesize that the emotion generation difficulties of adults with ADHD will be related to joy and fear. In view of results from a previous study on co-occurring ADHD + ASD (97), this group may present with the emotional particularities of both conditions with emotion generation difficulties related to joy, fear and anger. Regarding AVFT, given the primary deficits in predicting, and interpreting the actions of others (107), and motor issues involving fronto-striatal circuits (108) in ASD, adults with ADHD + ASD are expected to produce fewer and rarer (low frequency in the language) action verbs than adults with ADHD and NT controls, whereas adults with ADHD will produce fewer action verbs than NT adults. Nevertheless, due to the hyperactivity found in ADHD, the action verbs produced are expected to be more arousing than those found in NT.

2. Materials and methods

2.1. Participants

Fifty-two adults with ADHD aged 18–57 (M = 34.75; SD = 11.49), and 13 adults with co-occurring ADHD + ASD aged 19–67 (M = 32.15; SD = 13.95) without intellectual disability, communication difficulties, nor difference in gender distribution (see **Table 1**) were recruited from the University Hospital of Strasbourg. We were also interested in a group of 12 autistic adults aged 18–48 (M = 26.92; SD = 8.35) from another study on emotion dysregulation conducted by our team. After diagnoses of ADHD and ASD were established by senior psychiatrists according to DSM-5 criteria (3), subjects were offered participation in one of the two studies. These

TABLE 1 Demographic, clinical and neuropsychological characteristics of participants in each group.

	ADHD (AD) (*N* = 52)	ADHD + ASD (AD + AS) (*N* = 13)	Neurotypical controls (NT) (*N* = 24)	Statistical analyses	*P*	*Post hoc*
Demographic characteristics						
Age	34.75 (11.49)	32.15 (13.95)	32.71 (9.45)	χ^2 (2) = 1.699	0.428	/
Gender	46.14% F	38.46% F	66.66% F	χ^2 (2) = 3.452	0.178	/
Years of education	13.61 (3.29)	15.09 (1.81)	15.37 (1.53)	χ^2 (2) = 5.764	0.056	/
IQ estimation	111.19 (14.15)	108.55 (15.65)	105.00 (9.52)	F (2,65) = 1.635	0.203	/
WAIS-III–vocabulary (SS)	12.12 (2.74)	13.00 (2.00)	10.71 (1.60)	χ^2 (2) = 6.250	**0.044**	AD + AS > NT
Clinical characteristics						
WRAADDS—inattention	17.71 (3.85)	17.17 (3.92)	5.74 (4.34)	F (2,61) = 63.991	<**0.001***	AD, AD + AS > NT
WRAADDS—hyperactivity	7.54 (3.24)	7.33 (1.86)	2.52 (2.39)	χ^2 (2) = 25.138	<**0.001***	AD, AD, AD + AS > NT
WRAADDS—impulsivity	12.31 (3.77)	10.50 (5.32)	4.00 (3.33)	χ^2 (2) = 32.145	<**0.001***	AD, AD + AS > NT
WRAADDS—affective Lability	11.69 (2.92)	11.33 (4.55)	6.00 (4.17)	χ^2 (2) = 22.063	<**0.001***	AD, AD + AS > NT
WRAADDS—emotional over-reactivity	12.80 (3.16)	13.33 (3.20)	5.13 (3.33)	χ^2 (2) = 34.122	<**0.001***	AD, AD + AS > NT
WRAADDS—social maladjustment	21.83 (8.42)	32.33 (8.71)	6.26 (6.67)	F (2,61) = 39.442	<**0.001***	AD + AS > AD > NT
Autism spectrum quotient	4.29 (1.79)	6.50 (2.37)	2.87 (2.32)	F (2,64) = 12.801	<**0.001***	AD + AS > AD > NT
BDI—total	19.80 (10.64)	29.92 (18.28)	6.37 (5.95)	χ^2 (2) = 25.780	<**0.001***	AD, AD + AS > NT
GAD-7—total	9.29 (4.54)	5.80 (6.22)	2.79 (1.76)	χ^2 (2) = 18.738	<**0.001***	AD > NT
TEMPS-A—cyclothymic temperament	22.28 (7.98)	17.67 (10.31)	7.35 (5.80)	F (2,60) = 27.276	<**0.001***	AD, AD + AS > NT
Neuropsychological characteristics						
VFT free (number of words)	69.03 (22.27)	54.91 (17.66)	73.54 (19.50)	F (2,64) = 1.698	0.191	/
VFT letter (number of words)	28.46 (6.95)	26.45 (7.02)	29.92 (6.91)	F (2,64) = 0.635	0.533	/
VFT semantic (number of words)	37.09 (8.04)	32.36 (8.22)	38.50 (6.28)	F (2,64) = 1.136	0.328	/
TMT Part B minus Part A (seconds)	36.52 (18.35)	24.82 (15.53)	30.41 (14.45)	χ^2 (2) = 2.479	0.290	/
Stroop interference minus reading (sec)	34.37 (14.27)	29.02 (7.03)	25.83 (8.39)	χ^2 (2) = 5.368	0.068	/
Digit span forward (number of digits)	6.31 (1.16)	5.83 (0.41)	6.67 (0.76)	χ^2 (2) = 4.533	0.104	/
Digit span backward (number of digits)	5.20 (1.43)	4.67 (0.52)	5.79 (1.47)	χ^2 (2) = 3.868	0.145	/
Hayling test inhibition (seconds)	60.04 (32.29)	57.47 (13.84)	63.73 (33.93)	χ^2 (2) = 0.240	0.887	/
Hayling test inhibition (errors)	6.94 (3.73)	6.33 (3.33)	4.42 (2.43)	χ^2 (2) = 6.708	**0.035***	AD > NT
TAP Go-Nogo 1 out of 2 (errors)	0.97 (0.98)	1.17 (1.94)	0.75 (0.94)	χ^2 (2) = 1.114	0.573	/
TAP Go-Nogo 2 out of 5 (errors)	0.34 (0.72)	0.17 (0.41)	0.04 (0.20)	χ^2 (2) = 3.925	0.140	/
ToL (total correct)	4.86 (2.38)	4.33 (2.73)	4.25 (2.64)	F (2,62) = 0.919	0.640	/
ToL (additional movements)	26.80 (17.96)	28.33 (14.83)	28.46 (18.23)	χ^2 (2) = 0.195	0.907	/
ToL (initation time)	73.30 (47.24)	56.16 (25.97)	61.29 (44.97)	χ^2 (2) = 1.698	0.428	/

*$p < 0.05$; Years of education, number of years after the first grade in elementary school; SS, scaled score. *p*-values in bold = significant *post hoc* comparisons.

diagnostic interviews also involve establishing a differential diagnosis between ASD and ADHD. Among the ADHD group, 78% of subjects had combined ADHD presentation, 20% of them had inattentive subtype, and the last 2% had hyperactive presentation. Subjects with psychopathological disorders, i.e., depressive disorders and anxiety, were excluded in the ADHD group but not in the ASD and ADHD + ASD group. Ten subjects from the ASD group had experienced one or more past depressive episodes. Four of them also suffered from anxiety disorders. In the ADHD + ASD group, four adults had depression, whereas three had an anxiety disorder. In addition, one participant had bipolar disorder. Twenty-four age-matched NT controls aged 20–46 (M = 32.71; SD = 9.45) were recruited. These participants did not have any history of neurological, psychiatric, or substance use disorders and did not use any psychotropic medication. Adults with ADHD and ADHD + ASD had significantly higher ADHD symptoms (measured by the Wender-Reimherr Adult Attention Deficit Disorder Scale; (16), compared to NT controls ($p < 0.001$; **Table 1**). In contrast, the Autism Spectrum Quotient score (AQ–10 items); (109) and social adjustment difficulties (WRAADDS); (16) were higher in the ADHD + ASD group compared to the ADHD group ($p = 0.011$; $p = 0.010$, respectively) and to NT subjects ($p < 0.001$). All participants

were native French speakers. This study was approved by ethics committees (CPP South Mediterranean II; IDRCB: 2017-A01618-45 and CPP East of France; No. SI 21.01.21.41923).

2.2. Materials and procedures

All subjects, with the exception of the ASD group, participated in a neuropsychological assessment and completed self-report questionnaires. Autistic adults only completed the VFT conditions.

2.2.1. Questionnaires

Attention deficit hyperactivity disorder and ASD symptoms were, respectively measured by the Wender-Reimherr Adult Attention Deficit Disorder Scale (WRAADDS); (16) and the Autism spectrum Quotient (AQ–10 items); (109). The WRAADDS is a self-reported scale assessing the three core symptoms of ADHD, some dimensions related to emotion dysregulation (e.g., affective lability and emotional over-reactivity), as well as other areas frequently disrupted in ADHD, such as organization, academic problems and social adjustment. The 10-item Autism Spectrum Quotient is a self-administered questionnaire used to assess traits of autism in adults without intellectual disability. Items are related to domains of the dyad of impairments, i.e., social communication peculiarities (e.g., I find it easy to "read between the lines" when someone is talking to me), restricted and repetitive behaviors (i.e., I like to collect information about categories of things).

Depressive and anxiety symptoms were measured by the Beck Depression Inventory (BDI); (110), and the Generalized Anxiety Disorder assessment 7-item (GAD-7); (111). These screening tools are used to measure the severity of depression and generalized anxiety in normal and psychiatric populations.

Emotion dysregulation was evaluated by the cyclothymic dimension of the Temperament Evaluation of Memphis, Pisa, Paris and San Diego-autoquestionnaire version (TEMPS-A); (112) and the WRAADDS (16). The TEMPS-A is a questionnaire designed to measure affective temperaments with four subscales: cyclothymic (e.g., my ability to think varies greatly from sharp to dull for no apparent reason), irritable (e.g., people tell me I blow up out of nowhere), hyperthymic (e.g., I love to tackle new projects, even if risky), and anxious (e.g., when someone is late coming home, I fear they may have had an accident). To study ED, in particular emotional lability in ADHD, we were interested in the cyclothymic temperament subscale of the TEMPS-A. All the scales used were self-reported and validated in French (113–117) with Likert scales to consider the frequency of symptoms.

2.2.2. Neuropsychological assessment

Intelligence quotient: In ADHD, ADHD + ASD, and NT groups, IQ was estimated *via* the matrix and the vocabulary subtests of the Wechsler Adult Intelligence Scale–Third Edition (WAIS-III); (118), whereas the matrix and information subtests of the WAIS-IV (119) were used to estimate IQ in the ASD group.

Executive functions: verbal working memory was assessed by the digit-span task (WAIS-III); (108). The "Go-Nogo" subtests with 1-target and 2-target from the TAP 2.3 (120) and the Stroop task (D-KEFS); (121) were, respectively used to investigate motor inhibition and interference inhibition. In addition, the Hayling test (122) was administrated to evaluate semantic inhibition abilities. The "Flexibility" condition of the Stroop task and the Trail Making Test (TMT A&B); (123) has been proposed to measure, respectively cognitive flexibility, speed and attention switching. The Tower of London test (124) was administrated to analyse planning strategies. In addition to EWFT and AVFT, three other conditions of VFT were used, i.e., the free word generation condition (125), as well as the semantic and letter conditions (62).

2.2.3. EWFT and AVFT

Emotional Word Fluency Test (EWFT); (54): the EWFT measures the production of emotion words within 1 min. Similar to other VFT, three scores are generated, including the number of emotion words, the number of repetitions, and the number of rule breaks (i.e., non-emotion words). Feelings such as "love," or typical adjectives describing an emotion such as "happy" were included. In order to measure the variety of the emotional lexicon, words were systematically categorized into one of the six basic emotions defined by Ekman (126). We used Plutchik's wheel of emotions (127), which is based on primary emotions and associated secondary ones. A percentage of the number of words generated by the subject related to each primary emotion (see **Table 2**) was calculated to compare the type of emotion. For example, the percentage of words related to joy compared to the total number of words produced was reported. We were also interested in the frequency of occurrence of every emotions produced in the whole sample. We then measured the average frequency of the emotions generated by each subject. This measure allows to evaluate the ability to produce more varied and less common emotions.

Action Verb Fluency Test (AVFT); (101): This task measures the production of action verbs within 1 min. The number of action nouns, repetitions, and rule violations (i.e., non-action words, such as "appear") were calculated. The frequency of occurrence of action verbs was also averaged over the entire sample. Repetitions and errors were not analysed because their occurrences were too rare in the total sample.

Valence, arousal, dominance: Based on Osgood et al. (128), we analysed the valence, the arousal and the dominance of words. The valence refers to the pleasantness/unpleasantness of a word, while arousal refers to the intensity of the emotion provoked by the stimulus (129, 130). Dominance is the subjectively measured degree of control exerted by a word. This dimension expresses the extent to which the word denotes something that is weak/submissive or strong/dominant (130). This method, previously used in the analysis of the EWFT (61) and the AVFT (131), corresponds to study the average valence, arousal, and dominance of words generated by using a corpus of English words (130). This corpus established for a total of 13,915 words allows quantifying these variables for the

TABLE 2 Examples of words related to the six primary emotions defined by Ekman (126) and Plutchik (127).

Joy	Fear	Anger	Sadness	Surprise	Disgust
Joy	Fear	Anger	Sadness	Surprise	Disgust
Love	Anxiety	Envy	Melancholy	Astonishment	–
Happiness	Anguish	Hate	Depressed	–	–
Excitement	Jealousy	Indignation	Despair	–	–
Euphoria	Stress	Rage	Bitterness	–	–
Pleasure	Panic	Fury	–	–	–

majority of emotional and action words evoked. For each word, a score for valence, arousal and dominance is available based on the ratings of one million subjects. Specifically, ratings on (i) the valence of each word ranging from unhappy to happy; (ii) arousal from excited to calm and (iii) dominance from in control to controlled are available (130). The French words generated in the VFT were translated and then back-translated with translation dictionaries, i.e., WordReference and Linguee. Translations were chosen according to previously generated words when several equivalents existed.

2.3. Statistical analyses

Percentages for qualitative variables and means as well as standard deviations for quantitative variables were included in descriptive statistics. One-way ANOVA and *post hoc* Tukey were conducted to compare demographic and clinical variables between groups. One-way ANOVA and *post hoc* Tukey were also used for each VFT variables (i.e, quantity of words, errors, repetitions, valence, arousal, dominance, and percentage for each primary emotion). Assumptions for ANOVA were verified by the Shapiro-Wilk test for normality and by Levene's test for equality of variances. When the conditions could not be met, Kruskall-Wallis and Mann–Whitney U tests were used to compare the results between groups. The alpha level was adjusted for multiple *post hoc* comparisons of VFT characteristics between groups using the false discovery rate (FDR) method (131). Statistical significance was set at 0.05. Pearson's correlation coefficients were computed in the ADHD groups only between VFT characteristics, clinical symptoms, and cognitive performances. Statistical analyses were performed with the Jasp© and the Jamovi© software.

3. Results

3.1. Descriptive statistics

Compared to NT controls, adults with ADHD and with ADHD + ASD reported higher depressive symptoms [χ^2 (2) = 25.780; $p < 0.001$; $p = 0.001$] and cyclothymic traits [F (2,60) = 27.276; $p < 0.001$; $p = 0.012$, respectively]. This is also the case for anxiety in adults with ADHD [χ^2 (2) = 18,738; $p < 0.001$]. Regarding neuropsychological measures, adults with ADHD made more errors on the Hayling Test compared to NT subjects [χ^2 (2) = 6,708; $p = 0.013$].

3.2. EWFT and AVFT characteristics

First, one-way ANOVA shows that the arousal of emotions produced is significantly different between groups [F (2,86) = 4.074; p = 0.020]. *Post-hoc* test revealed that adults with ADHD generated significantly more arousing emotions (M = 5.29; $p = 0.033$; Figure 1) than NT subjects (M = 5.08). This result is no longer significant after FDR correction ($p = 0.099$). Regarding the type of emotions, adults with ADHD + ASD produced fewer anger-related words (M = 12%) compared to adults with ADHD [M = 24%; χ^2 (2) = 8.339; $p = 0.015$ with FDR; Table 3] and NT controls (M = 24%; $p = 0.022$ with FDR).

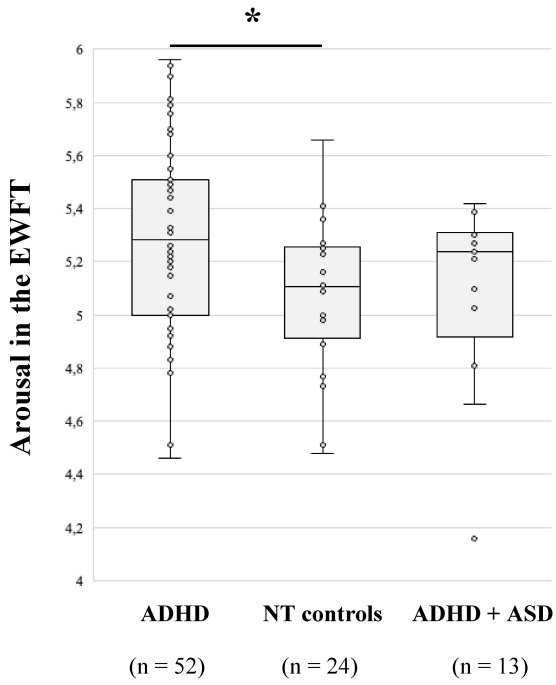


FIGURE 1
Median arousal of emotions produced in the emotion word fluency test (EWFT) according to diagnostic groups. *$p = 0.033$, after the false discovery rate (FDR) correction, $p = 0.099$.

Regarding disgust and surprise the number of occurrences of related words was too rare to be analysed.

Regarding the AVFT, differences between groups are significant for the number of words produced [F (2,85) = 3.655; $p = 0.030$]. Adults with ADHD + ASD produced fewer actions (M = 14.69) compared to NT subjects (M = 19.54; $p = 0.024$). However, this difference was no longer significant after the FDR correction ($p = 0.072$). The other measures, including rule breaks, repetitions, frequency, valence, arousal and dominance of generated words did not differ significantly between groups.

Secondly, we also looked at the VFT performance of a group of adults with ASD only (Table 4). Autistic adults produced more emotions (M = 10.67) and fewer rule breaks (M = 1.92) compared to subjects with ADHD (M = 7.52, $p = 0.003$; M = 3.73, $p = 0.032$, respectively; Table 4). Only the difference in the number of emotions produced remained significant after the FDR correction ($p = 0.018$).

3.3. Correlation analyses between symptoms, EWFT and AVFT

In the ADHD group only, we were interested in the relationship between core symptoms, emotion dysregulation, cold and hot executive functions (i.e., EWFT and AVFT). Regarding ADHD symptoms, hyperactivity was related to the frequency of reported emotions (Table 5). Adults with ADHD produced more frequent emotions if they had an higher level of hyperactivity (r = 0.420; $p = 0.012$). The other main symptom domains, i.e., inattention and impulsivity, were not related to EWFT characteristics ($p > 0.05$; Table 5). Furthermore, emotional over-reactivity in adults with ADHD was associated with the number of emotions (r = 0.440;

TABLE 3 Emotional word fluency test (EWFT) and action verb fluency test (AVFT) characteristics in each group.

	ADHD (AD) (*N* = 52)	ADHD + ASD (AS + AD) (*N* = 13)	Neurotypical controls (NT) (*N* = 24)	Statistical analyses	*P*	*Post hoc*
EWFT–number of words	7.52 (2.42)	8.15 (3.16)	8.67 (2.84)	F (2,86) = 1.598	0.208	/
EWFT–rule breaks	3.73 (3.36)	2.61 (3.15)	3.08 (2.38)	χ^2 (2) = 2.869	0.238	/
EWFT–repetitions	0.38 (0.74)	0.15 (0.38)	0.42 (0.83)	χ^2 (2) = 0.783	0.676	/
EWFT–frequency	0.05 (0.01)	0.05 (0.02)	0.04 (0.01)	F (2,86) = 0.469	0.628	/
EWFT–valence	4.59 (0.71)	4.64 (0.44)	4.41 (0.64)	F (2,86) = 0.715	0.492	/
EWFT–arousal	5.29 (0.35)	5.09 (0.36)	5.08 (0.28)	F (2,86) = 4.074	**0.020***	AD > NT
EWFT–dominance	5.05 (0.44)	4.91 (0.23)	4.88 (0.40)	F (2,86) = 1.555	0.217	/
EWFT–% joy	35% (14%)	35% (8%)	31% (13%)	F (2,86) = 0.692	0.503	/
EWFT–% sadness	20% (12%)	27% (19%)	19% (10%)	F (2,86) = 1.800	0.653	/
EWFT–% anger	24% (15%)	12% (9%)	24% (16%)	χ^2 (2) = 8.339	**0.015****	AD, NT > AS + AD
EWFT–% fear	18% (16%)	24% (16%)	22% (14%)	χ^2 (2) = 3.829	0.147	/
EWFT–% disgust	1% (4%)	1% (3%)	0% (2%)	χ^2 (2) = 0.411	/	/
EWFT–% surprise	2% (6%)	1% (4%)	3% (6%)	χ^2 (2) = 1.386	/	/
AVFT–number of words	18.22 (5.59)	14.69 (3.94)	19.54 (5.07)	F (2,85) = 3.655	**0.030***	NT > AS + AD
AVFT–rule breaks	0.41 (1.47)	0.31 (0.85)	0.00 (0.00)	/	/	/
AVFT–repetitions	0.41 (0.70)	0.15 (0.38)	0.67 (1.05)	χ^2 (2) = 2.619	0.270	/
AVFT–frequency	0.01 (0.00)	0.01 (0.00)	0.01 (0.00)	χ^2 (2) = 2.038	0.361	/
AVFT–valence	5.76 (0.42)	5.83 (0.39)	5.84 (0.33)	F (2,85) = 0.452	0.638	/
AVFT–arousal	4.30 (0.23)	4.27 (0.26)	4.31 (0.18)	F (2,85) = 0.107	0.898	/
AVFT–dominance	5.71 (0.29)	5.80 (0.30)	5.83 (0.21)	F (2,85) = 1.670	0.194	/

*$p < 0.05$.
**Comparisons that remain significant after the false discovery rate (FDR) correction.
P-values in bold = significant *post hoc* comparisons.

$p = 0.008$), as well as the frequency of these words in EWFT (r = –0.398; $p = 0.018$). The greater the emotional over-reactivity, the more numerous and infrequent the generated emotions were in adults with ADHD. In contrast, there was no relationship between EWFT performances and social adjustment (WRAADDS; $p > 0.05$).

Hyperactivity (r = 0.387; $p = 0.024$; Table 5) and impulsivity symptoms (r = 0.494; $p = 0.003$) were also associated with more arousing actions on the AVFT. In addition, affective lability symptoms tended to be related to the number of actions produced (r = 0.337; $p = 0.051$). This suggests that the more adults with ADHD experience mood swings, the more actions they generate spontaneously. Social maladjustment was negatively associated with the frequency of actions reported by adults with ADHD (r = –0.359; $p = 0.037$).

3.4. Correlation analyses between cold executive functions, EWFT and AVFT

Concerning cold executive functions, correlation analyses indicate significant associations between performance on free, letter, semantic, and action verb VFT ($p < 0.05$; Table 6). Nevertheless, VFT measures on the free, semantic, letter and action verb conditions were unrelated to those obtained in the EWFT. With this task, increased cognitive flexibility on the TMT-B is associated with an increased number of emotions (r = –0.443; $p < 0.008$). In addition, increased inhibition errors in the Hayling test is associated with fewer emotional words generated (r = –0.337; $p = 0.048$).

For the AVFT, the faster the automatic response inhibition on the Hayling task, the more action nouns adults with ADHD can generate (r = 0.346; $p = 0.045$). The number of action verbs produced was positively related to working memory performance on the digit span task. Moreover, the number of correct responses in the ToL task, reflecting planning abilities, was also positively associated (r = 0.365; $p = 0.034$) to the number of words generated in the AVFT.

4. Discussion

This study is the first to use a wide range of VFT, including two conditions assessing hot executive functions, in adults with ADHD, ASD, and co-occurring ADHD + ASD, compared to NT controls. The EWFT and the AVFT conditions revealed specific patterns regarding the particularities of affective language and its potential links with emotion dysregulation symptoms in ADHD, i.e., especially emotional over-reactivity and impulsivity. Consistent with our hypotheses, adults with ADHD + ASD produced fewer action verbs compared to NT subjects. These results are congruous with those obtained by Inokuchi and Kamio in autistic adults (105) and are in line with previous studies conducted in adults with ADHD + ASD (46), indicating that the combination of these two conditions is characterized by more severe deficits compared to separate presentations of ADHD or ASD (47–49).

Indeed, process-oriented measures of the emotional output on the EWFT differed between groups. Adults with ADHD produced more arousing emotions than NT subjects. However, this result was no longer significant after FDR correction, which may be due to a lack of statistical power. Nevertheless, this result may be explained by the heightened emotional experience of adults with ADHD. Indeed,

TABLE 4 Comparison of demographic and neuropsychological characteristics of autistic adults (n = 12) with the three other groups.

	ASD (AS) (N = 12)	Statistical analyses	*P*	*Post hoc*
Demographic characteristics				
Age	26.92 (8.35)	χ^2 (3) = 5.736	0.125	/
Gender	41.66% F	χ^2 (3) = 3.830	0.280	/
Years of education	14.67 (1.63)	χ^2 (3) = 6.047	0.109	/
IQ estimation	114.65 (15.82)	F (3,72) = 1.529	0.214	/
WAIS-IV–information (SS)	14.00 (2.88)	/	/	/
Neuropsychological characteristics				
VFT free (number of words)	54.00 (23.95)	F (3,65) = 1.171	0.021*	/
VFT letter (number of words)	23.73 (6.39)	F (3,65) = 0.465	0.087	/
VFT semantic (number of words)	33.36 (6.96)	F (3,65) = 0.893	0.072	/
EWFT–number of words	10.67 (3.31)	F (3,97) = 4.557	**0.005****	AS > AD
EWFT–rule breaks	1.92 (2.19)	χ^2 (3) = 6.432	**0.092**	AD > AS
EWFT–repetitions	0.17 (0.39)	χ^2 (3) = 1.243	0.544	/
EWFT–frequency	0.04 (0.01)	F (3,97) = 0.698	0.556	/
EWFT–valence	4.23 (0.81)	F (3,97) = 1.247	0.297	/
EWFT–arousal	5.17 (0.16)	F (3,97) = 3.015	**0.067**	AD > NT
EWFT–dominance	4.78 (0.34)	F (3,97) = 1.960	0.125	/
EWFT–% joy	25% (14%)	F (3,97) = 1.905	0.134	/
EWFT–% sadness	19% (9%)	χ^2 (3) = 0.987	0.804	/
EWFT–% anger	18% (11%)	χ^2 (3) = 9.806	**0.020****	AD, NT > AS + AD
EWFT–% fear	29% (12%)	χ^2 (3) = 7.969	**0.047***	AS > AD
EWFT–% disgust	7% (6%)	/	/	/
EWFT–% surprise	3% (5%)	/	/	/
AVFT–number of words	18.33 (4.72)	F (3,96) = 2.502	**0.064**	NT > AS + AD
AVFT–rule breaks	0.25 (0.87)	/	/	/
AVFT–repetitions	0.17 (0.58)	χ^2 (3) = 4.877	0.147	/
AVFT–frequency	0.01 (0.00)	F (3,96) = 1.093	0.356	/
AVFT–valence	5.92 (0.29)	F (3,96) = 0.686	0.562	/
AVFT–arousal	4.27 (0.25)	F (3,96) = 0.116	0.951	/
AVFT–dominance	5.81 (0.20)	F (3,96) = 1.325	0.271	/

*$p < 0.05$.

**Comparisons that remains significant after the false discovery rate (FDR) correction.

P-values in bold = significant *post hoc* comparisons.

TABLE 5 Correlation analyses between emotional word fluency test (EWFT), action verb fluency test (AVFT) and clinical characteristics in attention deficit hyperactivity disorder (ADHD) group (n = 52).

	WRAADDS inattention	WRAADDS hyperactivity	WRAADDS impulsivity	WRAADDS affective lability	WRAADDS emotional over-reactivity	WRAADDS social maladjustment	TEMPS-A cyclothymic temperament
EWFT–Number of words	−0.075	−0.275	−0.175	0.185	**0.440***	−0.089	0.052
EWFT–Rule breaks	−0.002	0.169	0.052	0.061	−0.118	0.156	0.291
EWFT–Frequency	0.088	**0.420***	0.237	−0.004	**−0.398***	−0.103	−0.138
EWFT–Valence	−0.236	−0.076	−0.177	−0.131	0.107	−0.072	0.151
EWFT–Arousal	−0.109	−0.204	−0.229	−0.268	−0.123	−0.373	−0.395
EWFT–Dominance	−0.170	−0.072	−0.095	−0.068	0.114	−0.010	−0.115
AVFT–Number of words	−0.097	−0.056	0.019	**0.337***	0.003	0.105	0.228
AVFT–Rule breaks	0.026	0.048	0.041	0.031	0.163	−0.059	−0.270
AVFT–Frequency	0.014	0.154	0.112	−0.171	0.082	**−0.359***	−0.125
AVFT–Valence	−0.236	−0.076	−0.177	−0.131	0.107	−0.072	−0.067
AVFT – Arousal	0.320	**0.387***	**0.494***	0.287	−0.170	0.043	0.047
AVFT–Dominance	−0.122	−0.000	−0.148	−0.200	−0.043	−0.210	−0.115

*$p < 0.05$.

TABLE 6 Pearsons's correlation matrix for performance on executive tests, number of emotions and actions in attention deficit hyperactivity disorder (ADHD) group (n = 52).

	EWFT (number of words)	AF (number of words)	VF free (number of words)	VF semantic (number of words)	VF phono-logical (number of words)	Digit span forward (number of digits)	Digit span backward (number of digits)	Hayling test inhibition (seconds)	Hayling test inhibition (errors)	TAP Go-Nogo 1 out of 2 (errors)	TAP Go-Nogo 2 out of 5 (errors)	Stroop test inter-ferenc–reading (seconds)	TMT Part B–Part A (seconds)	ToL (total correct)	ToL (additional move-ments)	ToL (initation time)
EWFT (number of words)	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–
AVFT (number of words)	0.236	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–
VFT free (number of words)	0.049	**0.613***	–	–	–	–	–	–	–	–	–	–	–	–	–	–
VFT semantic (number of words)	−0.001	**0.517***	**0.552***	–	–	–	–	–	–	–	–	–	–	–	–	–
VFT letter (number of words)	0.168	**0.595***	**0.504***	**0.716***	–	–	–	–	–	–	–	–	–	–	–	–
Digit span forward (number of digits)	0.116	**0.468***	0.291	0.319	0.304	–	–	–	–	–	–	–	–	–	–	–
Digit span backward (number of digits)	0.222	**0.391***	**0.342***	0.303	0.263	**0.369***	–	–	–	–	–	–	–	–	–	–
Hayling test inhibition (seconds)	−0.108	**−0.346***	**−0.343***	**−0.480***	**−0.452***	−0.162	0.037	–	–	–	–	–	–	–	–	–
Hayling test inhibition (errors)	**−0.337***	−0.232	−0.059	−0.079	−0.291	−0.050	**−0.401***	0.141	–	–	–	–	–	–	–	–
TAP Go-Nogo 1 out of 2 (errors)	−0.137	−0.191	−0.103	−0.085	0.011	−0.173	−0.017	0.252	0.024	–	–	–	–	–	–	–
TAP Go-Nogo 2 out of 5 (errors)	−0.109	−0.194	−0.154	−0.056	−0.026	−0.272	0.300	0.308	−0.221	**0.467***	–	–	–	–	–	–

(Continued)

10.3389/fpsyt.2023.1098210

TABLE 6 (Continued)

	EWFT (number of words)	AF (number of words)	VF free (number of words)	VF semantic (number of words)	VF phono-logical (number of words)	Digit span forward (number of digits)	Digit span backward (number of digits)	Hayling test inhibition (seconds)	Hayling test inhibition (errors)	TAP Go-Nogo 1 out of 2 (errors)	TAP Go-Nogo 2 out of 5 (errors)	Stroop test inter-ferenc–reading (seconds)	TMT Part B–Part A (seconds)	ToL (total correct)	ToL (additional move-ments)	ToL (initation time)
Stroop test interference–reading (seconds)	−0.273	**−0.427***	**−0.351***	−0.340	−0.209	**−0.344***	**−0.426***	**0.359***	0.131	0.032	−0.113	–	–	–	–	–
TMT Part B–Part A (seconds)	**−0.443***	−0.038	0.098	−0.176	−0.004	−0.167	−0.274	0.202	0.262	0.047	−0.034	**0.353***	–	–	–	–
ToL (total correct)	0.263	**0.365***	0.260	0.282	**0.385***	0.166	0.320	−0.154	**−0.416***	−0.090	0.046	−0.032	−0.180	–	–	–
ToL (additional movements)	−0.290	−0.101	−0.065	−0.288	−0.306	−0.035	−0.306	−0.024	**0.418***	0.035	−0.144	−0.020	0.165	**−0.805***	–	–
ToL (initation time)	−0.193	0.336	0.165	**0.413***	**0.360***	0.274	**0.343***	−0.167	−0.299	−0.121	−0.128	0.156	−0.201	**0.428***	**−0.419***	–

*$p < 0.05$.

related to symptoms of emotional over-reactivity, individuals with ADHD report experiencing emotions more intensely, which could translate in the EWFT by more arousing emotional words (16–18). Consistent with Barkley's model, which links hot executive functions, cold executive functions, and actions, it is possible that the inhibitory control impairment in ADHD may also involve emotion self-regulation abilities. Hence, inhibitory impairments in ADHD could be seen as widespread, leading to both emotional and non-emotional impulsivity (37, 42). In our study, in adults with ADHD, verbal inhibition abilities on the Hayling task were positively correlated with emotion generation in the EWFT, that is, the more people with ADHD have difficulties inhibiting a verbal response, the fewer the emotional words spontaneously generated. Given the link between emotion dysregulation and executive functions (132), it is possible that increased verbal impulsivity, especially in social interactions, is related to a lack of control over the retrieval of emotional words in ADHD, which translates here by fewer emotional words produced compared to the ASD group (69–71). This hypothesis is consistent with the functional impact of emotion dysregulation symptoms in adults with ADHD, especially in terms of social functioning (27, 133). Indeed, symptoms of ED, including emotional and verbal impulsivity, may lead adults with ADHD to say things impulsively, which can damage their social, marital, and professional relationships (27, 133). Furthermore, in past studies impulsivity in people with ADHD has been linked to increased sensation-seeking personality traits (106). As an example, heightened sensation-seeking in people with ADHD has been linked to the attraction to horror movies (104), which elicits the intense and arousing emotion of fear. Given that impulsivity was found to be positively correlated with intense actions produced in the AVFT in our ADHD group, we argue that this facet of impulsivity (i.e., sensation-seeking) in particular may be associated with intense emotional and movement-related feelings.

Regarding the emotional words related to the six primary emotions (126, 127), the ADHD + ASD group produced significantly fewer emotions related to anger, compared to NT controls and adults with ADHD. This result is consistent with studies showing deficits in the recognition of negative emotions in autistic individuals (136, 137). In particular, autistic people have deficits in the early stages of emotion recognition such as anger (97). According to Tye et al.'s (97) results and embodied cognition theories, difficulties in generating anger-related emotions are associated with the cognitive identification and processing (i.e., alexithymia) of emotions rather than a lower intensity of subjective feeling (138). Indeed, alexithymia leads to reduced emotional word production in a negative context (89). Alexithymia is frequent in ASD (139) and in ADHD (140) and appears to underlie symptoms of emotion dysregulation in autism, which are characterized by intense episodes of anger (139). Given that alexithymia and emotion dysregulation were not assessed in autistic adults in our study, the links between emotional word production, alexithymia, and emotion dysregulation would require further exploration in ASD and ADHD. This is especially important since alexithymia is related to self-harming behaviors (141, 142), used to regulate unidentified anger in ASD (31, 143).

Concerning AVFT, adults with ADHD + ASD generated fewer actions compared to NT controls. These results are consistent with those obtained by Inokuchi and Kamio (105), who found a deficit in the semantic clustering process in VFT in autistic adults compared to NT controls. A decrease in the number of action names spontaneously generated could be caused by executive dysfunction

(105), motor impairment (108), or deficits in social cognition in ASD and ADHD. In addition, the performance of adults with ADHD in this task was correlated with an unconstrained VFT condition (the free condition; **Table 6**) which is particularly dependent on initiation strategies. Nevertheless, the difference with NT subjects was only significant in the ADHD + ASD group. These results are in line with studies postulating that the ADHD + ASD co-occurrence is not a simple addition of the cognitive alterations specific to each disorder (144). Indeed, the combination of these two conditions can lead to more severe disturbance of hot executive functions.

In ADHD, contrary to our hypothesis, participants did not produce more arousing actions compared to the other groups. Nevertheless, symptoms of hyperactivity and impulsivity were positively correlated with the generation of arousing actions. Moreover, we found a negative association between the frequency of actions generated and self-reported difficulties of social adjustment on the WRAADDS. This suggests that unusual actions are related to difficulties in social adaptation in adults with ADHD, which is consistent with the relationship between motor hyperactivity and impaired social functioning in adults with ADHD (11, 145). In our study, motor hyperactivity was also strongly correlated with emotion dysregulation and the number of actions generated was positively correlated with affective lability. Both findings are in line with those from results from past studies suggesting that emotion dysregulation is heightened in the combined ADHD subtype compared to the inattentive subtype (18, 146–149). Dysregulated arousal states have been put forward as an explanation to this association (150). It is therefore possible that dysregulated arousal states underlie affective lability in ADHD–i.e., brief and unpredictable shifts from ordinary mood to depression or mild excitation (151)—and lead to an unstable daily routine where adults with ADHD can rapidly alternate between initiated actions, akin to hyperactivity symptoms.

We also checked the EWFT and AVFT performance of autistic adults without the ADHD co-occurrence. We found that autistic adults produced significantly more emotion words compared to adults with ADHD (**Table 4**). Considering the observed links between the symptoms of emotion dysregulation and performance in the EWFT, these findings suggest that having increased affective language is not necessarily more adaptive, on the one hand, and that autistic adults do not have difficulties in spontaneously generating emotion words, but rather in processing and recognizing emotional states in themselves and others, on the other hand. This might also be due to the fact that people with ADHD are more impulsive, which are related to increased errors on the Hayling task, compared to autistic adults. Indeed, in an arousing context, such as the EWFT, increased verbal impulsivity might prevent people with ADHD from using efficient strategies to retrieve emotion nouns, instead of unrelated off-task words. Moreover, due to the time limited of VFT, errors prevent access to correct answers and result in decreased word output, which seems to have been the case here. This explanation is supported by the negative correlation between the number of words generated in the EWFT and verbal response inhibition errors in the ADHD group. Additionally, in our study, emotional over-reactivity in adults with ADHD was positively associated with the spontaneous generation of numerous and unusual emotions. As a matter of fact, emotional over-reactivity can lead to responses to minor stimuli in the environment that typically do not result in emotional reactions (151). Thus, adults with ADHD may have a broader repertoire of emotional experiences, both in terms of reactivity and experiences, that do not result in an increased word output in the EWFT due to their verbal impulsivity.

This study has several limitations. First, this study is limited by the small sample of adults with co-occurring ADHD + ASD and autistic adults, which results in a lack of statistical power. Analysing VFT performance in a larger sample could allow to tackle the specificities of the ASD group, including in the generation of anger-related words and action verbs. Second, we did not exclude participants with psychopathological comorbidities in the ADHD + ASD and ASD groups. These comorbidities, including anxiety and depressive disorders, are very common in these conditions (152) and are intrinsically linked to emotion dysregulation (33). In addition, hot executive functions are closely related to mood (153), and patients with mood disorders often present executive dysfunction (154–156), notably in verbal fluency (157). There also are specificities of emotion dysregulation in different presentations of ADHD—i.e., inattentive, combined and hyperactive (15). It is therefore important to explore the links between VFT performance and emotion dysregulation in different presentations of ADHD. Finally, given the potential relationship between alexithymia and sensation-seeking in spontaneous emotional and action word production in ADHD and ADHD + ASD, future studies should directly target these dimensions in relation to VFT performance in all groups.

5. Conclusion and perspectives

Taken together, our results suggest that even though emotional processing difficulties is a shared symptom domain between autistic adults, adults with ADHD and adults with ASD + ADHD, several differences can be found using hot executive functions conditions of VFT. First of all, adults with co-occurring ADHD + ASD could present unique affective features that are different from those found in separate presentations of ADHD or ASD, and may have more severe cognitive difficulties, especially in verbal initiation. Most importantly, these results point to the fact that their emotional symptoms do not seem to be a simple addition of both conditions. Secondly, increased emotional word output can also be dysfunctional and linked to emotion dysregulation. While future studies are needed with increased sample sizes and measures of emotion dysregulation symptoms in autistic adults, these findings suggest that different mechanisms are involved in emotional processing in each subgroup and different treatment options could be needed to target them.

Data availability statement

The raw data supporting the conclusions of this article will be made available by the authors, without undue reservation.

Ethics statement

The studies involving human participants were reviewed and approved by CPP South Mediterranean II. The patients/participants provided their written informed consent to participate in this study.

Author contributions

AW: study conception, data collection and coding, statistical analysis, and write the first draft of the manuscript. EM: study conception, manuscript writing and revision, and data collection. SW: study conception, manuscript writing and revision, and data collection. LW: study conception, manuscript writing and revision, and data collection. All authors contributed to the article and approved the submitted version.

Acknowledgments

The authors thank to the participants for their cooperation during the research, and Elena Costache for her help with the acquisition of data.

Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

Publisher's note

References

1. World Health Organization. *The World Health Report: 2004: Changing History*. Geneva: World Health Organization. (ı2004)ı.

2. Boyle CA, Boulet S, Schieve LA, Cohen RA, Blumberg SJ, Yeargin-Allsopp M, et al. Trends in the prevalence of developmental disabilities in US children, 1997-2008. (2011) *Pediatrics.* 127:1034–42. doi: 10.1542/peds.2010-2989.

3. American Psychiatric Association, DSM-5 Task Force. *Diagnostic and Statistical Manual of Mental Disorders: DSM-5™* 5th Edn, Washington, DC: American Psychiatric Publishing, Inc. (2013).

4. Hansen BH, Oerbeck B, Skirbekk B, Petrovski BÉ, Kristensen H. Neurodevelopmental disorders: prevalence and comorbidity in children referred to mental health services. *Nordic J Psychiatry.* (2018) 72:285–91. doi: 10.1080/08039488.2018.1444087

5. Danielson ML, Bitsko RH, Ghandour RM, Holbrook JR, Kogan MD, Blumberg SJ. Prevalence of parent-reported ADHD diagnosis and associated treatment among U.S. children and adolescents, 2016. *J Clin Child Adolescent Psychol.* (2018) 47, 199–212. doi: 10.1080/15374416.2017.1417860.

6. Wender PH. Attention-deficit hyperactivity disorder in adults. *Psychiatric Clin North Am.* (1998) 21, 761–4, v. doi: 10.1016/s0193-953x(05)70039-3

7. Faraone SV, Biederman J, Mick E. The age-dependent decline of attention deficit hyperactivity disorder: a meta-analysis of follow-up studies. *Psychol Med.* (2006) 36, 159–65. doi: 10.1017/S003329170500471X.

8. Simon V, Czobor P, Bálint S, Mészáros A, Bitter I. Prevalence and correlates of adult attention-deficit hyperactivity disorder: meta-analysis. *Br J Psychiatry.* (2009) 194, 204–11. doi: 10.1192/bjp.bp.107.048827.

9. Fayyad J, De Graaf R, Kessler R, Alonso J, Angermeyer M, Demyttenaere K, et al. Cross-national prevalence and correlates of adult attention-deficit hyperactivity disorder. *Br J Psychiatry.* (2007) 190, 402–9. doi: 10.1192/bjp.bp.106.034389.

10. Song P, Zha M, Yang Q, Zhang Y, Li X, Rudan I. The prevalence of adult attention-deficit hyperactivity disorder: a global systematic review and meta-analysis. *J Global Health.* (2021) 11:04009. doi: 10.7189/jogh.11.04009.

11. Martz E, Bertschy G, Kraemer C, Weibel S, Weiner L. Beyond motor hyperactivity: racing thoughts are an integral symptom of adult attention deficit hyperactivity disorder. *Psychiatry Res.* (2021) 301:113988. doi: 10.1016/j.psychres.2021.113988.

12. Biederman J, Mick E, Faraone SV. Age-dependent decline of symptoms of attention deficit hyperactivity disorder: impact of remission definition and symptom type. *Am J Psychiatry.* (2000) 157, 816–8. doi: 10.1176/appi.ajp.157.5.816.

13. Mick E, Faraone SV, Biederman J. Age-dependent expression of attention-deficit/hyperactivity disorder symptoms. *Psychiatric Clin North Am.* (2004) 27, 215–24. doi: 10.1016/j.psc.2004.01.003.

14. Wilens TE, Biederman J, Faraone SV, Martelon M, Westerberg D Spencer, TJ. Presenting ADHD symptoms, subtypes, and comorbid disorders in clinically referred adults with ADHD. *J Clin Psychiatry.* (2009) 70:1557–62.

15. Shaw P, Stringaris A, Nigg J, Leibenluft E. Emotion dysregulation in attention deficit hyperactivity disorder. *Am J Psychiatry.* (2014) 171, 276–93. doi: 10.1176/appi.ajp.2013.13070966.

16. Wender PH. *Attention-deficit Hyperactivity Disorder in Adults*. New York, NY: Oxford University Press. (1995).

17. Cavelti M, Corbisiero S, Bitto H, Moerstedt B, Newark P, Faschina S, et al. A comparison of self-reported emotional regulation skills in adults with attention-deficit/hyperactivity disorder and borderline personality disorder. *J Attent Disord.* (2019) 23, 1396–406. doi: 10.1177/1087054717698814.

18. Skirrow C, Asherson P. Emotional lability, comorbidity and impairment in adults with attention-deficit hyperactivity disorder. *J Affect Disord.* (2013) 147:80–6. doi: 10.1016/j.jad.2012.10.011.

19. Bodalski EA, Knouse LE, Kovalev D. Adult ADHD, emotion dysregulation, and functional outcomes: examining the role of emotion regulation strategies. *J Psychopathol Behav Assess.* (2019) 41:81–92. doi: 10.1007/s10862-018-9695-1.

20. Kooij JJ, Huss M, Asherson P, Akehurst R, Beusterien K, French A. et al. Distinguishing comorbidity and successful management of adult ADHD. *J Attent Disord.* (2012) 16(Suppl. 5), 3S–19S. doi: 10.1177/1087054711435361

21. Jacob CP, Romanos J, Dempfle A, Heine M, Windemuth-Kieselbach C, Kruse A, et al. Co-morbidity of adult attention-deficit/hyperactivity disorder with focus on personality traits and related disorders in a tertiary referral center. *Eur Arch Psychiatry Clin Neurosci.* (2007) 257:309–17. doi: 10.1007/s00406-007-0722-6

22. Bonati M, Cartabia M, Zanetti M, Reale L, Didoni A, Costantino MA, et al. Age level vs grade level for the diagnosis of ADHD and neurodevelopmental disorders. *Eur Child Adolescent Psychiatry.* (2018) 27:1171–80. doi: 10.1007/s00787-018-1180-6

23. Jensen CM, Steinhausen HC. Comorbid mental disorders in children and adolescents with attention-deficit/hyperactivity disorder in a large nationwide study. *Attent Deficit Hyperact Disord.* (2015) 7:27–38. doi: 10.1007/s12402-014-0142-1

24. Chiarotti F, Venerosi A. Epidemiology of autism spectrum disorders: a review of worldwide prevalence estimates since 2014. *Brain Sci.* (2020) 10:274. doi: 10.3390/brainsci10050274

25. Parsons OE, Bayliss AP, Remington A. A few of my favorite things: circumscribed interests in autism are not accompanied by increased attentional salience on a personalized selective attention task. *Mol Autism.* (2017) 8:20. doi: 10.1186/s13229-017-0132-1

26. Christakou A, Murphy CM, Chantiluke K, Cubillo AI, Smith AB, Giampietro V, et al. Disorder-specific functional abnormalities during sustained attention in youth with attention deficit hyperactivity disorder (ADHD) and with autism. *Mol Psychiatry.* (2013) 18:236–44. doi: 10.1038/mp.2011.185

27. Robin L. The impact of ADHD on marriage. *ADHD Rep.* (2002) 10:9–14. doi: 10.1521/adhd.10.3.9.20553

28. de Boo GM, Prins PJ. Social incompetence in children with ADHD: possible moderators and mediators in social-skills training. *Clin Psychol Rev.* (2007) 27:78–97. doi: 10.1016/j.cpr.2006.03.006

29. Little LM, Dean E, Tomchek S, Dunn W. Sensory processing patterns in autism, attention deficit hyperactivity disorder, and typical development. *Phys Occup Therapy Pediatr.* (2018) 38:243–54. doi: 10.1080/01942638.2017.1390809

30. Lecavalier L, Leone S, Wiltz J. The impact of behaviour problems on caregiver stress in young people with autism spectrum disorders. *J Intell Disabil Res.* (2006) 50(Pt. 3):172–83. doi: 10.1111/j.1365-2788.2005.00732.x

31. Laurent AC, Rubin E. Challenges in emotional regulation in asperger syndrome and high-functioning autism. *Top Lang Disord.* (2004) 24:286–97. doi: 10.1097/00011363-200410000-00006

32. Mayes SD, Waxmonsky JD, Calhoun SL, Bixler EO. Disruptive mood dysregulation disorder symptoms and association with oppositional defiant and other disorders in a general population child sample. *J Child Adolescent Psychopharmacol.* (2016) 26:101–6. doi: 10.1089/cap.2015.0074

33. White SW, Mazefsky CA, Dichter GS, Chiu PH, Richey JA, Ollendick TH. Social-cognitive, physiological, and neural mechanisms underlying emotion regulation impairments: understanding anxiety in autism spectrum disorder. *Int J Dev Neurosci.* (2014) 39:22–36. doi: 10.1016/j.ijdevneu.2014.05.012

34. Mazefsky CA, Herrington J, Siegel M, Scarpa A, Maddox BB, Scahill L, et al. The role of emotion regulation in autism spectrum disorder. *J Am Acad Child Adolescent Psychiatry.* (2013) 52:679–88. doi: 10.1016/j.jaac.2013.05.006

35. Samson AC, Wells WM, Phillips JM, Hardan AY, Gross JJ. Emotion regulation in autism spectrum disorder: evidence from parent interviews and children's daily diaries. *J Child Psychol Psychiatry Allied Disciplines.* (2015) 56:903–13. doi: 10.1111/jcpp.12370

36. Eisenberg N, Spinrad TL. Emotion-related regulation: sharpening the definition. *Child Dev.* (2004) 75:334–9. doi: 10.1111/j.1467-8624.2004.00674.x

37. Barkley RA. Differential diagnosis of adults with ADHD: the role of executive function and self-regulation. *J Clin Psychiatry.* (2010) 71:e17. doi: 10.4088/JCP.9066tx1c

38. Samson AC, Huber O, Gross JJ. Emotion regulation in Asperger's syndrome and high-functioning autism. *Emotion.* (2012) 12:659–65. doi: 10.1037/a0027975

39. Miyake A, Friedman NP, Emerson MJ, Witzki AH, Howerter A, Wager TD. The unity and diversity of executive functions and their contributions to complex "Frontal Lobe" tasks: a latent variable analysis. *Cogn Psychol.* (2000) 41:49–100. doi: 10.1006/cogp.1999.0734

40. Chan RC, Shum D, Toulopoulou T, Chen EY. Assessment of executive functions: review of instruments and identification of critical issues. *Arch Clin Neuropsychol.* (2008) 23:201–16. doi: 10.1016/j.acn.2007.08.010

41. McDonald S. Impairments in social cognition following severe traumatic brain injury. *J Int Neuropsychol Soc.* (2013) 19:231–46. doi: 10.1017/S1355617712001506

42. Mitchell JT, Robertson CD, Anastopolous AD, Nelson-Gray RO, Kollins SH. Emotion dysregulation and emotional impulsivity among adults with attention-deficit/hyperactivity disorder: results of a preliminary study. *J Psychopathol Behav Assess.* (2012) 34:510–9. doi: 10.1007/s10862-012-9297-2

43. Mazefsky CA, Oswald DP, Day TN, Eack SM, Minshew NJ, Lainhart JE. ASD, a psychiatric disorder, or both? Psychiatric diagnoses in adolescents with high-functioning ASD. *J Clin Child Adolescent Psychol.* (2012) 41:516–23. doi: 10.1080/15374416.2012.686102

44. Samson AC, Phillips JM, Parker KJ, Shah S, Gross JJ, Hardan AY. Emotion dysregulation and the core features of autism spectrum disorder. *J Autism Dev Disord.* (2014) 44:1766–72. doi: 10.1007/s10803-013-2022-5

45. Mazefsky CA, White SW. Emotion regulation: concepts and practice in autism spectrum disorder. *Child Adolescent Psychiatric Clin North Am.* (2014) 23:15–24. doi: 10.1016/j.chc.2013.07.002

46. Ashwood KL, Tye C, Azadi B, Cartwright S, Asherson P, Bolton P. Brief report: adaptive functioning in children with ASD, ADHD and ASD + ADHD. *J Autism Dev Disord.* (2015) 45:2235–42. doi: 10.1007/s10803-014-2352-y

47. Rao PA, Landa RJ. Association between severity of behavioral phenotype and comorbid attention deficit hyperactivity disorder symptoms in children with autism spectrum disorders. *Autism.* (2014) 18:272–80. doi: 10.1177/1362361312470494

48. Sikora DM, Vora P, Coury DL, Rosenberg D. Attention-deficit/hyperactivity disorder symptoms, adaptive functioning, and quality of life in children with autism spectrum disorder. *Pediatrics.* (2012) 130(Suppl. 2):S91–7. doi: 10.1542/peds.2012-0900G

49. Yerys BE, Wallace GL, Sokoloff JL, Shook DA, James JD, Kenworthy L. Attention deficit/hyperactivity disorder symptoms moderate cognition and behavior in children with autism spectrum disorders. *Autism Res.* (2009) 2:322–33. doi: 10.1002/aur.103

50. Milner B, Branch C, Rasmussen T. Observations on cerebral dominance. In: de Reuck AV, O'Connor M editors. *Ciba Foundation Symposium - Bioassay of Anterior Pituitary and Adrenocortical Hormones (Colloquia on Endocrinology).* London: J. and A. Churchill (1964).

51. Whiteside DM, Kealey T, Semla M, Luu H, Rice L, Basso MR, et al. Verbal fluency: language or executive function measure? *Appl Neuropsychol Adult.* (2016) 23:29–34. doi: 10.1080/23279095.2015.1004574

52. Henry JD, Crawford JR. Verbal fluency deficits in Parkinson's disease: a meta-analysis. *J Int Neuropsychol Soc.* (2004) 10:608–22. doi: 10.1017/S1355617704104141

53. Abbassi E, Kahlaoui K, Wilson MA, Joanette Y. Processing the emotions in words: the complementary contributions of the left and right hemispheres. *Cogn Affect Behav Neurosci.* (2011) 11:372–85. doi: 10.3758/s13415-011-0034-1

54. Abeare CA, Chauvin KA, Kaploun KA, Chu O, Dumitrescu C, Pascual-Leone A. Validation of the emotion word fluency test: preliminary evidence. *J Int Neuropsychol Soc.* (2009) 15:24–5. doi: 10.1037/pas0000965

55. Borod JC, Koff E, Yecker S, Santschi C, Schmidt JM. Facial asymmetry during emotional expression: gender, valence, and measurement technique. *Neuropsychologia.* (1998) 36:1209–15. doi: 10.1016/s0028-3932(97)00166-8

56. Croker V, McDonald S. Recognition of emotion from facial expression following traumatic brain injury. *Brain Injury.* (2005) 19:787–99. doi: 10.1080/02699050500110033

57. Klumpp H, Deldin P. Review of brain functioning in depression for semantic processing and verbal fluency. *Int J Psychophysiol.* (2010) 75:77–85. doi: 10.1016/j.ijpsycho.2009.10.003

58. Spalletta G, Troisi A, Alimenti S, di Michele F, Pau F, Pasini A, et al. Reduced prefrontal cognitive activation associated with aggression in schizophrenia. *Schizophrenia Res.* (2001) 50:134–5. doi: 10.1016/s0920-9964(00)00164-x

59. Lartseva A, Dijkstra T, Buitelaar JK. Emotional language processing in autism spectrum disorders: a systematic review. *Front Hum Neurosci.* (2015) 8:991. doi: 10.3389/fnhum.2014.00991

60. Keatley E, Hanks R, Sander AM, Kratz AL, Tulsky DS, Ianni P, et al. Group differences among caregivers of civilians and service members or veterans with traumatic brain injury. *Arch Phys Med Rehabil.* (2019) 100:S52–7. doi: 10.1016/j.apmr.2018.11.009

61. Abeare CA, Freund S, Kaploun K, McAuley T, Dumitrescu C. The emotion word fluency test (EWFT): initial psychometric, validation, and physiological evidence in young adults. *J Clin Exp Neuropsychol.* (2017) 39:738–52. doi: 10.1080/13803395.2016.1259396

62. Cardebat D, Doyon B, Puel M, Goulet P, Joanette Y. Formal and semantic lexical evocation in normal subjects. per- formance and dynamics of production as a function of sex, age and educational level. *Acta Neurol Belg.* (1990) 90:207–17.

63. Troyer AK, Moscovitch M, Winocur G. Clustering and switching as two components of verbal fluency: evidence from younger and older healthy adults. *Neuropsychology.* (1997) 11:138–46. doi: 10.1037//0894-4105.11.1.138

64. Oldfield RC, Wingfield A. Response latencies in naming objects. *Quarterly J Exp Psychol.* (1965) 17:273–81. doi: 10.1080/17470216508416445

65. Xiao X, Zhao D, Zhang Q, Guo CY. Retrieval of concrete words involves more contextual information than abstract words: multiple components for the concreteness effect. *Brain Lang.* (2012) 120:251–8. doi: 10.1016/j.bandl.2011.09.006

66. Ergis A-M, Gierski F. Les fluences verbales : aspects theìoriques et nouvelles approches. *L'anneìe Psychol.* (2004) 104:331–59. doi: 10.3406/psy.2004.29670

67. Walker AJ, Shores EA, Trollor JN, Lee T, Sachdev PS. Neuropsychological functioning of adults with attention deficit hyperactivity disorder. *J Clin Exp Neuropsychol.* (2000) 22:115–24.

68. Tucha O, Mecklinger L, Laufkötter R, Kaunzinger I, Paul GM, Klein HE, et al. Clustering and switching on verbal and figural fluency functions in adults with attention deficit hyperactivity disorder. *Cogn Neuropsychiatry.* (2005) 10:231–48. doi: 10.1080/13546800444000047

69. Boonstra AM, Oosterlaan J, Sergeant JA, Buitelaar JK. Executive functioning in adult ADHD: a meta-analytic review. *Psychol Med.* (2005) 35:1097–108. doi: 10.1017/s003329170500499x

70. Schoechlin C, Engel RR. Neuropsychological performance in adult attention-deficit hyperactivity disorder: meta-analysis of empirical data. *Arch Clin Neuropsychol.* (2005) 20:727–44. doi: 10.1016/j.acn.2005.04.005

71. Andreou G, Trott K. Verbal fluency in adults diagnosed with attention-deficit hyperactivity disorder (ADHD) in childhood. *Attent Deficit Hyperact Disord.* (2013) 5:343–51. doi: 10.1007/s12402-013-0112-z

72. Johnson DE, Epstein JN, Waid LR, Latham PK, Voronin KE, Anton RF. Neuropsychological performance deficits in adults with attention deficit/ hyperactivity disorder. *Arch Clin Neuropsychol.* (2001) 16:587–604.

73. Martz E, Weibel S, Weiner L. An overactive mind: investigating racing thoughts in ADHD, hypomania and comorbid ADHD and bipolar disorder via verbal fluency tasks. *J Affect Disord.* (2022) 300:226–34. doi: 10.1016/j.jad.2021.12.060

74. Dichter GS, Lam KS, Turner-Brown LM, Holtzclaw TN, Bodfish JW. Generativity abilities predict communication deficits but not repetitive behaviors in autism spectrum disorders. *J Autism Dev Disord.* (2009) 39:1298–304. doi: 10.1007/s10803-009-0742-3

75. Spek A, Schatorjé T, Scholte E, van Berckelaer-Onnes I. Verbal fluency in adults with high functioning autism or Asperger syndrome. *Neuropsychologia.* (2009) 47:652–6. doi: 10.1016/j.neuropsychologia.2008.11.015

76. Ehlen F, Roepke S, Klostermann F, Baskow I, Geise P, Belica C, et al. Small semantic networks in individuals with autism spectrum disorder without intellectual impairment: a verbal fluency approach. *J Autism Dev Disord.* (2020) 50:3967–87. doi: 10.1007/s10803-020-04457-9

77. Tóth O, Pesthy O, Farkas K, Guttengéber A, Komoróczy E, Réthelyi JM, et al. Intact fluency in autism? a comprehensive approach of verbal fluency task including word imageability and concreteness. *Autism Res.* (2022) 15:677–86. doi: 10.1002/aur.2672

78. Barnard L, Muldoon K, Hasan R, O'Brien G, Stewart M. Profiling executive dysfunction in adults with autism and comorbid learning disability. *Autism.* (2008) 12:125–41. doi: 10.1177/1362361307088486

79. Minshew NJ, Goldstein G, Siegel DJ. Speech and language in high-functioning autistic individuals. *Neuropsychology.* (1995) 9:255–61. doi: 10.1037/0894-4105.9.2.255

80. Mottron L, Bzdok D. Autism spectrum heterogeneity: fact or artifact? *Mol Psychiatry.* (2020) 25:3178–85. doi: 10.1038/s41380-020-0748-y

81. Torenvliet C, Groenman AP, Radhoe TA, Agelink, van Rentergem JA, Geurts HM. One size does not fit all: an individualized approach to understand heterogeneous cognitive performance in autistic adults. *Autism Res.* (2022). [Epub ahead of print].

82. Carmo JC, Duarte E, Pinho S, Marques JF, Filipe CN. Verbal fluency as a function of time in autism spectrum disorder: an impairment of initiation processes? *J Clin Exp Neuropsychol.* (2015) 37:710–21. doi: 10.1080/13803395.2015.1062082

83. Dunn M, Gomes H, Sebastian M. Prototypicality of responses of autistic, language disordered, and normal children in a word fluency task. *Child Neuropsychol.* (1996) 2:99–108. doi: 10.1080/09297049608401355

84. Kelley E, Paul JJ, Fein D, Naigles LR. Residual language deficits in optimal outcome children with a history of autism. *J Autism Dev Disord.* (2006) 36:807–28. doi: 10.1007/s10803-006-0111-4

85. Eigsti IM, Bennetto L, Dadlani MB. Beyond pragmatics: morphosyntactic development in autism. *J Autism Dev Disord.* (2007) 37:1007–23. doi: 10.1007/s10803-006-0239-2

86. Prelock PJ, Nelson NW. Language and communication in autism: an integrated view. *Pediatric Clin North Am.* (2012) 59:129–45, xi. doi: 10.1016/j.pcl.2011.10.008.

87. Hubbard DJ, Faso DJ, Assmann PF, Sasson NJ. Production and perception of emotional prosody by adults with autism spectrum disorder. *Autism Res.* (2017) 10:1991–2001. doi: 10.1002/aur.1847

88. Kinnaird E, Stewart C, Tchanturia K. Investigating alexithymia in autism: a systematic review and meta-analysis. *Eur Psychiatry.* (2019) 55:80–9. doi: 10.1016/j.eurpsy.2018.09.004

89. Camia C, Desmedt O, Luminet O. Exploring autobiographical memory specificity and narrative emotional processing in alexithymia. *Narrat Inquiry.* (2020) 30:59–79. doi: 10.1075/ni.18089.kob

90. Wilbarger JL, McIntosh DN, Winkielman P. Startle modulation in autism: positive affective stimuli enhance startle response. *Neuropsychologia.* (2009) 47:1323–31. doi: 10.1016/j.neuropsychologia.2009.01.025

91. Corden B, Chilvers R, Skuse D. Avoidance of emotionally arousing stimuli predicts social-perceptual impairment in Asperger's syndrome. *Neuropsychologia.* (2008) 46:137–47. doi: 10.1016/j.neuropsychologia.2007.08.005

92. Wong K, Lee K, Tsze S, Yu WS, Ng IH, Tong M, et al. Comparing early pragmatics in typically developing children and children with neurodevelopmental disorders. *J Autism Dev Disord.* (2022) 52:3825–39. doi: 10.1007/s10803-021-05261-9

93. Bora E, Pantelis C. Meta-analysis of social cognition in attention-deficit/hyperactivity disorder (ADHD): comparison with healthy controls and autistic spectrum disorder. *Psychol Med.* (2016) 46:699–716. doi: 10.1017/S0033291715002573

94. Schönenberg M, Schneidt A, Wiedemann E, Jusyte A. Processing of dynamic affective information in adults with ADHD. *J Attent Disord.* (2019) 23:32–9. doi: 10.1177/1087054715577992

95. Da Fonseca D, Seguier V, Santos A, Poinso F, Deruelle C. Emotion understanding in children with ADHD. *Child Psychiatry Hum Dev.* (2009) 40:111–21. doi: 10.1007/s10578-008-0114-9

96. Greene RW, Biederman J, Faraone SV, Ouellette CA, Penn C, Griffin SM. Toward a new psychometric definition of social disability in children with attention-deficit hyperactivity disorder. *J Am Acad Child Adolescent Psychiatry.* (1996) 35:571–8. doi: 10.1097/00004583-199605000-00011

97. Tye C, Battaglia M, Bertoletti E, Ashwood KL, Azadi B, Asherson P, et al. Altered neurophysiological responses to emotional faces discriminate children with ASD, ADHD and ASD+ADHD. *Biol Psychol.* (2014) 103:125–34. doi: 10.1016/j.biopsycho.2014.08.013

98. Moseley R, Carota F, Hauk O, Mohr B, Pulvermüller F. A role for the motor system in binding abstract emotional meaning. *Cereb Cortex.* (2012) 22:1634–47. doi: 10.1093/cercor/bhr238

99. Pulvermüller F, Fadiga L. Active perception: sensorimotor circuits as a cortical basis for language. *Nat Rev. Neurosci.* (2010) 11:351–60. doi: 10.1038/nrn2811

100. Damasio AR, Tranel D. Nouns and verbs are retrieved with differently distributed neural systems. *Proc Natl Acad Sci USA.* (1993) 90:4957–60. doi: 10.1073/pnas.90.11.4957

101. Piatt AL, Fields JA, Paolo AM, Tröster AI. Action (verb naming) fluency as an executive function measure: convergent and divergent evidence of validity. *Neuropsychologia.* (1999) 37:1499–503. doi: 10.1016/s0028-3932(99)00066-4

102. Fettes P, Schulze L, Downar J. Cortico-Striatal-Thalamic loop circuits of the orbitofrontal cortex: promising therapeutic targets in psychiatric illness. *Front Systems Neurosci.* (2017) 11:25. doi: 10.3389/fnsys.2017.00025

103. Cubillo A, Halari R, Smith A, Taylor E, Rubia K. A review of fronto-striatal and fronto-cortical brain abnormalities in children and adults with attention deficit hyperactivity disorder (ADHD) and new evidence for dysfunction in adults with ADHD during motivation and attention. *Cortex.* (2012) 48:194–215. doi: 10.1016/j.cortex.2011.04.007

104. Nishitani N, Avikainen S, Hari R. Abnormal imitation-related cortical activation sequences in Asperger's syndrome. *Ann Neurol.* (2004) 55:558–62. doi: 10.1002/ana.20031

105. Inokuchi E, Kamio Y. Qualitative analyses of verbal fluency in adolescents and young adults with high-functioning autism spectrum disorder. *Res Autism Spectrum Disord.* (2013) 7:1403–10. doi: 10.1016/j.rasd.2013.08.010

106. Lopez K. *Pragmatics and Semantics in Children with Autism Spectrum Disorder*. New York, NY: CUNY Academic Works (2015).

107. Baron-Cohen S. *Mindblindness: an Essay on Autism and Theory of Mind*. Cambridge, MA: MIT Press (1995).

108. Hillus J, Moseley R, Roepke S, Mohr B. Action semantic deficits and impaired motor skills in autistic adults without intellectual impairment. *Front Hum Neurosci.* (2019) 13:256. doi: 10.3389/fnhum.2019.00256

109. Allison C, Auyeung B, Baron-Cohen S. Toward brief "Red Flags" for autism screening: the short autism spectrum quotient and the short quantitative checklist for autism in toddlers in 1,000 cases and 3,000 controls [corrected]. *J Am Acad Child Adolescent Psychiatry.* (2012) 51:202–212.e7. doi: 10.1016/j.jaac.2011.11.003.

110. Beck AT, Ward CH, Mendelson M, Mock J, Erbaugh J. An inventory for measuring depression. *Arch General Psychiatry.* (1961) 4:561–71. doi: 10.1001/archpsyc.1961.01710120031004

111. Spitzer RL, Kroenke K, Williams JB, Löwe B. A brief measure for assessing generalized anxiety disorder: the GAD-7. *Arch Int Med.* (2006) 166:1092–7. doi: 10.1001/archinte.166.10.1092

112. Akiskal HS, Placidi GF, Maremmani I, Signoretta S, Liguori A, Gervasi R, et al. TEMPS-I: delineating the most discriminant traits of the cyclothymic, depressive, hyperthymic and irritable temperaments in a nonpatient population. *J Affect Disord.* (1998) 51:7–19. doi: 10.1016/s0165-0327(98)00152-9

113. Weibel S, Bicego F, Muller S, Martz E, Costache ME, Kraemer C, et al. Two facets of emotion dysregulation are core symptomatic domains in adult ADHD: results from the SR-WRAADDS, a broad symptom self-report questionnaire. *J Attent Disord.* (2022) 26:767–78. doi: 10.1177/10870547211027647

114. Sonié S, Kassai B, Pirat E, Bain P, Robinson J, Gomot M, et al. The French version of the autism-spectrum quotient in adolescents: a cross-cultural validation study. *J Autism Dev Disord.* (2013) 43:1178–83. doi: 10.1007/s10803-012-1663-0

115. Byrne BM, Baron P. Measuring adolescent depression: tests of equivalent factorial structure for english and french versions of the beck depression inventory. *Appl Psychol.* (1994) 43:33–47. doi: 10.1111/j.1464-0597.1994.tb00808.x

116. Micoulaud-Franchi JA, Lagarde S, Barkate G, Dufournet B, Besancon C, Trébuchon-Da Fonseca A, et al. Rapid detection of generalized anxiety disorder and major depression in epilepsy: validation of the GAD-7 as a complementary tool to the NDDI-E in a French sample. *Epilepsy Behav.* (2016) 57(Pt. A):211–6. doi: 10.1016/j.yebeh.2016.02.015

117. Krebs MO, Kazes M, Olié JP, Loo H, Akiskal K, Akiskal H. The French version of the validated short TEMPS-A: the temperament evaluation of Memphis, Pisa, Paris and San Diego. *J Affect Disord.* (2006) 96:271–3. doi: 10.1016/j.jad.2006.11.001

118. Wechsler D. *Wechsler Adult Intelligence Scale*. 3rd Edn. San Antonio, CA: The Psychological Corporation (1997).

119. Wechsler D. *WAIS IV Nouvelle Version de l'échelle D'intelligence de Weschler Pour Adultes*. 5 ed. Paris: Edition du Centre de Psychologie Appliquée (2011).

120. Zimmermann P, Fimm B. *Test D'évaluation de L'attention (TEA). Version 2.1*. Würselen: Psytest (2009).

121. Delis DC, Kaplan E, Kramer JH. *Delis-Kaplan Executive Function System§(DKEFS§): Examiner's Manual: Flexibility of Thinking, Concept Formation, Problem Solving, Planning, Creativity, Impluse Control, Inhibition*. London: Pearson (2001).

122. Burgess PW, Shallice T. *The Hayling and Brixton Tests*. Bury St Edmunds: Thames Valley Test Company (1997).

123. Tombaugh TN. Trail making test A and B: normative data stratified by age and education. *Arch Clin Neuropsychol.* (2004) 19:203–14. doi: 10.1016/S0887-6177(03)00039-8

124. Shallice T. SpeciÆc impairments of planning. *Philos Trans R Soc London Biol.* (1982) 298:199–209.

125. Joanette Y, Ska B, Côté H. *Protocole Montréal d'Évaluation de la Communication*. Isbergues: Ortho Édition (2004).

126. Ekman P. Cross-cultural studies of facial expressions. In: Ekman P editor. *Darwin and Facial Expressions*. London: Academic Press (1973). p. 169–222.

127. Plutchik R. A general psychoevolutionary theory of emotion. In: Plutchik R, Kellerman H editors. *Emotion: Theory, Research, and Experience*. New York, NY: Academic Press (1980). p. 3–33. doi: 10.1016/B978-0-12-558701-3.50007-7

128. Osgood CE, Suci GJ, Tannenbaum PH. *The Measurement of Meaning*. Champaign, IL: University of Illinois Press (1957). doi: 10.1177/001654926000600217

129. Söderholm C, Häyry E, Laine M, Karrasch M. Valence and arousal ratings for 420 Finnish nouns by age and gender. *PLoS One.* (2013) 8:e72859. doi: 10.1371/journal.pone.0072859

130. Warriner AB, Kuperman V, Brysbaert M. Norms of valence, arousal, and dominance for 13,915 English lemmas. *Behav Res Methods.* (2013) 45:1191–207. doi: 10.3758/s13428-012-0314-x

131. Benjamini Y, Hochberg Y. Controlling the false discovery rate: a practical and powerful approach to powerful testing. *J R Statist Soc B.* (1995) 57:289–300.

132. Petrovic P, Castellanos FX. Top-Down dysregulation-from ADHD to emotional instability. *Front Behav Neurosci.* (2016) 10:70. doi: 10.3389/fnbeh.2016.00070

133. Barkley RA, Murphy KR. Impairment in occupational functioning and adult ADHD: the predictive utility of executive function (EF) ratings versus EF tests. *Arch Clin Neuropsychol.* (2010) 25:157–73. doi: 10.1093/arclin/acq014

134. López-Bonilla J, Lopez L. Sensation seeking and the use of the internet: a confirmatory analysis of the brief sensation seeking scale (BSSS) by gender. *Soc Sci Comp Rev.* (2010) 28:177–93. doi: 10.1177/0894439309341

135. Martin GN. (Why) do you like scary movies? a review of the empirical research on psychological responses to horror films. *Front Psychol.* (2019) 10:2298. doi: 10.3389/fpsyg.2019.02298

136. Ashwin C, Chapman E, Colle L, Baron-Cohen S. Impaired recognition of negative basic emotions in autism: a test of the amygdala theory. *Soc Neurosci.* (2006) 1:349–63. doi: 10.1080/17470910601040772

137. Uljarevic M, Hamilton A. Recognition of emotions in autism: a formal meta-analysis. *J Autism Dev Disord.* (2013) 43:1517–26. doi: 10.1007/s10803-012-1695-5

138. Rogers K, Dziobek I, Hassenstab J, Wolf OT, Convit A. Who cares? revisiting empathy in Asperger syndrome. *J Autism Dev Disord.* (2007) 37:709–15. doi: 10.1007/s10803-006-0197-8

139. Jahromi LB, Meek SE, Ober-Reynolds S. Emotion regulation in the context of frustration in children with high functioning autism and their typical peers. *J Child Psychol Psychiatry Allied Disciplines.* (2012) 53:1250–8. doi: 10.1111/j.1469-7610.2012.02560.x

140. Edel MA, Rudel A, Hubert C, Scheele D, Brüne M, Juckel G, et al. Alexithymia, emotion processing and social anxiety in adults with ADHD. *Eur J Med Res.* (2010) 15:403–9. doi: 10.1186/2047-783x-15-9-403

141. Duerden EG, Oatley HK, Mak-Fan KM, McGrath PA, Taylor MJ, Szatmari P, et al. Risk factors associated with self-injurious behaviors in children and adolescents with autism spectrum disorders. *J Autism Dev Disord.* (2012) 42:2460–70. doi: 10.1007/s10803-012-1497-9

142. Culpin I, Mars B, Pearson RM, Golding J, Heron J, Bubak I, et al. Autistic traits and suicidal thoughts, plans, and self-harm in late adolescence: population-based cohort study. *J Am Acad Child Adolescent Psychiatry.* (2018) 57:313–320.e6. doi: 10.1016/j.jaac.2018.01.023

143. Moseley RL, Gregory NJ, Smith P, Allison C, Baron-Cohen S. A 'choice', an 'addiction', a way 'out of the lost': exploring self-injury in autistic people without intellectual disability. *Mol Autism.* (2019) 10:18. doi: 10.1186/s13229-019-0267-3

144. Nydén A, Niklasson L, Stahlberg O, Anckarsater H, Wentz E, Rastam M, et al. Adults with autism spectrum disorders and adhd neuropsychological aspects. *Res Dev Disabil.* (2010) 31:1659–68. doi: 10.1016/j.ridd.2010.04.010

145. Canu WH, Carlson CL. ADHD and social adapta- tion: from childhood to adulthood. *ADHD Report.* (2004) 12:1–6. doi: 10.1521/adhd.12.2.1.30979

146. Reimherr FW, Marchant BK, Gift TE, Steans TA, Wender PH. Types of adult attention-deficit hyperactivity disorder (ADHD): baseline characteristics, initial response, and long-term response to treatment with methylphenidate. *Attent Deficit Hyperact Disord.* (2015) 7:115–28. doi: 10.1007/s12402-015-0176-z

147. Reimherr FW, Roesler M, Marchant BK, Gift TE, Retz W, Philipp-Wiegmann F, et al. Types of adult attention-deficit/hyperactivity disorder: a replication analysis. *J Clin Psychiatry.* (2020) 81:19m13077. doi: 10.4088/JCP.19m13077

148. Marchant BK, Reimherr FW, Robison D, Robison RJ, Wender PH. Psychometric properties of the wender-reimherr adult attention deficit disorder scale. *Psychol Assessment.* (2013) 25:942–50. doi: 10.1037/a0032797

149. Corbisiero S, Mörstedt B, Bitto H, Stieglitz RD. Emotional dysregulation in adults with attention-deficit/hyperactivity disorder-validity, predictability, severity, and comorbidity. *J Clin Psychol.* (2017) 73:99–112. doi: 10.1002/jclp.22317

150. Martella D, Aldunate N, Fuentes LJ, Sánchez-Pérez N. Arousal and executive alterations in attention deficit hyperactivity disorder (ADHD). *Front Psychol.* (2020) 11:1991. doi: 10.3389/fpsyg.2020.01991

151. Davidson R. Affective style and affective disorders: perspectives from affective neuroscience. *Cogn Emot.* (1998) 12:307–30. doi: 10.1080/026999398379628

152. Lundström S, Reichenberg A, Melke J, Råstam M, Kerekes N, Lichtenstein P, et al. Autism spectrum disorders and coexisting disorders in a nationwide Swedish twin study. *J Child Psychol Psychiatry Allied Disciplines.* (2015) 56:702–10. doi: 10.1111/jcpp.12329

153. Bernabei L, Bersani FS, Delle Chiaie R, Pompili E, Casula S, D'Aniello G, et al. A preliminary study on hot and cool executive functions in bipolar disorder and on their association with emotion regulation strategies. *Riv Psichiatria.* (2018) 53:331–5. doi: 10.1708/3084.30767

154. Robinson LJ, Ferrier IN. Evolution of cognitive impairment in bipolar disorder: a systematic review of cross-sectional evidence. *Bipolar Disord.* (2006) 8:103–16. doi: 10.1111/j.1399-5618.2006.00277.x

155. Torres, IJ, Boudreau VG, Yatham LN. Neuropsychological functioning in euthymic bipolar disorder: a meta-analysis. *Acta Psychiatrica Scand Suppl.* (2007) 116:17–26. doi: 10.1111/j.1600-0447.2007.01055.x

156. Trivedi MH, Greer TL. Cognitive dysfunction in unipolar depression: implications for treatment. *J Affect Disord.* (2014) 152-154:19–27. doi: 10.1016/j.jad.2013.09.012

157. Raucher-Chéné D, Achim AM, Kaladjian A, Besche-Richard C. Verbal fluency in bipolar disorders: a systematic review and meta-analysis. *J Affect Disord.* (2017) 207:359–66. doi: 10.1016/j.jad.2016.09.039

frontiers | Frontiers in Psychiatry

TYPE Original Research
PUBLISHED 24 February 2023
DOI 10.3389/fpsyt.2023.1134396

OPEN ACCESS

EDITED BY
Ru Ying Cai,
Autism Spectrum Australia, Australia

REVIEWED BY
Gellan Ahmed,
Assiut University, Egypt
Maria Giuseppina Petruzzelli,
University of Bari Aldo Moro, Italy

*CORRESPONDENCE
Xiaoying Zheng
✉ zhengxiaoying@sph.pumc.edu.cn

SPECIALTY SECTION
This article was submitted to Autism, a section of the journal Frontiers in Psychiatry

RECEIVED 30 December 2022
ACCEPTED 06 February 2023
PUBLISHED 24 February 2023

CITATION
Zhao Y, Luo Y, Zhang R and Zheng X (2023) The impact of COVID-19 pandemic on emotional and behavioral problems of children with autism spectrum disorder and developmental delay aged 1–6 years in China.
Front. Psychiatry 14:1134396.
doi: 10.3389/fpsyt.2023.1134396

The impact of COVID-19 pandemic on emotional and behavioral problems of children with autism spectrum disorder and developmental delay aged 1–6 years in China

Yanan Zhao[1,2,3], Yanan Luo[4], Rong Zhang[5,6,7] and Xiaoying Zheng[1,3,7]*

[1]School of Population Medicine and Public Health, Chinese Academy of Medical Sciences/Peking Union Medical College, Beijing, China, [2]China Center for Health Development Studies, Peking University, Beijing, China, [3]Asia-Pacific Economic Cooperation Health Sciences Academy, Peking University, Beijing, China, [4]Department of Global Health, School of Public Health, Peking University, Beijing, China, [5]Neuroscience Research Institute, Peking University, Beijing, China, [6]Key Laboratory for Neuroscience of the Ministry of Education, Key Laboratory for Neuroscience of the Ministry of National Health Commission, Department of Neurobiology, School of Basic Medical Sciences, Peking University Health Science Center, Beijing, China, [7]Autism Research Centre, Peking University Health Science Centre, Beijing, China

Introduction: The COVID-19 pandemic outbreak have caused increased levels of emotional and behavioral problems, particularly among people with pre-existing mental health conditions. Young individuals with autism spectrum disorders (ASD) and developmental delay (DD) are particularly at risk due to their vulnerability. The purpose of this study was to look into the different effects of the COVID-19 pandemic on 1–6-year-old children with ASD and DD.

Methods: Parents and guardians of children with ASD completed an online survey that included questions about their children's socio-demographics characteristics, the effects of the COVID-19 outbreak on their health, and what they needed in order to deal with the conditions of the pandemic.

Results: This study compared 4,138 children with ASD to 711 children with DD. Children with ASD had a higher risk of having more emotional and behavioral problems than children with DD (OR 1.38, 95% CI 1.12–1.70). Compared to parent-oriented rehabilitation at home, discontinuing rehabilitation had a higher likelihood of negative emotional and behavioral change (OR 1.67, 95% CI 1.41–1.98). Having teachers' online support had a higher likelihood of negative emotional and behavioral change for ASD children (OR 1.26, 95% CI 1.03–1.54).

Conclusions: This article provided evidence that children with developmental disabilities, particularly ASD, were at risk for a variety of challenges to their emotional functioning during the COVID-19 period, and that online support was not an ideal way for children with ASD to receive effective educational intervention in China.

KEYWORDS

COVID-19, Child and Adolescent Psychiatry, development delay, autism spectrum disorder, emotional and social development

Introduction

The COVID-19 pandemic is a global health crisis that has resulted in a public health emergency. It has caused a high level of psychological stress in the general population (1) as well as an increased risk of emotional and behavioral problems in people who already have a mental health condition (2).

The massive effects of the COVID-19 pandemic have forced both the formal and informal sectors to halt operations, including the educational sector (3). Individuals with special needs are receiving fewer, if any, crucial therapy hours (e.g., speech therapy, behavioral therapy) and classroom time than they would normally (4), particularly children. Many parents have considered home education as an alternative mode of intervention during the pandemic (5). Several studies have confirmed that home education was an additional useful learning method for children during the COVID lockdown (3, 5, 6). However, implementing home education presents numerous challenges (7), and the provision of online educational intervention is insufficient (3, 8). The need for novel approaches and the continuity of care for those with chronic health problems during the pandemic cannot be overstated (9).

Among vulnerable populations, children with severe developmental disabilities are of particular concern regarding how the COVID-19 outbreak may have impacted their wellbeing and health (7, 8). Two common developmental disorders are autism spectrum disorders (ASD) and developmental delay (DD). ASD is a range of neurodevelopmental disorders that are characterized by impairments in social interaction and communication and restricted, repetitive behaviors (10). A child with developmental delay (DD) has delays in language, fine and gross motor functioning, sensory integration, cognitive functioning, or communication, as well as behavioral and socio-psychological problems during infancy or early childhood (11, 12). The COVID-19 outbreak has undoubtedly resulted in a rapidly shifting social situation, which may exacerbate the difficulties of children with developmental disabilities (13), particularly children with ASD. Because they are vulnerable to the effects of prolonged isolation or quarantine, they may struggle to adapt to this new norm, especially given the disorder's inflexibility and insistence on sameness (10). Furthermore, children with ASD are more likely to have comorbidities such as anxiety and learning disabilities, which present additional challenges to be dealt with during the COVID-19 pandemic (4, 7, 14, 15). We may discover that people with developmental disabilities are more vulnerable in general, and that disruptions in routine caused by COVID-19 can cause major emotional and behavioral upheaval (16); however, this has not been thoroughly researched in potentially higher-risk groups, such as children with ASD and DD. We don't know how the effects differ between severe developmental disorders. Children with ASD may have expressive communication challenges, making it difficult to communicate pain (17, 18), while children with DD may have delays in development of language and intellectual abilities. We can better study the significance of online educational intervention and gain a better understanding of different neurodevelopmental conditions by comparing the performance of the two types of children during the epidemic.

The government in China has taken strict measures to control the pandemic in order to protect vulnerable people from COVID-19 while not overburdening the health service. From February 2020, most Chinese residents were required to stay at home and maintain social distance. Although some institutions reopened after April, the primary focus remained on online education. Most institutions or schools reopened after June, but the hours of operation were not consistent due to the epidemic. Suspension of therapy courses for children and adolescents with specific requirement may result in missed opportunities for basic skill development (4, 13, 15–19). Worse, staying at home puts children with special educational needs and their families in an unusually stressful situation (20). According to recent surveys in China, the prevalence of ASD was 0.7% (21), while the prevalence of DD was 4.5% (22). As a developing country with a large population and an ineffective social security system, China faces a health crisis.

Beginning with the negative emotional and behavioral effects of the COVID-19 outbreak on children with ASD and DD, the overall goal of this study is to investigate child statute changes in Chinese families with children with ASD and DD during lockdown. We aimed to (1) investigate the different effects of the COVID-19 pandemic on the emotional and behavioral change of children with ASD and DD; (2) investigate whether any pre-pandemic sociodemographic or child characteristics would predict a negative impact of the pandemic on children with ASD and DD; and (3) investigate the effects of home education and family needs in the COVID-19 lockdown. Based on previous evidence (23), emotional and behavioral problems during the pandemic were hypothesized to predict a poor outcome.

Materials and methods

Data source

This study used data from the Survey on Family Circumstances and Demand for Support and Resources among Autistic Children in China (FCDSR). It was a survey that was distributed to members of the AlsoLife online patient community. The Quality Assurance staff at the China Association of Rehabilitation of Disabled Persons (CARDP) reviewed the survey for editorial and technical suggestions, which aimed to describe the family information, treatment, rehabilitation subsidies, and health expenditure of children with ASD. The other details of the survey have been described elsewhere (24). We did not use a sampling design because there is no nationwide ASD survey in China. A pilot field study ($N = 20$) was conducted to refine the instrument and data collection procedures, and the results indicated that respondents generally understood the questionnaire, so only minor wording changes were made.

The online survey was completed by parents and guardians of people with ASD or DD, of which 78.1% were mothers, 19.7% were fathers, and 2.2% were other guardians. Additionally, ASD advocacy and family support networks were used to distribute and directly encourage survey participation. During the questionnaire collection and data cleaning process, data with obvious errors

Abbreviations: ASD, autism spectrum disorder; LFA, low functioning autism; MFA, middle functioning autism; HFA, high functioning autism; N, number; M, mean; SD, standard deviation.

or omissions were removed. A total of 8,014 households were analyzed, with 4,849 households included in this study, in which 4,138 households had children with ASD and 711 households had children with DD. Families with children diagnosed or suspected of having ASD or DD were invited to participate if their children met the following criteria: (1) age 1–6 years and diagnosed or suspected of having ASD through a hospital diagnosis; (2) diagnosing hospital and diagnosing hospital department both having diagnostic qualifications. Exclusion criteria were patients with physical disabilities, cerebral palsy, or epilepsy as comorbidities. The selection procedure is depicted in **Figure 1**. Thirty-one provinces and a total of 385 cities in China were included. Given the lack of a large-scale survey of children with ASD or DD in China, the second national sample survey of disabled people in China could also be used as a reference. These samples are representative (see **Supplementary Tables 1, 2**).

Measures

The survey included questions about the impact of the COVID-19 outbreak on their wellbeing as well as the requirements for dealing with the pandemic. Each multiple-choice question allowed participants to choose only one item. This measure of emotional and behavioral change was reported by parents who were asked about their children's emotional and behavioral changes (e.g., emotional reactions to stress, emotional self-regulation system's stability and emotional and behavioral problems related to ASD or DD) during the COVID-19 pandemic lockdown. Specifically, a scale of 1 = "improved," 2 = "no change," and 3 = "worse" was used. Demographic variables, family socioeconomic variables, and family treatment history variables were used as control variables in this study. The demographic variables included the age of the children, their gender, and the number of children in the household, and having comorbidities or not. The age was the age at the time of the survey. The comorbidities referred to neurodevelopment disorders, including intellectual disabilities (ID) and attention deficit and hyperactivity disorder (ADHD) in this study. Information on family sociodemographic and medical history was gathered. The income of families was divided into three categories: below average, average, and above average. According to the data distribution, the below average group had an annual income of less than $12,327 (RMB80,000), the average group had an annual income between $12,327 (RMB80,001) and $23,112 (RMB150,000), and the above average group had an annual income greater than $23,112 (RMB150,000).

Statistical analysis

The final raw data were downloaded from WJX Forms into a Microsoft Excel file for analysis using SPSS software. Descriptive statistics were used to provide baseline information concerning survey participants' children with ASD. Logistic regressions were performed to investigate whether the socio-demographic or clinical characteristics of any individuals with ASD would predict a greater frequency and intensity of emotional and behavioral problems following the COVID-19 outbreak. Associations between predictors and independent variables were reported by odds ratios (ORs) and their 95% confidence intervals (CIs). All the estimated costs were converted to US dollar (US$) values in January 2021, when one US$ was equivalent to about 6.49 Chinese yuan. All statistical analyses were conducted using SPSS 22.0 for Windows (SPSS Inc., Chicago, IL, USA).

Consent and ethics approval

All families provided electronic informed consent before enrollment. We assert that all procedures contributing to this work comply with the ethical standards of the relevant national and institutional committees on human experimentation. All procedures involving human subjects/patients were approved by the ethics committee of Peking University Institutional Review Board and approval number is IRB00001052-20016.

Results

Study population

A total of 4,849 eligible children (78.8%) were recruited for the study. **Table 1** presents characteristics of the participating children. Compared with children with DD, children with ASD had a greater proportion of parents with higher education (father with a higher education degree, 66.92% vs. 59.63%; mother with a higher education degree, 66.67% vs. 59.35%; both, $p < 0.01$). Compared with children with DD, children with ASD had a smaller proportion of having comorbidities (19.33% vs. 24.47%, $p < 0.01$). Age, gender, and family income did not differ significantly between the two groups.

Emotional and behavioral impact at the outbreak of the pandemic

Table 2 shows the impact of the pandemic outbreak on children and parents. For the ASD group and the DD group, most of the families engaged in parent-oriented educational intervention (59.35% vs. 54.57%), while more than 20% of the families ceased intervention in the lockdown (22.40% vs. 27.14%). Changes in behavioral or emotional problems had some differences between the two groups, where more children in the ASD group showed a decline than the DD group (22.31% vs. 18.00%).

Most families in both groups increased their parental time spent with their children. In both groups, parental pressures were greater than before.

Predictors of negative impact on emotional and behavioral changes in the pandemic

The logistic regression model results showed that children with ASD had a higher likelihood of experiencing negative emotional

10.3389/fpsyt.2023.1134396

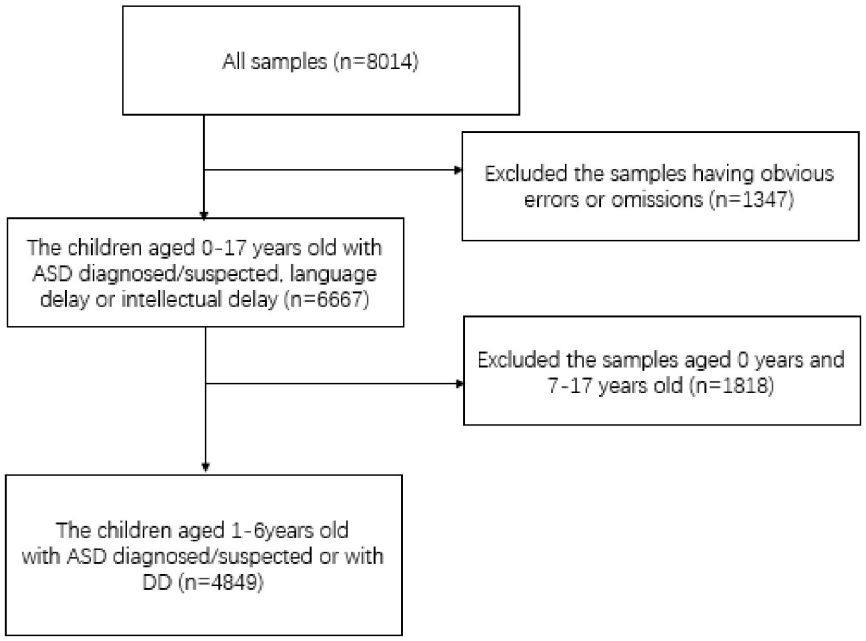


FIGURE 1
Flow chart.

TABLE 1 Characteristics of ASD and DD groups.

	ASD (N = 4,138)		DD (N = 711)		P-value
	Mean/Number	SD/%	Mean/Number	SD/%	
Age (years)	4.21	1.23	4.21	1.26	0.130
Gender of child					
Male	3,451	83.40	542	76.23	<0.001
Female	687	16.60	169	23.77	
Only children					
No	1,969	47.58	355	49.93	0.255
Yes	2,169	52.42	356	50.07	
Comorbidity[a]					
No	3,338	80.67	537	75.53	0.002
Yes	800	19.33	174	24.47	
Higher education degree-father					
No	1,369	33.08	287	40.37	<0.001
Yes	2,769	66.92	424	59.63	
Higher education degree-mother					
No	1,379	33.33	289	40.65	<0.001
Yes	2,759	66.67	422	59.35	
Family annual income[b]	22448.87	25124.34	19521.42	19295.19	0.067

ASD, autism spectrum disorder; DD, developmental delay. [a]Including intellectual disabilities (ID) and attention deficit and hyperactivity disorder (ADHD). [b]USD, $.

and behavioral change (OR 1.38, 95% CI 1.12–1.70; Table 3). Compared with parent-oriented educational intervention at home, ceasing intervention had a higher likelihood of negative emotional and behavioral change (OR 1.67, 95% CI 1.41–1.98; Table 3). Children with comorbidities were associated with a more than one-fold likelihood of having negative emotional and behavioral change (OR 1.52; 95% CI 1.29–1.80; Table 3), and having conflicts with other family members was associated with a more than one-fold likelihood of having a poor mental status (OR 1.44; 95% CI 1.22–1.70; Table 3). Having a family income of average and above average was associated with a lower likelihood of negative emotional and behavioral change (OR 0.82; 95% CI 0.68–0.98 for

10.3389/fpsyt.2023.1134396

TABLE 2 The changes of children and parents after the outbreak of COVID-19.

	ASD (*N* = 4,138)		DD (N = 711)	
	N	%	*N*	%
Rehabilitation type				
Parent oriented	2,456	59.35	388	54.57
Teachers' online support	755	18.25	130	18.28
Cease	927	22.40	193	27.14
Emotional and behavioral change				
Improved	1,573	38.01	286	40.23
No change	1,642	39.68	297	41.77
Worse	923	22.31	128	18.00
Time spent between parents and children				
Shorter	195	4.71	18	2.53
No change	1,102	26.63	212	29.82
Longer	2,841	68.66	481	67.65
Parental pressures change				
More	2,577	62.28	430	60.48
No change	1,241	29.99	231	32.49
Less	320	7.73	50	7.03

ASD, autism spectrum disorder; DD, developmental delay.

TABLE 3 Logistic regression analysis for variables predicting the negative emotional and behavioral change* of children.

Characteristics	(*N* = 4,849)		
	Odds ratio	95% CI	
Disorder			
DD	1.00	reference	
ASD	1.38	1.12	1.70
Rehabilitation type			
Parent oriented	1.00	reference	
Teachers' online support	1.19	0.99	1.44
Cease	1.67	1.41	1.98
Rehabilitation hours			
< 4 h a day	1.00	reference	
≥ 4 h a day	0.85	0.68	1.05
Only children			
No	1.00	reference	
Yes	0.96	0.83	1.11
Gender of child			
Male	1.00	reference	
Female	1.03	0.85	1.11
Age of child			
1–3 years	1.00	reference	
4–6 years	1.15	0.98	1.34
Comorbidity[a]			
No	1.00	reference	
Yes	1.52	1.29	1.80
Household income			
Below average	1.00	reference	
Around average	0.82	0.68	0.98
Above average	0.81	0.67	0.98
Higher education degree-father			
No	1.00	reference	
Yes	1.04	0.86	1.27
Higher education degree-mother			
No	1.00	reference	
Yes	1.11	0.91	1.34
Time spent between parents and children			
Shorter	1.00	reference	
No change	0.43	0.31	0.61
Longer	0.41	0.30	0.57

*Emotional and behavioral change is "worse" in the survey. ASD, autism spectrum disorder; DD, developmental delay. [a]Including intellectual disabilities (ID) and attention deficit and hyperactivity disorder (ADHD).

average income families and OR 0.81; 95% CI 0.67–0.98 for above average income families; Table 3) compared to those having an income below average. Parental time spent with children had an effect on children's emotions and behaviors. If the amount of time spent had not changed or had increased, the likelihood of having more negative emotional and behavioral changes decreased (OR 0.43; 95% CI 0.31–0.61 for families where there was no change in time spent with children; OR 0.41; 95% CI 0.30–0.57 for families where time spent with children increased; see Table 3).

Child gender, age, number of children in the household, and parent's educational level were not significantly associated with negative emotional or behavioral change of children (Table 3).

Stratifying by age groups resulted in significant differences ($p < 0.05$). For the 4–6-year-old group, the likelihood of negative emotional and behavioral change for children with ASD was higher than children with DD ($P < 0.05$). However, for the 1–3-year-old group, the difference between children with ASD and DD was not significant. For the 4–6-year-old group, the amount of time spent with parents had significant influence on the likelihood of negative emotional and behavioral change in two age groups. The longer the accompanying time the less likely the children had negative emotional and behavioral change (Figure 2).

Logistic regression was performed on children with ASD and DD to identify the factors that influenced each group. Table 4 shows the outcomes.

For the ASD group, the logistic regression model results showed that compared with parent-oriented intervention at home, ceasing rehabilitation had a higher likelihood of negative emotional and behavioral change (OR 1.67, 95% CI 1.41–1.98; Table 4) and having teachers' online support had a higher likelihood of negative emotional and behavioral change in children (OR 1.26, 95% CI 1.03–1.54; Table 4). Having children with comorbidities was associated with a more than one-fold likelihood of having negative emotional and behavioral change in children (OR 1.59; 95% CI 1.33–1.90; Table 4). If the accompanying time did not change or was longer than it previously had been, the likelihood

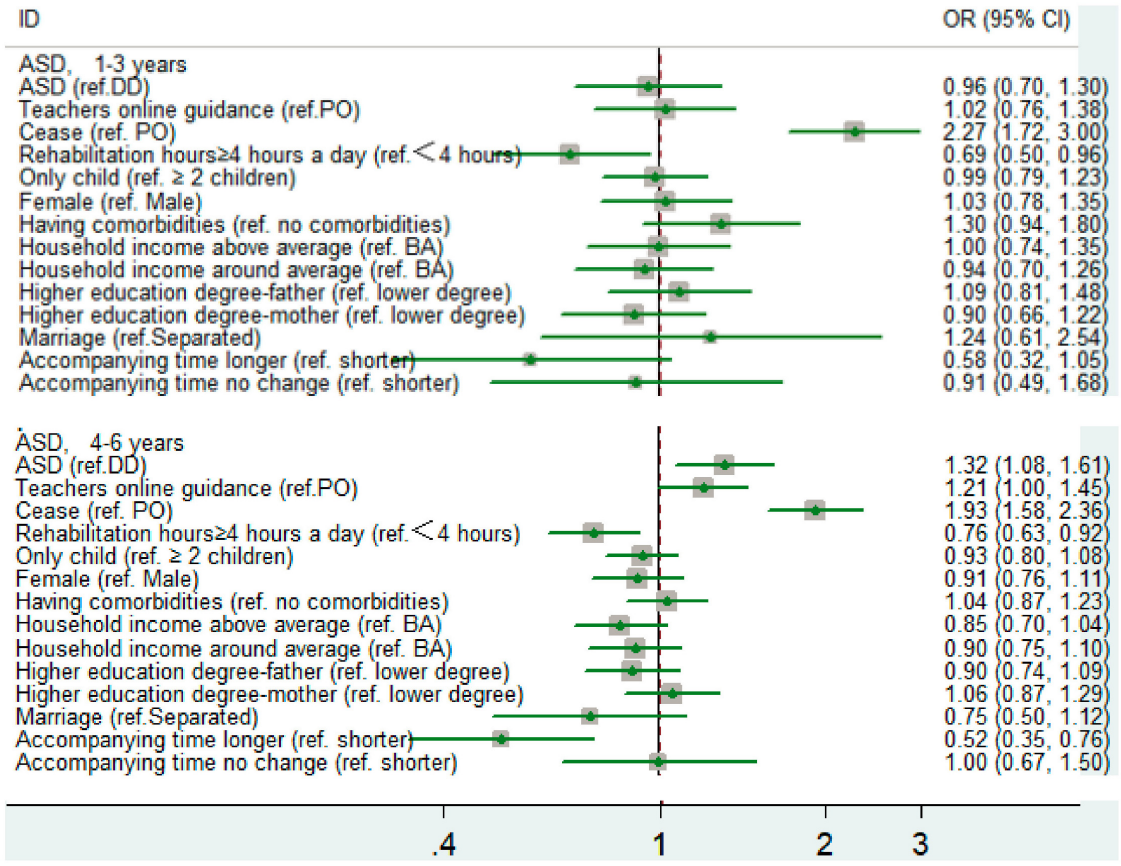


FIGURE 2
The odds of negative emotional and behavioral change in two group.

of negative emotional and behavioral changes occurring was lower (OR 0.40; 95% CI 0.29–0.55 for families where time spent did not change and OR 0.35; 95% CI 0.26–0.48 for families where time spent was longer than previously; see Table 4). Child gender, age, number of children in the household, parents' education, and household income were not significantly associated with negative emotional and behavioral change in children (Table 4).

For the DD group, however, only ceasing rehabilitation had a higher likelihood of negative emotional and behavioral change (OR 1.95, 95% CI 1.33–2.88; Table 4).

What is needed to deal with conditions of the pandemic

A total of 4,849 survey participants responded to the open question about what could be done to help deal with the ongoing pandemic. The most commonly reported requirement was more professional one-to-one online support (53.93%), followed by financial support (28.05%), online knowledge training (16.33%), and others (1.69%); see Table 5.

In the ASD group, more professional one-to-one online support was needed by a higher percentage of parents compared to the DD group.

Discussion

This was the first study in China to use a national survey to investigate COVID-19's negative impact on children with ASD and DD. This study yielded several key findings.

First, we discovered that 22.31% of children with ASD exhibited increased emotional and behavioral issues during the lockdown. For the children with DD, 18% experienced increased emotional and behavioral problems. According to the theory of mind, the age of 4–5 years old is critical for the development of social abilities (25, 26). However, governments and policymakers all over the world, including China, chose school closure and home confinement as two necessary measures to limit the spread of COVID-19 infection, making it impossible for preschoolers with ASD and DD to improve their ability during their critical developmental period. Child isolation was detrimental to the emotional wellbeing of children with developmental disabilities (27, 28). Due to COVID-19, home schooling occurred involuntarily and was associated with feelings of loneliness, negatively impacting the children's mental health and worsening pre-existing psychiatric disorders and behavioral problems (14, 23, 29). Our findings were slightly lower than other studies; for example, 45% of the sample had a worsening of their pre-existing psychiatric disorder (30), and 35.5% and 41.5% of children with ASD presented with more intense and frequent behavior problems, respectively (31). On the one hand, we used

10.3389/fpsyt.2023.1134396

TABLE 4 Logistic regression analysis for variables predicting the negative emotional and behavioral change* of children in two groups.

Characteristics	ASD group (n = 4,138)			DD group (n = 711)		
	Odds ratio	95% CI		Odds ratio	95% CI	
Rehabilitation type						
Parent oriented	1.00	reference		1.00	reference	
Teachers' online support	1.26	1.03	1.54	0.80	0.45	1.42
Cease	1.72	1.44	2.07	1.95	1.33	2.88
Rehabilitation hours						
< 4 h a day	1.00	reference		1.00	reference	
≥ 4 h a day	0.84	0.67	1.05	0.91	0.46	1.78
Only children						
No	1.00	reference		1.00	reference	
Yes	0.97	0.84	1.13	0.91	0.61	1.36
Gender of child						
Male	1.00	reference		1.00	reference	
Female	1.00	0.81	1.22	1.13	0.72	1.78
Age of child						
1–3 years	1.00	reference		1.00	reference	
4–6 years	1.15	0.97	1.35	1.11	0.72	1.71
Comorbidity[a]						
No	1.00	reference		1.00	reference	
Yes	1.59	1.33	1.90	1.28	0.82	2.00
Household income						
Below average	1.00	reference		1.00	reference	
Around average	0.84	0.69	1.02	0.74	0.45	1.22
Above average	0.83	0.68	1.01	0.72	0.43	1.22
Higher education degree-father						
No	1.00	reference		1.00	reference	
Yes	1.06	0.86	1.30	1.02	0.61	1.72
Higher education degree-mother						
No	1.00	reference		1.00	reference	
Yes	1.13	0.92	1.39	0.94	0.55	1.59
Time spent between parents and children						
Shorter	1.00	reference		1.00	reference	
No change	0.40	0.29	0.55	0.62	0.19	2.04
Longer	0.35	0.26	0.48	0.80	0.25	2.54

*Emotional and behavioral change is "worse" in the survey. ASD, autism spectrum disorder; DD, developmental delay. [a]Including intellectual disabilities (ID) and attention deficit and hyperactivity disorder (ADHD).

a larger sample, which may reduce the percentage of negative outcomes; on the other hand, differences between countries may cause some difference, as a Chinese small sample survey found that one-third of children's social and emotional status had worsened (32). In our study, a recall bias could lead to underestimation.

Second, the COVID-19 pandemic affected children with ASD differently than children with DD. Children with ASD had a higher likelihood of negative emotional and behavioral changes than children with DD, particularly those aged 3–6 years. The abrupt removal of resources during the first national lockdown, as well as the prolonged isolation, may have had a disproportionate impact on this vulnerable group, putting them at a higher risk of behavioral problems exacerbations (33). DD is defined as having a significantly lower-than-average intellectual disability, and research shows that, when compared to children and adolescents with intellectual disabilities, children and adolescents with ASD have significantly higher rates of behavioral and emotional problems (34). ASD is more severe when compared to other disabilities. Individuals with ASD have differences in receptive communication skills and may experience delays in processing information (35),

TABLE 5 Responses to the open-ended question about what could be of help to deal with the conditions of the ongoing pandemic.

	ALL	ASD	DD
Financial support	1,360 (28.05)	1,134 (27.40)	226 (31.79)
Professional one-to-one online support	2,615 (53.93)	2,251 (54.40)	364 (51.20)
Online knowledge training	792 (16.33)	684 (16.53)	108 (15.19)
Others	82 (1.69)	69 (1.67)	13 (1.83)
Sum	4,849 (100.00)	4,138 (100.00)	711 (100.00)

ASD, autism spectrum disorder; DD, developmental delay.

which affects their ability to respond to the pandemic in an accepting and efficient manner (8, 36). Our findings add to the growing body of evidence that people with severe disabilities, such as ASD, are disproportionately affected by negative events (37, 38).

Third, some families stopped recovering, but most of the families were undertaking the rehabilitation recovery (77.6% for ASD group vs. 72.86% for DD group). The implementation of an online form of education during the COVID-19 pandemic was less than optimal for children. The literature on the efficacy of telehealth for individuals with ASD is mixed (39, 40). The implementation of home education presents its challenges, even for the professional developmental behavioral pediatrician (8). Home education for children with special needs should be more focused on parental roles (41), as the online advice from others only decreases parents' responsibilities. For children who rely on gestures or picture exchange, they often require a parent or additional support to help them to communicate responses. This study also collected parents' narratives on their perceived needs throughout the pandemic through an open-ended question. About half of the participants reported needing one-to-one support and guidance from professionals online, where the answers were consistent with those of earlier studies that stated that healthcare services, especially in-home services, were badly needed (31); this also reflected that those parents were not satisfied with the current online support. Delivery of programs that are easily implemented and meet the needs of children and their families is needed (42). Although clinical trials and systematic reviews have shown telehealth interventions to result in promising improvements in learning under the circumstances of limited access to in-person services, we have not obtained optimistic results in the case of a real scenario (COVID-19). It also reminds us of the differences between the real world and experiments.

Fourth, we found that low family income increased the risk for negative changes in children's emotions and behavior. This finding aligned with recent research highlighting significant socioeconomic health inequities in other populations (e.g., adults) during the pandemic (43, 44). Parents with low income who have children with ASD may be more vulnerable to employment loss and may experience greater challenges finding care for their child. This in turn can exacerbate behavioral challenges in these children, adding to the myriad of challenges already faced by caregivers (8, 45, 46).

Fifth, the increase in time spent with the child was conducive to the improvement of the emotional functions and skills of children with ASD during the pandemic. With long-term restrictions at home, parents had more time to spend with their children, thus more family activities could be done. The results of the British Millennium Cohort study found that in the early development of children, a series of activities with parents and children had a significant effect on children's cognitive development (47). Long-term home life also increases the chances of home language input. Home language input is positively correlated with children's cognitive ability and vocabulary diversity (48). This had a positive effect on the development of children with ASD. But for children with DD, we did not find the accompanying time as an influence. This finding showed that different factors play a role in different kinds of developmental disorders.

Limitations

First, the sample is drawn from a network survey. All families were invited to participate in this study and completed an electronic questionnaire, which did not allow for control stratification in sampling. Although the sex ratio and family location distribution of this study were consistent with the main data of the Second National Survey of Disabled People (SNSDP), we should adopt more rigorous sampling methods to improve our research in the future. Second, the data's accuracy should be considered. A considerable amount of information was in the form of parents self-reporting, which might deviate from the real situation. Our questionnaire did not assess whether worsening changes reached a level of clinical significance or represented normative variations over time. Third, more studies following the cohorts of children longitudinally should be conducted to understand the long-term impacts of the COVID-19 pandemic on the functioning of children with ASD and DD as well as on their families. Fourth, the impacts of specific comorbidities of the children and parental genders were not further discussed in this article. In the future, better classification of comorbidities and respondent bias caused by gender will aid in the development of more appropriate procedures.

Conclusion

In conclusion, the current survey study indicates that the ongoing COVID-19 pandemic has resulted in a difficult period for the majority of families with preschoolers with ASD and DD, showing increased emotional and behavioral problems. During the COVID-19 pandemic, the implementation of online education was less than ideal for children and even teachers' online instruction was not as effective as parental-oriented instruction. More professional one-to-one online support and psychiatric care should be made available in the future. Our findings could help shape future policies and interventions to reduce the negative effects of COVID-19 on young people with developmental disorders and their families. More importantly, we should better evaluate the impact of policies on people's health, take timely measures to protect the interests of vulnerable groups, and reduce health inequities.

Data availability statement

The raw data supporting the conclusions of this article will be made available by the authors, without undue reservation.

Ethics statement

The studies involving human participants were reviewed and approved by the Ethics Committee of Peking University Institutional Review Board and approval number is IRB00001052-20016. Written informed consent to participate in this study was provided by the participants or their legal guardian/next of kin.

Author contributions

YZ conceptualized and designed the study, drafted the initial manuscript, reviewed, and revised the manuscript. XZ and RZ designed the data collection instruments, collected the data, reviewed, and revised the manuscript. YL coordinated and supervised the data collection and critically reviewed the manuscript for important intellectual content. All authors approved the final manuscript as submitted and agreed to be accountable for all aspects of the work.

Funding

All phases of this study were supported by Beijing Municipal Science and Technology Commission grant, Z181100001518005 and the Key Realm R&D Program of Guangdong Province, 2019B030335001.

Acknowledgments

We thank the Liu Daiyue and Xu Disha at ALSOLIFE for their contribution.

Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

Publisher's note

All claims expressed in this article are solely those of the authors and do not necessarily represent those of their affiliated organizations, or those of the publisher, the editors and the reviewers. Any product that may be evaluated in this article, or claim that may be made by its manufacturer, is not guaranteed or endorsed by the publisher.

Supplementary material

The Supplementary Material for this article can be found online at: https://www.frontiersin.org/articles/10.3389/fpsyt.2023.1134396/full#supplementary-material

References

1. Lima CKT, Carvalho PMM, Lima IAAS, Nunes JVAO, Saraiva JS, de Souza RI, et al. The emotional impact of coronavirus 2019-nCoV (new coronavirus disease). *Psychiatry Res.* (2020) 287:112915. doi: 10.1016/j.psychres.2020.112915

2. Yao H, Chen JH, Xu YF. Patients with mental health disorders in the COVID-19 epidemic. *Lancet Psychiatry.* (2020) 7:e21. doi: 10.1016/S2215-0366(20)30090-0

3. Cahapay MB. How Filipino parents home educate their children with autism during COVID-19 period. *Int J Dev Disabil.* (2020) 68:395–8. doi: 10.1080/20473869.2020.1780554

4. Eshraghi AA, Li C, Alessandri M, Messinger DS, Eshraghi RS, Mittal R, et al. COVID-19: overcoming the challenges faced by individuals with autism and their families. *Lancet Psychiatry.* (2020) 7:481–3. doi: 10.1016/S2215-0366(20)30197-8

5. Majoko T, Dudu A. Parents' strategies for home educating their children with Autism Spectrum Disorder during the COVID-19 period in Zimbabwe. *Int J Dev Disabil.* (2020) 68:474–8. doi: 10.1080/20473869.2020

6. Degli Espinosa F, Metko A, Raimondi M, Impenna M, Scognamiglio EA. Model of support for families of children with autism living in the COVID-19 lockdown: lessons from Italy. *Behav Anal Pract.* (2020) 13:550–8. doi: 10.1007/s40617-020-00438-7

7. Narzisi A. Handle the autism spectrum condition during coronavirus (COVID-19) stay at home period: ten tips for helping parents and caregivers of young children. *Brain Sci.* (2020) 10:207. doi: 10.3390/brainsci10040207

8. Aishworiya R, Kang YQ. Including children with developmental disabilities in the equation during this COVID-19 pandemic. *J Autism Dev Disord.* (2021) 51:2155–8. doi: 10.1007/s10803-020-04670-6

9. Laupacis A. Working together to contain and manage COVID-19. *CMAJ.* (2020) 192:E340–1. doi: 10.1503/cmaj.200428

10. American Psychiatric Association. *Diagnostic and Statistical Manual of Mental Disorders*. 5th ed. Arlington, VA: American Psychiatric Association Press (2013).

11. Battaglia A, Bianchini E, Carey JC. Diagnostic yield of the comprehensive assessment of developmental delay/mental retardation in an institute of child neuropsychiatry. *Am J Med Genet.* (1999) 82:60–6.

12. Saigal S, Doyle LW. An overview of mortality and sequelae of preterm birth from infancy to adulthood. *Lancet.* (2008) 371:261–9. doi: 10.1016/S0140-6736(08)60136-1

13. Duan L, Shao X, Wang Y, Huang Y, Miao J, Yang X, et al. An investigation of mental health status of children and adolescents in china during the outbreak of COVID-19. *J Affect Disord.* (2020) 275:112–8. doi: 10.1016/j.jad.2020.06.029

14. Petruzzelli MG, Furente F, Colacicco G, Annecchini F, Margari A, Gabellone A, et al. Implication of COVID-19 pandemic on adolescent mental health: an analysis of the psychiatric counseling from the emergency room of an Italian university hospital in the years 2019-2021. *J Clin Med.* (2022) 11:6177. doi: 10.3390/jcm11206177

15. Racine N, McArthur BA, Cooke JE, Eirich R, Zhu J, Madigan S. Global prevalence of depressive and anxiety symptoms in children and adolescents during COVID-19: a meta-analysis. *JAMA Pediatr.* (2021) 175:1142–50. doi: 10.1001/jamapediatrics.2021.2482

16. Ammerman BA, Burke TA, Jacobucci R, McClure K. Preliminary investigation of the association between COVID-19 and suicidal thoughts and behaviors in the U.S. *J Psychiatr Res.* (2021) 134:32–8. doi: 10.1016/j.jpsychires.2020.12.037

17. Hubbard K, Trauner DA. Intonation and emotion in autistic spectrum disorders. *J Psycholinguist Res.* (2007) 36:159–73. doi: 10.1007/s10936-006-9037-4

18. Rattaz C, Dubois A, Michelon C, Viellard M, Poinso F, Baghdadli A. How do children with autism spectrum disorders express pain? A comparison with

developmentally delayed and typically developing children. *Pain.* (2013) 154:2007–13. doi: 10.1016/j.pain.2013.06.011

19. Lee J. Mental health effects of school closures during COVID-19. *Lancet Child Adolesc Health.* (2020) 4:421. doi: 10.1016/S2352-4642(20)30109-7

20. Asbury K, Fox L, Deniz E, Code A, Toseeb U. How is COVID-19 affecting the mental health of children with special educational needs and disabilities and their families? *J Autism Dev Disord.* (2021) 51:1772–80. doi: 10.1007/s10803-020-04577-2

21. Zhou H, Xu X, Yan W, Zou X, Wu L, Luo X, et al. Prevalence of autism spectrum disorder in China: a nationwide multi-center population-based study among children aged 6 to 12 years. *Neurosci Bull.* (2020) 36:961–71. doi: 10.1007/s12264-020-00530-6

22. Yang Y, Shi L, Jin X, Tong S. Association of perinatal factors with suspected developmental delay in urban children aged 1-36 months–a large-scale cross-sectional study in China. *BMC Pediatr.* (2023) 23:11. doi: 10.1186/s12887-022-03819-9

23. Brooks SK, Webster RK, Smith LE, Woodland L, Wessely S, Greenberg N, et al. The psychological impact of quarantine and how to reduce it: rapid review of the evidence. *Lancet.* (2020) 395:912–20. doi: 10.1016/S0140-6736(20)30460-8

24. Luo Y, Zhao Y, Wang Y, Liang R, Hong C, Yang Y, et al. Characteristics and treatment patterns of autism spectrum disorder in China, 2020. *Psychiatry Res.* (2022) 317:114879. doi: 10.1016/j.psychres.2022.114879

25. Korkmaz B. Theory of mind and neurodevelopmental disorders of childhood. *Pediatr Res.* (2011) 69(Pt 2):101R–8R. doi: 10.1203/PDR.0b013e318212c177

26. Chevallier C, Volkmar RP, Pelphrey K (editors). *Encyclopedia of Autism Spectrum Disorders*. New York, NY: Springer (2013). p. 3111–5.

27. Allen K, Kern ML, Vella-Brodrick D, Hattie J, Waters L. What schools need to know about fostering school belonging: a meta-analysis. *Educ Psychol Rev.* (2018) 30:1–34.

28. Jose PE, Ryan N, Pryor J. Does social connectedness promote a greater sense of well-being in adolescence over time? *J Res Adolesc.* (2012) 22:235–51. doi: 10.1111/famp.12514

29. Loades ME, Chatburn E, Higson-Sweeney N, Reynolds S, Shafran R, Brigden A, et al. Rapid systematic review: the impact of social isolation and loneliness on the mental health of children and adolescents in the context of COVID-19. *J Am Acad Child Adolesc Psychiatry.* (2020) 59:1218–39.e3. doi: 10.1016/j.jaac.2020.05.009

30. Vasa RA, Singh V, Holingue C, Kalb LG, Jang Y, Keefer A. Psychiatric problems during the COVID-19 pandemic in children with autism spectrum disorder. *Autism Res.* (2021) 14:2113–9. doi: 10.1002/aur.2574

31. Colizzi M, Sironi E, Antonini F, Ciceri ML, Bovo C, Zoccante L. Psychosocial and behavioral impact of COVID-19 in autism spectrum disorder: an online parent survey. *Brain Sci.* (2020) 10:341. doi: 10.3390/brainsci10060341

32. Huang S, Sun T, Zhu Y, Song S, Zhang J, Huang L, et al. Impact of the COVID-19 pandemic on children with ASD and their families: an online survey in China. *Psychol Res Behav Manag.* (2021) 14:289–97. doi: 10.2147/PRBM.S293426

33. Summers J, Baribeau D, Mockford M, Goldhopf L, Ambrozewicz P, Szatmari P, et al. Supporting children with neurodevelopmental disorders during the COVID-19 pandemic. *J Am Acad Child Adolesc Psychiatry.* (2021) 60:2–6. doi: 10.1016/j.jaac.2020.09.011

34. Tonge BJ, Einfeld SL. Psychopathology and intellectual disability: the Australian child to adult longitudinal study. *Int Rev Res Ment Retard.* (2003) 26:61–91.

35. Wallace GL, Kenworthy L, Pugliese CE, Popal HS, White EI, Brodsky E, et al. Real-world executive functions in adults with autism spectrum disorder: profiles of impairment and associations with adaptive functioning and co-morbid anxiety and depression. *J Autism Dev Disord.* (2016) 46:1071–83. doi: 10.1007/s10803-015-2655-7

36. Baweja R, Brown SL, Edwards EM, Murray MJ. COVID-19 pandemic and impact on patients with autism spectrum disorder. *J Autism Dev Disord.* (2022) 52:473–82. doi: 10.1007/s10803-021-04950-9

37. Campbell VA, Gilyard JA, Sinclair L, Sternberg T, Kailes JI. Preparing for and responding to pandemic influenza: implications for people with disabilities. *Am J Public Health.* (2009) 99(Suppl. 2):S294–300. doi: 10.2105/AJPH.2009.162677

38. Neece C, McIntyre LL, Fenning R. Examining the impact of COVID-19 in ethnically diverse families with young children with intellectual and developmental disabilities. *J Intellect Disabil Res.* (2020) 64:739–49. doi: 10.1111/jir.12769

39. Hepburn SL, Blakeley-Smith A, Wolff B, Reaven JA. Telehealth delivery of cognitive-behavioral intervention to youth with autism spectrum disorder and anxiety: a pilot study. *Autism.* (2016) 20:207–18. doi: 10.1177/1362361315575164

40. Knutsen J, Wolfe A, Burke BL, Hepburn S, Lindgren S, Coury D. A systematic review of telemedicine in autism spectrum disorders. *J Autism Dev Disord.* (2016) 3:330–44.

41. Jolly JL, Matthews MS, Nester J. Homeschooling the gifted: a parent's perspective. *Gifted Child Q.* (2013) 57:121–34.

42. Smile SC. Supporting children with autism spectrum disorder in the face of the COVID-19 pandemic. *CMAJ.* (2020) 192:E587. doi: 10.1503/cmaj.75399

43. Ettman, CK, Abdalla SM, Cohen GH, Sampson L, Vivier PM, Galea S. Low assets and financial stressors associated with higher depression during COVID-19 in a nationally representative sample of US adults. *J Epidemiol Commun Health.* (2020) 1–8. doi: 10.1136/jech-2020-215213

44. Rudenstine S, McNeal K, Schulder T, Ettman CK, Hernandez M, Gvozdieva K, et al. Depression and anxiety during the COVID-19 pandemic in an urban, low-income public university sample. *J Trauma Stress.* (2021) 34:12–22. doi: 10.1002/jts.22600

45. Shafran R, Coughtrey A, Whittal M. Recognising and addressing the impact of COVID-19 on obsessive-compulsive disorder. *Lancet Psychiatry.* (2020) 7:570–2. doi: 10.1016/S2215-0366(20)30222-4

46. Davide P, Andrea P, Martina O, Andrea E, Davide D, Mario A. The impact of the COVID-19 pandemic on patients with OCD: effects of contamination symptoms and remission state before the quarantine in a preliminary naturalistic study. *Psychiatry Res.* (2020) 291:113213. doi: 10.1016/j.psychres.2020.113213

47. Hernández-Alava M, Popli G. Children's development and parental input: evidence from the UK millennium cohort study. *Demography.* (2017) 54:485–511. doi: 10.1007/s13524-017-0554-6

48. d'Apice K, Latham RM, von Stumm S. A naturalistic home observational approach to children's language, cognition, and behavior. *Dev Psychol.* (2019) 55:1414–27. doi: 10.1037/dev0000733

frontiers | Frontiers in Psychology

TYPE Original Research
PUBLISHED 03 August 2023
DOI 10.3389/fpsyg.2023.1098449

OPEN ACCESS

EDITED BY
Ru Ying Cai,
Autism Spectrum Australia, Australia

REVIEWED BY
Fabrizio Stasolla,
Giustino Fortunato University, Italy
Mary Hanley,
Durham University, United Kingdom

*CORRESPONDENCE
Brianna N. Young
✉ youngbn@uwm.edu

RECEIVED 17 November 2022
ACCEPTED 13 June 2023
PUBLISHED 03 August 2023

CITATION
Young BN, Mohanty E, Levine K and Klein-Tasman BP (2023) Addressing fears of children with Williams syndrome: therapist and child behavior in the context of a novel play-and humor-infused exposure therapy approach. *Front. Psychol.* 14:1098449. doi: 10.3389/fpsyg.2023.1098449

Addressing fears of children with Williams syndrome: therapist and child behavior in the context of a novel play-and humor-infused exposure therapy approach

Brianna N. Young[1]*, Ellora Mohanty[1], Karen Levine[2] and Bonita P. Klein-Tasman[1]

[1]Child Neurodevelopment Research Lab, Department of Psychology, University of Wisconsin-Milwaukee, Milwaukee, WI, United States, [2]Department of Psychiatry, Harvard Medical School, Boston, MA, United States

Many children with Williams syndrome struggle with fears and phobias that significantly impact their daily lives. Yet, there is sparse literature about the impact of behavioral interventions to treat anxiety and phobias among children with Williams syndrome. Using observational coding of intervention videos, the current study examines patterns of the therapist's use of play and humor and relations to child behavioral responses for four children with Williams syndrome who were identified as treatment responders to humor- and play-infused exposure therapy for fears and anxieties. Sessions were coded for therapist behaviors (exposure with or without play/humor, stimulus type used during exposure, passive or invited attention to feared stimulus, and spontaneous parent participation in exposure) as well as positive, negative, and neutral child behaviors (verbalizations and behaviors). Temporal patterns between therapist and child behaviors were analyzed using lag sequential analyses. The results showed that tolerance of feared stimuli improved for two of the four children following this play- and humor-infused exposure therapy approach, and the remaining two participants demonstrated progress beyond tolerating the feared stimulus and showed increased positive behaviors with the feared stimulus across sessions. Findings also showed patterns of therapist attunement to the child's anxiety level demonstrated through efforts to flexibly adjust the degrees of exposure. Therapist-initiated invited attention behaviors, indicative of the therapist's use of narration and priming, were associated with child tolerance and positive behaviors during exposure to the feared stimulus. Limitations of this study include a very small sample size, short duration of intervention, and a single-subject research design, which limit the generalizability of findings. Implications and future directions of this research are discussed.

KEYWORDS

Williams syndrome, children, phobias, anxiety, play therapy, exposure therapy

Introduction

Williams syndrome is a rare neurodevelopmental disorder caused by a hemizygous microdeletion on chromosome 7q11.23 and characterized by physical, cognitive, and behavioral markers (Mervis and John, 2010). Williams syndrome has a prevalence of 1 in every 7,500 births and with roughly equal rates across males and females (Strømme et al., 2002). People with Williams syndrome have distinct facial features, cardiovascular complications, and often co-occurring mild-to-moderate intellectual disability, attention

problems, and anxiety (Mervis, 2000; Collins et al., 2010). Children with Williams syndrome also exhibit strong motivation for social interaction, indiscriminate friendliness, empathy for others (Järvinen et al., 2013), and relative strengths in verbal short-term memory and concrete expressive language (Mervis and John, 2010).

Anxiety is one of the most prevalent co-occurring conditions among people with Williams syndrome (Stinton et al., 2012; Royston et al., 2017; Ng-Cordell et al., 2018). A comprehensive examination of anxiety among children and adolescents ages 4–16 with Williams syndrome ($n = 119$) found that 53.8% met diagnostic criteria for specific phobia and 11.8% met criteria for generalized anxiety disorder (Leyfer et al., 2006). Furthermore, examination of the type of phobias experienced showed that loud noises were the most prevalent (27.7%) compared to common phobia types (e.g., situational, natural environment). Longitudinal research has provided evidence that anxiety and fears experienced by children and adolescents with Williams syndrome tend to persist over time, interfere with daily life, and may negatively impact socioemotional development, pointing to the need for interventions for children with Williams syndrome and co-occurring fears and anxiety (Einfeld et al., 1997, 1999, 2001; Woodruff-Borden et al., 2010).

Intervention studies with individuals with developmental and intellectual disabilities that specifically target anxiety, fears, or phobias are scant, with the majority of research conducted with autism spectrum disorder (ASD) populations (Kreslins et al., 2015; Ung et al., 2015). CBT interventions have demonstrated some effectiveness for individuals with higher functioning ASD (Ung et al., 2015), but few studies have tested the effectiveness of behavioral interventions on specific fears among people with ASD and co-occurring intellectual and developmental disabilities (Rosen et al., 2016). Single-subject design studies utilized systematic desensitization, graduated exposure, reinforcement procedures (some implemented by parents), modeling, hierarchy/stimulus fading, among other behavioral strategies (Rosen et al., 2016). Notably Koegel et al. (2004) conducted a within-subjects study targeting auditory-based fears (fear of toilet-flushing, animal toy sounds, and noises related to vacuum cleaners, blenders, and hand mixers) with three children with ASD and co-occurring developmental and/or intellectual disabilities. The results showed that all three children in this study were comfortable in the presence of their previously feared stimuli following the systematic desensitization intervention and at follow-up (based on child progress through their fear hierarchies) (Koegel et al., 2004). The treatment of anxiety and co-occurring intellectual disability is largely understudied. Case studies provide some evidence of effectiveness of graded exposure, response prevention, psychoeducation, and relaxation techniques to treat specific phobias in adults with intellectual disabilities (Hurley, 2004; Cowdrey and Walz, 2015; Dagnan et al., 2018). Further treatment effectiveness research is warranted to address the needs of individuals with intellectual disabilities and co-occurring anxiety.

Treatment studies with people with Williams syndrome and anxiety are exiguous, with the vast majority including case studies with small samples (1–3 participants) (Klein-Tasman and Albano, 2007; Phillips and Klein-Tasman, 2009; Conelea and Klein-Tasman, 2013). These case reports demonstrate some success treating anxiety symptoms using CBT-based interventions, with a tailored emphasis on behavioral aspects such as repeated practice of skills and role-playing replacement behaviors and less emphasis on cognitive components such as cognitive restructuring. Essau and Longhi (2013) found improvements in emotional and social skills with reduced anxiety following a case study analysis using a six-session CBT-based approach that taught skills connecting thoughts, emotions, and behaviors: Emotional and Social Skills Training for Individuals with Williams Syndrome (ESST-WS). Recent research of an adapted virtually delivered CBT-based group intervention for adults with Williams syndrome and anxiety ($n = 4$) showed promise as a feasible and effective approach (Thom et al., 2022). In sum, the research literature on evidence-based interventions with children and adults with Williams syndrome and co-occurring anxiety is remarkably scant, with few published studies, none with younger children, and none (outside very recent work by our group) specifically focused on fears and phobias.

In younger children with Williams syndrome, fears are a central manifestation of anxiety. While for typically developing children anxiety symptoms relating to specific phobias decrease over time with development (Costello et al., 2011), children with Williams syndrome and/or co-occurring intellectual disability are likely to experience worsening anxiety symptoms across development (Maiano et al., 2018; Ng-Cordell et al., 2018). Cognitive behavioral interventions have been identified as empirically supported approaches to addressing fears and phobias in typically developing children (Hirshfeld-Becker et al., 2010; Ollendick and Davis, 2013; Whiteside et al., 2020) and have also shown benefit for children with ASD (Davis et al., 2007; Ollendick et al., 2021) and children with intellectual disabilities (Hagopian et al., 2001; Moskowitz et al., 2019; Dovgan et al., 2020; Fodstad et al., 2021). Furthermore, evidence is accumulating that these interventions are also useful, with developmentally appropriate incorporation of play activities, with young typically developing children (Oar et al., 2015; Kershaw et al., 2017; Farrell et al., 2018). Leveraging the socially motivated tendencies of individuals with Williams syndrome (Mervis et al., 2001) by using social, adult–child play and humor in the context of a behavioral intervention may be useful to treat anxiety in children with Williams syndrome. We are engaged in a research program to examine the impact of play- and humor-infused exposure therapy on fears in children ages 4 through 10 with Williams syndrome and have described preliminary evidence of the utility of this approach (Klein-Tasman et al., 2022). The approach used is based in well-established graduated exposure (APA Division 12 Society of Clinical Psychology, 2022), uses functional behavior assessment to tailor treatment (Davis et al., 2009), and is informed by client-directed principles of outcome-informed therapy (Duncan and Miller, 2000). In our prior study, we demonstrated reductions in fear and anxiety (based on clinician and/or parent-report metrics) with four of the eight participants with complete data who participated in a brief, play- and humor-infused exposure-based intervention. Hence, this play- and humor-based behavioral approach shows promise as a developmentally attuned systematic desensitization intervention to treat fears and phobias in children

with Williams syndrome. However, one significant limitation of the prior work was reliance on therapist and caregiver ratings of improvement rather than direct observation.

The current study employs behavioral coding to examine patterns of therapist use of graduated exposure and systematic desensitization using play and humor, and child anxiety behaviors in a subset of children with Williams syndrome who showed a positive response to the brief intervention in the pilot study (Klein-Tasman et al., 2022). For the purposes of this study, fear was operationally defined as observable child behaviors that indicated the child was experiencing emotional and/or physiological responses reflecting reluctance to engage with or an aversion to the stimulus. We hypothesized that children with Williams syndrome in this sample would exhibit observable reductions in fear within and across sessions, facilitated by this play- and humor-infused exposure therapy approach. We anticipated that the therapist techniques (use of play and humor during exposures, gradual exposure up the fear hierarchy by stimulus type (media, toy, and actual), and invited attention and passive exposure) would facilitate reduced anxiety and improved regulation, evidenced by increased frequency of tolerating and/or positive behaviors in the presence of the feared stimulus.

Materials and methods

Participants

Participants were recruited through the Williams Syndrome Association's research registry as well as referrals from the PI of a largescale study of the cognitive and behavioral phenotype in Williams syndrome. Participants included four children with Williams syndrome who initially had high levels of anxiety and then showed reduced fear and anxiety based on parent and/or clinician report in our preliminary work (Klein-Tasman et al., 2022). Participants range from 4 to 9 years of age and include three boys and 1 girl. See Table 1 for participant characteristics. Inclusion criteria included a diagnosis of Williams syndrome (confirmed through prior genetic testing), age 4–10, the presence of fear or a strong emotional response to a definable stimulus that could feasibly be addressed in a university-based research laboratory, and English as the primary language spoken.

TABLE 1 Participant characteristics.

Participant pseudonym	Sex	Age (years; months)	GCA SS	Non-verbal reasoning SS	Verbal SS	ASD clinical diagnosis	Primary fear	Total sessions	Total sessions w/primary fear	Session length (range)
Ashton	M	6;8	63	79	85	No	Hand Dryer	3	2	23–35 min
Beth	F	6;5	82	108	83	No	Blenders	3	3	45–55 min
Colton	M	4;1	63	97	59	Yes	Blood Pressure	4	4	25–30 min
Danny	M	9;2	69	66	85	No	Vacuum	4	3	40–45 min

GCA, general cognitive ability; non-verbal and verbal standard scores (SS) on the DAS-II.

Measures

Background questionnaire

This parent-report questionnaire includes questions about child demographics, child's academic background, family information, birth and developmental history, medical history, and parent's primary concerns.

Differential Ability Scales – Second Edition

The Differential Ability Scales – Second Edition (DAS-II; Elliott, 2007) is a clinical measure of cognitive functioning for children ranging from age 2 years and 6 months to 17 years and 11 months. Children younger than age 9 were administered the Early

TABLE 2 Therapist behavior coding scheme.

Behavior code (Duration)	Operational definition
Therapist exposure type	
Pure exposure	The therapist presents the feared stimulus without play or humor (e.g., blood pressure cuff) with or without invited attention to the feared stimulus, or prompts the parent/ caregiver to present the child with the feared stimulus without play or humor.
Exposure with play and humor	The therapist presents the feared stimulus, with or without invited attention to the feared stimulus, with play and humor attempts to engage with the stimulus. This may be observed as a video playing in the background, and the therapist prompts play or humor (e.g., playful narrating techniques with humor) to engage with the media example. This may also be coded if the therapist prompts the parent/ caregiver to use play or humor with the child during exposure.
Stimulus type	
Media	Use of media version of feared stimulus in either pure exposure or exposure with play and humor. The therapist presents the child with a media example (image, video, or other media version) of the feared stimulus or the media example is present/ in view.
Toy	Use of a toy version of the feared stimulus in either pure exposure or exposure with play and humor. The therapist presents the child with a toy example (doll and toy vacuum cleaner) of the feared stimulus or the toy example is present/ in view.
Actual	Use of actual (realistic) version of feared stimulus in either pure exposure or exposure with play and humor. The therapist presents the child with their feared stimulus (e.g., blood pressure cuff) or the feared stimulus is present/ in view.
Attention to feared stimulus	
Invited attention	Therapist-initiated verbal or gestural cues or prompts attempting to direct the child's attention to the feared stimulus indicates presence of invited attention. Invited attention can also be observed in the form of therapist narration immediately before (primer) exposure, during exposure, or immediately after the exposure (within 3 seconds of exposure termination). Invited attention cannot occur during an independent child behavior.
Passive exposure	Invited attention toward another stimulus that is not the feared stimulus, thereby making the feared stimulus secondary to the invited attention stimulus.
Parent exposure type	
Pure exposure	The parent spontaneously presents the child with their feared stimulus, with or without invited attention to the exposure stimulus, without play or humor.
Exposure with play and humor	The parent spontaneously presents the feared stimulus, with or without invited attention to the exposure stimulus, with play and humor attempts to engage with the stimulus. This may be observed as a video playing in the background, and the parent prompts play or humor (e.g., playful narrating techniques with humor) to engage with the media example.

Years battery, and children older than age 8 were administered the School Age battery. This instrument is used to identify where a child's abilities fall in comparison with their peers as well as an individual profile of their cognitive strengths and weakness. Scores are represented as standard scores. See Table 1 for participant scores.

Procedure

The play- and humor-infused exposure-based intervention

The therapy approach is summarized as follows: The therapist provides a space for free play with toys that are tailored to the child's interests. The therapist encourages the parent(s) and caregiver(s) to participate and/or observe throughout sessions. After the child acclimates to the therapy room and happily plays together with the therapist, the therapist begins introducing the feared stimulus using the least intimidating level of exposure, which may be talking about the item, showing video of the item, or incorporating some aspect or representation of the item into the adult–child play (e.g., a pretend form of the item) and gauges the intensity of their reaction. As the child exhibits increased comfort in the presence of the feared stimulus, the therapist uses graduated exposure techniques to work up a fear ladder (based on functional assessment discussion with the parent prior to the intervention) toward direct contact with the feared stimulus. The therapist flexibly moves up and down levels of the fear hierarchy depending on the child's response following presentation or encouraged engagement with the feared stimulus. Throughout the sessions, socially engaging interactive play with the child is the consistent backdrop to help maintain contact and positive engagement with the feared stimulus. When anxiety becomes heightened as exposures incrementally move toward direct contact with a phobic stimulus, the therapist is likely to use socially attuned humor with the child (e.g., playful exaggerated emotion as a form of systematic desensitization) to help co-regulate, that is, to help the child sustain an emotionally regulated state, and to align themselves with the child, model emotional expression, and introduce a new emotional tone to interaction with the feared stimulus. In summary, the therapist spontaneously decreases the degree of exposure and/or increases playfulness and humor if the child shows signs of dysregulation and then increases the degree of exposure if the child appears regulated. Across sessions, the goal is for the child to sustain an emotionally regulated state, as manifested by exhibiting fewer fearful and anxious reactions to their feared stimulus to reduce functional impairment.

Behavioral coding procedure

Archival video-recorded intervention sessions of four participants (total of 12 sessions) were coded and analyzed using Noldus Observer XT 15.0 software to determine how

TABLE 3 Child behavior coding scheme.

Behavior code	Operational definition
Negative behaviors	
Negative verbalizations	Child verbally protests the interaction with the feared stimulus (utterance) or otherwise makes sounds indicating discomfort during exposure, such as saying "No!", "I don't like that",etc.(vocalization).
Negative physical	Child displays a negative physical reaction to the feared stimulus, such as cowering, backing up, or placing their hands over their ears (i.e.,avoidance).
Neutral behaviors	
Tolerating presence of feared stimulus	Child does not display any observable positive or negative reaction to the exposure (feared stimulus), indicating tolerance of the feared stimulus.
Positive behaviors	
Positive verbalizations	Child verbally exhibits excitement or engagement with the feared stimulus through articulated speech (utterance) or produces verbal indication (vocalization), grunt, or other verbal approximation indicating excitement or enjoyment during exposure, such as saying, "I want to try", "wow!", etc.
Positive physical	Child displays a positive physical reaction to the feared stimulus, such as moving their body toward the feared stimulus or engaging in exposure with physical contact, which followed a therapist/parent/caregiver prompt or cue.

child fear and anxiety-related behaviors change within session and across sessions, and characterize therapist use of play and humor and exposure techniques in this therapy. Behavioral coding (systematically defining and identifying overt behaviors) was conducted based on the coding scheme described in Tables 2, 3.

Therapist behaviors were coded using duration and frequency recording. The duration of therapist use of play- and humor-infused exposure therapy and pure exposure was coded to identify patterns of therapist-led exposure by type, as well as spontaneous parent use of exposure by type. The use of different types of feared stimuli (media, toy, and actual) was also coded during pure or play- and humor-infused exposure therapy. Therapist behaviors also included passive and invited attention to the feared stimulus. For example, the therapist may direct attention to a non-feared stimulus (e.g., toy car), thereby making the feared stimulus secondary to the stimulus where attention is invited (i.e., passive exposure). If the therapist used verbal and/or non-verbal cues to guide the child's gaze toward the feared stimulus (e.g., "Hey want to check this out?"), this was coded as invited attention to the stimulus. See Table 2 for the detailed therapist behavior coding scheme.

Child behaviors were coded as frequency of positive, tolerating (i.e., neutral), and negative behaviors within intervals, to capture observable fear-related behaviors. Verbalizations (e.g., articulated speech or verbal approximations) and physical behaviors (e.g., physical approach or avoidance) were also coded. See Table 3 for the detailed child behavior coding scheme.

Data analytic approach

Interrater reliability process

Interrater reliability was calculated using Noldus Observer XT 15.0, including Cohen's kappa and agreement percentage. Cohen's kappa ranges from −1 to +1, and kappa values should be interpreted using the following ranges: Values of ≤0 suggest no interrater agreement, values of 0.01–0.20 suggest no agreement to slight agreement, values of 0.21–0.40 suggest fair agreement, values of 0.41–0.60 suggest moderate agreement, values of 0.61–0.80 suggest substantial agreement, and values of 0.81–1.00 suggest almost perfect interrater agreement. For the current study, interrater reliability was considered met when k ≥0.70 and agreement percentage ≥80%.

The first author and a graduate student trained in the coding scheme separately coded one intervention session video (of a participant that is not included in the current sample) to pilot the developed coding scheme. It was expected that the coding scheme would need revisions as the development of a behavior coding scheme is an iterative process, often requiring discussions about revisions and refinement of operational definitions (Chorney et al., 2015). Macrocoding (i.e., codes that may apply to the broader context of the behaviors) and microcoding approaches (i.e., codes that are more specific and time-intensive) to the coding scheme were considered during the revision process (Chorney et al., 2015). The two coders discussed areas of disagreement during the preliminary coding process and revised the coding scheme accordingly. When all child codes and all therapist codes were analyzed as respective groups, minimum kappa values were reached. For all child codes, agreement percentage for frequency recording was 87.90%, and k = 0.84. For all therapist codes, agreement percentage for frequency was 82.46%, and k = 0.79 and agreement percentage for duration was 92.69%, and k = 0.91. See Table 4 for detailed reliability coding.

Once interrater agreement of the behavior coding scheme was established (agreement ≥80% and/or k ≥0.70) with one intervention session video of a participant that is not included in the current sample, one randomly selected intervention video for each participant was coded to ensure continued interrater reliability for all participants in this sample. Once interrater reliability was established with four intervention session videos (one for each participant), series data trends of frequency and duration of behaviors (per session and across) were coded, graphed, and visually inspected. Based on the interrater reliability findings, we elected to concentrate on frequency of child behavior and duration and frequency of therapist behavior. Specifically, the number of child behavior occurrences in each session of each participant

TABLE 4 Interrater reliability.

Code	Preliminary reliability	Preliminary reliability	Ashton		Beth		Colton		Danny	
	Frequency	Duration*	Frequency	Duration*	Frequency	Duration*	Frequency	Duration*	Frequency	Duration*
Negative child behaviors	88.89 %	89.10%	80%	72.67%	92.86%	94.78%	85.71%	60.44%	NP	NP
Negative vocalizations	80%	96.61%	100%	55.58%	NP	NP	NP	NP	NP	NP
Negative utterances	100%	90.27%	0% **	0% **	80%	45.66%	100%	53.57%	NP	NP
Negative physical	84.62%	87.71%	100%	77.68%	95.65%	96.88%	83.33%	61.05%	NP	NP
Positive child behaviors	89.74%	94.06%	86.67%	56.67%	88.89%	74.19%	86.49%	90.15%	88.89%	75.16%
Positive vocalizations	88.89%	87.51%	100%	59.63%	NP	NP	83.33%	56.91%	NP	NP
Positive utterances	84.62%	89.38%	83.33%	95.71%	95.45%	90.87%	NP	NP	100%	91.67%
Positive physical	94.12%	98.56%	83.33%	37.38%	82.61%	71.75%	88.00%	94.22%	87.50%	74.56%
Neutral/Tolerating child behaviors	86.21%	65.26%	91.67%	86.18%	92.31%	79.21%	86.67%	80.57%	NP	NP
All child codes	**87.90%**	**71.78%**	**87.23%**	**65.41%**	**90.70%**	**81.44%**	**86.44%**	**83.79%**	**88.89%**	**75.16%**
Therapist – pure exposure	92.86%	84.45%	NP	NP	100%	95.51%	NP	NP	NP	NP
Therapist – play and humor	83.33%	98.05%	100%	99.15%	100%	95.25%	75.00%	100%	100%	88.56%
Therapist – stimulus type: media	89.47%	97.66%	NP	NP	100%	95.56%	NP	NP	NP	NP
Therapist – stimulus type: toy	50%	94.52%	50%	99.48%	100%	81.75%	100%	80.79%	100%	89.42%
Therapist – stimulus type: actual	66.67%	89.06%	NP	NP	66.67%	83.30%	66.67%	94.74%	100%	95.29%
Therapist – directed attention	88.41%	92.67%	72.22%	85.23%	89.58%	96.03%	94.44%	82.97%	61.54%	92.29%
Therapist – passive exposure	100%	86.51%	NP	NP	NP	NP	66.67%	85.28%	NP	NP
Parent – pure exposure	81.25%	95.23%	NP	NP	NP	NP	NP	NP	NP	NP
Parent – play and humor	N/A	N/A	NP	NP	NP	NP	100%	93.17%	NP	NP
All therapist codes	**82.46%**	**92.69%**	**70.83%**	**95.70%**	**91.78%**	**91.14%**	**80.43%**	**93.54%**	**68.75%**	**91.10%**

Interrater reliability values are reflected as percent agreement (%).

*Duration interrater reliability values are conservative, reflecting agreement on milliseconds(ms).

**Negative utterances disagreement was a disagreement of 1 observation (frequency) and 1 second(duration).

N/A: not applicable to the videocoded.

NP: behavior code not present in this session video.

10.3389/fpsyg.2023.1098449

TABLE 5 Description of fear hierarchies.

Participant pseudonym	Targeted fear	Fear hierarchy
Ashton	Hand dryers	1. Watched videos of a variety of hand dryers in action, with increasing volume levels 2. Used portable hand dryer in the therapy room to blow scarves, push toy school bus first with the therapist placing the objects 3. Over time used portable hand dryer in the therapy room to blow scarves, push toy school bus with the child placing objects 4. Visited hand dryers in nearby restrooms, taking along scarves to play with, and therapist facilitates exposures 5. Finally, used hand dryer in naturalistic setting (i.e., public restroom) on hands, and child initiating exposures
Beth	Blenders	1. Played with toy blender with therapist initiation 2. Played with toy blender without therapist initiation 3. Watched videos of varying blenders with increasing volume 4. Therapist placed actual blender in the therapy room (unplugged) in the child's view 5. Therapist activated actual blender in the therapy room (without content) for short durations, with child observing/listening from next door and eventually entering the room 6. Therapist activated actual blender in the therapy room (without content) for increasingly longer durations, with child observing/listening from next door and eventually entering the room 7. Therapist activated actual blender in the therapy room with ice/berries, with child observing from next door and eventually entering the room 8. Therapist activated actual blender while child was in the room 9. Therapist activated actual blender while child was in the room, with child in closer proximity to the blender (< 3′) 10. Therapist activated actual blender while child added berries 11. Child activated the blender
Colton	Blood pressure (BP) cuff	1. Therapist and child played with toy BP cuff on stuffed animal, with therapist initiating 2. Therapist and child played with real BP cuff on stuffed animal, with therapist initiating 3. Therapist and child played with toy BP cuff on stuffed animal, with child initiating 4. Therapist initiated play with toy BP cuff on therapist (e.g., arms and legs) 5. Child initiated play with toy BP cuff on therapist (e.g., arms and legs) 6. Therapist initiated play with real BP cuff on therapist 7. Child initiated or joined play with real BP cuff on therapist 8. Therapist initiated play with toy BP cuff on child 9. Therapist initiated play with real BP cuff on child
Danny	Vacuum cleaners	1. Therapist initiated play with toy vacuum cleaner without sound 2. Child initiated play with toy vacuum cleaner with sound 3. Watched videos of varying vacuum cleaners with increasing volume 4. Therapist presented (actual) vacuum cleaner in the room, unplugged 5. Therapist presented (actual) vacuum cleaner in the room plugged in but turned off. 6. Therapist initiated play with (actual) vacuum cleaner (turned on) in the therapy room with the child in a separate room 7. Therapist initiated play with (actual) vacuum cleaner (turned on) in the hallway with the child seated on rolling chair and gradually brought closer (with child's permission) by throwing a stuffed animal closer and closer toward the vacuum cleaner 8. Therapist playful use of (actual) vacuum cleaner (modeling) to "vacuum" up bubbles 9. Therapist initialed playful use of (actual) vacuum cleaner by the child to "vacuum" up bubbles 10. Child initiated use of (actual) vacuum cleaner (by the child) to vacuum debris from the carpet

(frequency) was calculated within 5-minute intervals and graphed and visually inspected to demonstrate patterns of change. Similarly, the duration and frequency of therapist behaviors (e.g., use of play and humor) in each session were coded and calculated within 5-minute intervals and graphed and visually inspected. Lag sequential analyses (event state lag of 1) were also conducted to identify sequential patterns between therapist and child behaviors as well as sequential therapist behaviors (e.g., invited attention followed by exposure with play and humor).

Results

Child demographics and overview of course of treatment

Each of the four participants' demographic information as well as verbal and non-verbal norm-referenced cognitive functioning are displayed in Table 1. The fear hierarchy (i.e., course of treatment) for each participant is displayed in Table 5.

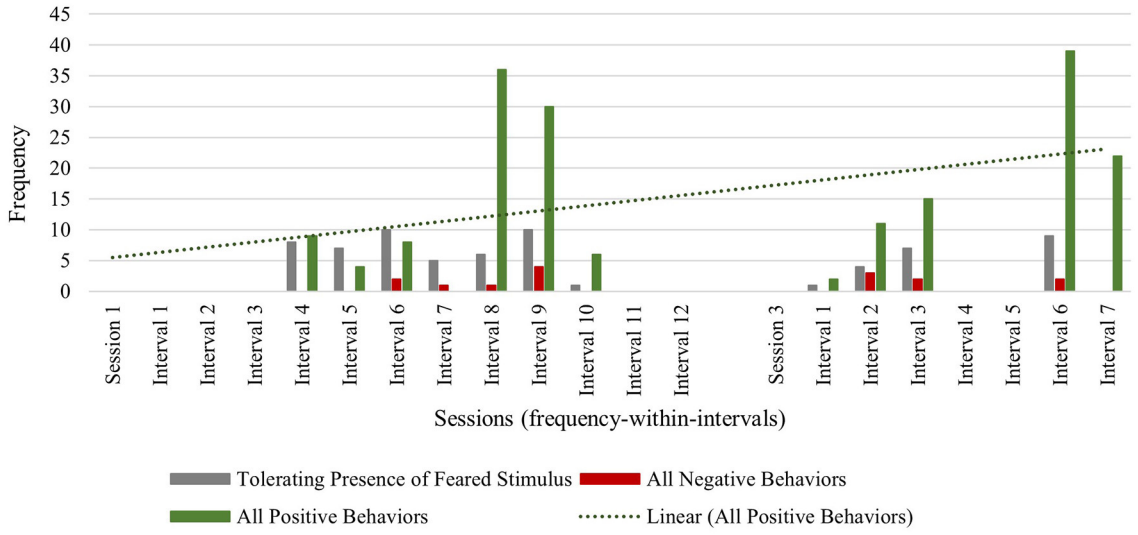


FIGURE 1
Ashton: across session child behavior.

Behavioral coding analyses

The duration of each therapist behavior code was analyzed using duration-within-interval recording. Intervals were also 5-minute each across the duration of each session.

The frequency of each child behavior code was calculated, graphed, and visually inspected using frequency-within-interval recording. Intervals were defined as 5-minute each across the duration of each session. Each child participated in 3–4 therapy sessions. For two participants, one session video was excluded because the intervention focus was on a secondary feared stimuli (i.e., not the primary targeted fear identified). Each session video duration ranged from 30 to 65 min as indicated in Table 1.

Analyses of temporal patterns were conducted using lag sequential analyses in Noldus Observer XT 15.0. Lag sequential data are defined as identifying an event that immediately follows an event (i.e., state lag event of 1). Lag sequential data for therapist behavior codes following child behavior codes and child behavior codes following therapist behaviors were analyzed to identify high frequencies of sequential behavior patterns. First, lag sequential data for therapist behaviors following child behavior codes were analyzed to identify ways in which the therapist responded to the child, within and across sessions. Second, lag sequential data for child behaviors following therapist behaviors were analyzed to identify how each child responded to each therapist technique within the context of exposure therapy.

Each participant's behavioral coding data analyses will be presented individually, using pseudonyms to protect privacy for each child, in a case series design format. Of note, passive exposure was only occasionally observed and is therefore not interpreted here. To illustrate the unique response to the intervention observed for the different participants in this sample, we have individually tailored presentations of the findings to best capture the nature of each participant's response. Common behavioral coding trends across participants will be presented in the discussion.

Ashton

Ashton's primary targeted fear was hand dryers found in public restrooms. Therapy sessions 1 and 3 were coded and analyzed as these sessions focused on this primary feared stimulus. Below child behavior (frequency coding) will be discussed first, followed by therapist behaviors (duration coding) and last, lag sequential findings (state event lag of 1).

Across sessions, Ashton exhibited low frequencies of all negative behaviors (verbalizations and physical avoidance), with a total frequency of 8 in session 1 and a total frequency of 7 in session 3. This highlights the flexible and attuned nature of the intervention, where the therapist works to minimize overly distressing the child during exposures. The observed frequency of tolerating the feared stimulus decreased from a total frequency of 47 in session 1, to a total session frequency of 21 in session 3. However, this decrease in tolerance of the feared stimulus can be explained by the maintained frequency of positive verbalizations (frequency of 34 during both sessions 1 and 3) and even an increase in positive physical behaviors (e.g., approach or direct contact with the feared stimulus) from session 1 (frequency of 33) to session 3 (frequency of 35) (see Figure 1).

Furthermore, Ashton's observed positive behaviors maintained and increased in the face of increasingly challenging stimulus types used by the therapist across sessions. In session 1, the therapist used pure exposure with media stimulus types (i.e., least challenging stimulus type), followed by exposure with play and humor using a toy stimulus type (i.e., a portable hand dryer) (see Figure 2). In the first half of session 3, the use of play and humor was paired with the toy version of the hand dryer along with allowing scarves to be blown by the hand dryer, to engage with the stimulus in a new and unconventional way. In the latter half of session 3, this child engaged in increasingly longer durations of pure exposure with the most challenging stimulus type: an actual hand dryer in a community restroom (see Figure 2). The most frequent child behavior group in session 3 was positive behavior (frequency of 89),

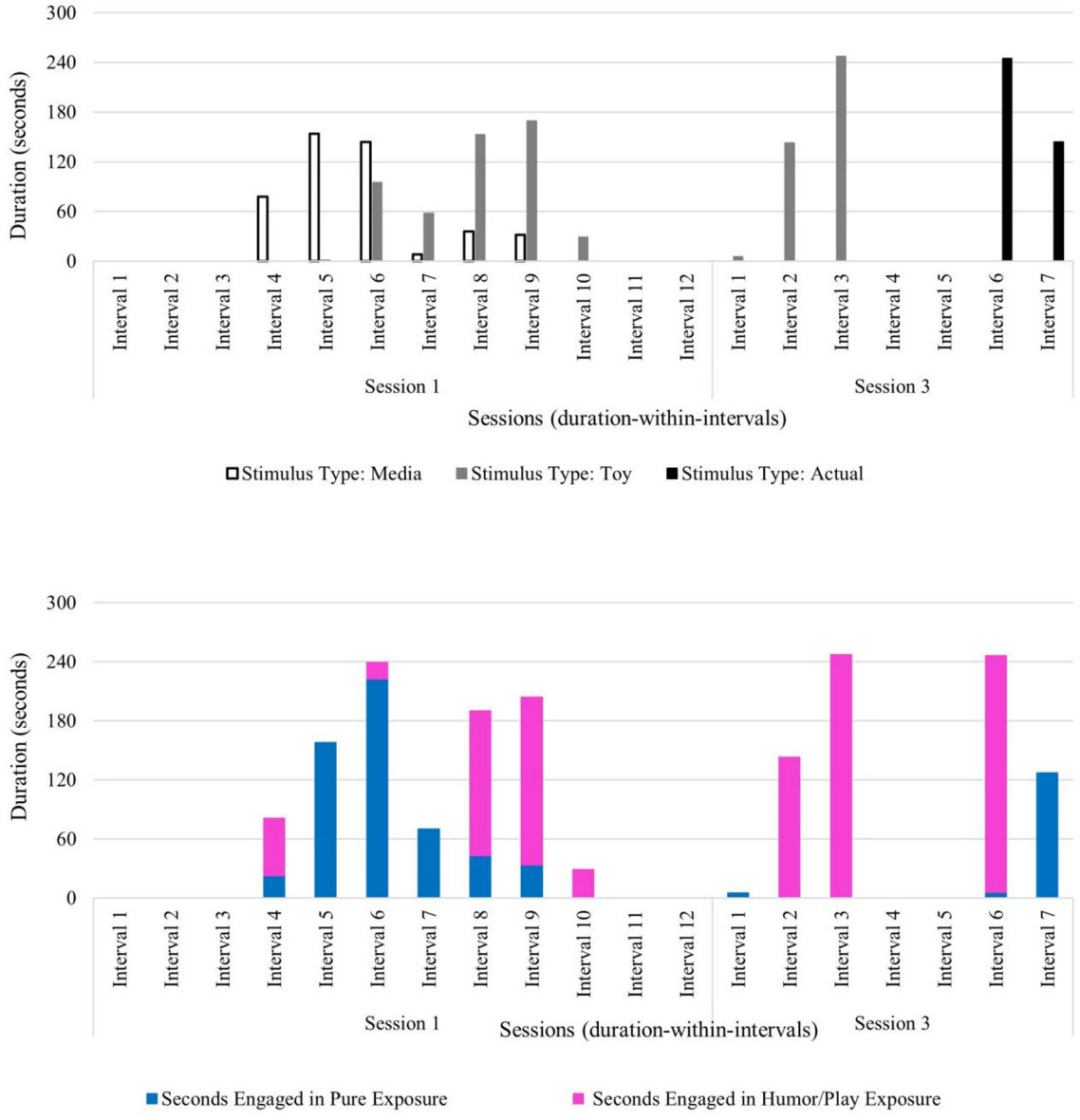


FIGURE 2
Ashton: therapist stimulus type and exposure type across sessions.

relative to tolerant behavior (frequency of 21) and negative behavior (frequency of 7). Across sessions, Ashton's progress was observed through the frequency of positive verbalizations, and an increase in positive physical behaviors (e.g., approach or direct contact with the feared stimulus) in the presence of his feared stimulus. These data reveal that over the course of this brief play- and humor-infused exposure therapy, this child was able to not only tolerate the feared stimulus, but also engage with the previously feared stimulus in positive ways with no observable signs of distress—even when faced with the most challenging level of his fear hierarchy (i.e., using an activated hand dryer in a community restroom) (see Figure 2).

Next, temporal patterns between child and therapist behavior codes were analyzed using lag sequential data analyses. First, temporal patterns between therapist use of invited attention and all child behaviors were analyzed. The results show that therapist use of invited attention most frequently followed the child tolerating the feared stimulus and also followed positive child behaviors and was less common following negative child behaviors. This differential use of invited attention demonstrates how the therapist uniquely tailors the timing of such techniques (e.g., redirection, narration, and exaggerated emotion to facilitate co-regulation). When a child is observed to tolerate a feared stimulus, it can appear quite ambiguous to the therapist. For instance, the child's absence of negative or positive behaviors can suggest to the therapist that a redirection to the exposure activity may be warranted to prevent an occurrence of negative behavior (i.e., proactive strategy) and to continue tolerance of the feared stimulus. The use of invited attention following the child's positive behaviors suggests that the therapist uses techniques to encourage continued positive engagement with the feared stimulus (see Table 7). Importantly,

10.3389/fpsyg.2023.1098449

TABLE 6 Frequency of child behaviors immediately following therapist use of invited attention.

Session	Negative response				Tolerance of the feared stimulus				Positive response			
	Ashton	Beth	Colton	Danny	Ashton	Beth	Colton	Danny	Ashton	Beth	Colton	Danny
Session 1	0	**13**	5	7	0	4	4	7	**10**	**19**	5	**12**
Session 2	N/A	**20**	0	4	N/A	4	4	4	N/A	**12**	9	**16**
Session 3	1	4	3	4	2	2	5	7	**19**	7	**18**	**19**
Session 4			7				6				7	

N/A, session was not coded for this participant.
Lag sequential frequencies of 10 or more are bolded.

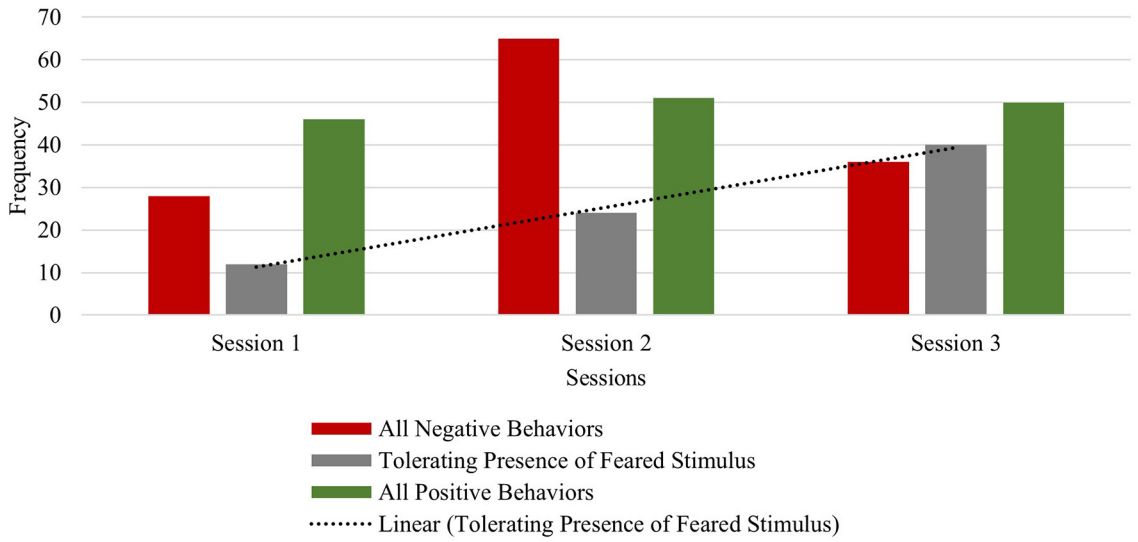


FIGURE 3
Beth: across session child behavior.

the results also show that following the therapist's use of invited attention, the most frequent child behavior code included positive physical behaviors (e.g., approaching or making direct contact with the hand dryer) (see Table 6). This pattern is suggestive that the therapist's technique to gently entice the child to participate in play-based exposures most frequently yields positive child behavioral responses.

Beth

Beth's primary targeted fear was identified as blenders. Therapy sessions 1, 2, and 3 were coded and analyzed as all sessions focused on this primary feared stimulus. Below child behavior (frequency coding) will be discussed first, followed by therapist behaviors (duration coding) and last, lag sequential findings (state event lag of 1).

Beth exhibited lower frequencies of within-session negative behaviors (verbalizations and physical avoidance) relative to within-session positive behaviors in the first session (negative behavior frequency of 28 and positive behavior frequency of 46) and in the final session (negative behavior frequency of 36 and positive behavior frequency of 50). Across session data show increasing frequencies of both negative and positive behaviors. It was observed that negative physical behaviors (e.g., covering ears) often co-occurred with positive physical behaviors (e.g., approaching the feared stimulus) within each of the three sessions. This behavior pattern has been highlighted in the literature as unique among children with Williams syndrome who experience fears and phobias (Gallo et al., 2008). Her verbalizations (both negative and positive) increased from session 1 (negative verbalizations: frequency of 4; positive verbalizations: frequency of 21) to session 3 (negative protest: frequency of 17; positive verbalizations: frequency of 34), while her physical behaviors decreased from session 1 (negative: frequency of 24; positive: frequency of 25) to session 3 (negative: frequency of 19; positive: frequency of 16). This increase in verbal protesting may be suggestive of attempted avoidance (similar to an extinction burst) of the feared stimulus as increasingly challenging levels of the fear hierarchy are presented over the course of this brief intervention (i.e., media to toy to actual blender) (see Figures 3, 4). Beth did exhibit some negative verbalizations while she simultaneously engaged in positive physical behaviors (e.g., approaching the blender). Notably, the observed frequency of Beth tolerating the feared stimulus showed a consistent increase across all sessions (frequency of 12 in session 1; frequency of 24 in session 2; frequency of 40 in session 3), with Beth making direct contact and operating an activated blender by session 3 (see Figure 3).

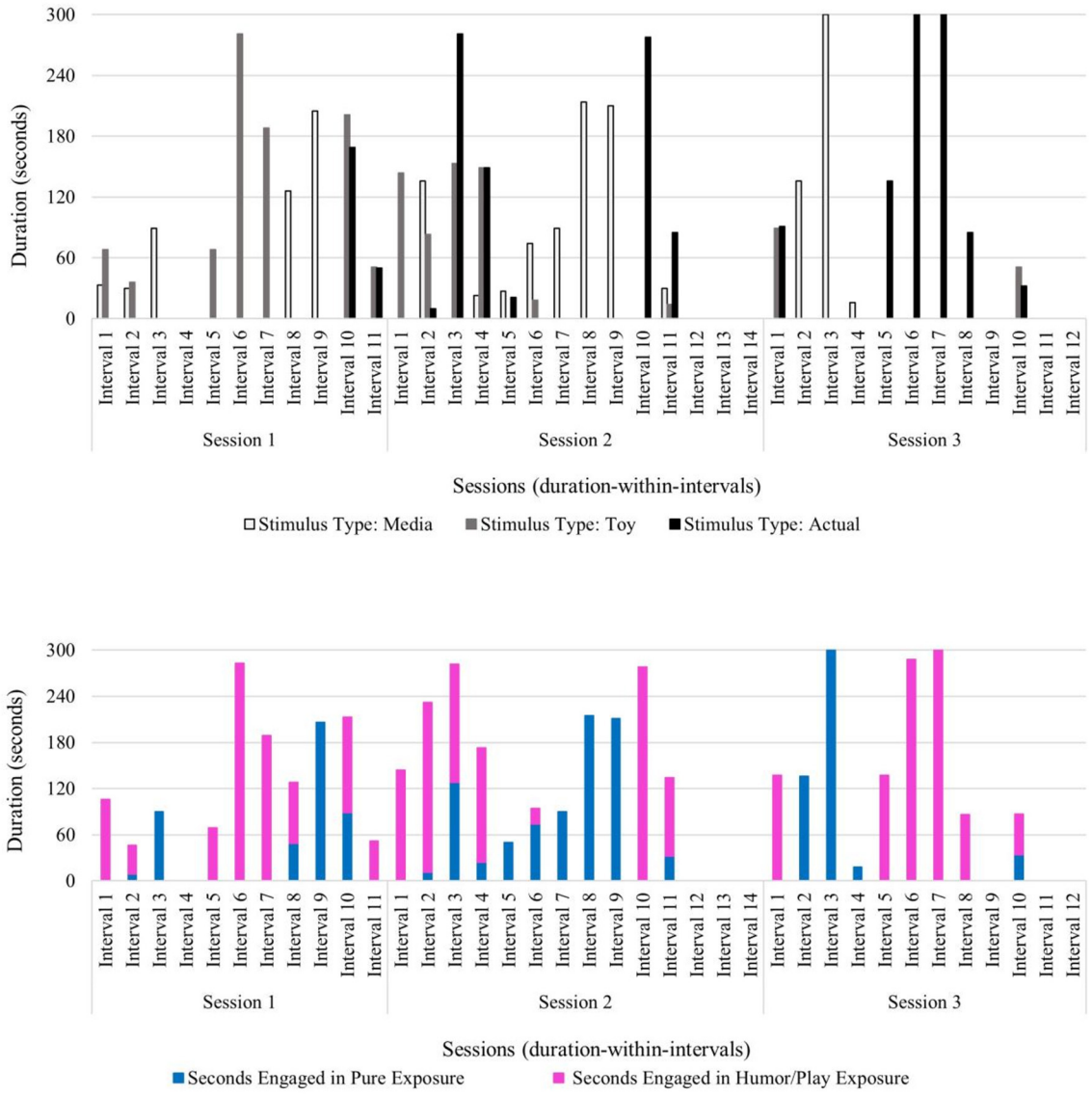


FIGURE 4
Beth: therapist stimulus type and exposure type across sessions.

Beth's pattern of co-occurring negative and positive behaviors along with a steady increase in tolerating the feared stimulus likely resulted from the therapist's deliberate use of specific exposure type(s) paired with specific stimulus type(s) across sessions. The therapist used play and humor to co-regulate, to help Beth sustain emotional regulation, and to prevent anxiety from becoming overwhelming. In session 1, the therapist infused play and humor into exposures with all stimulus types (i.e., media, toy, and actual blender) and implemented pure exposure with media examples of blenders and brief durations of the actual stimulus. The actual blender was introduced near the end of session 1 and unplugged and turned off to begin exposure to the sight of the object before working up the fear hierarchy to auditory exposure of the blender turned on later in the course of treatment (see Figure 4).

Next, temporal patterns between child and therapist behavior codes were analyzed using lag sequential data analyses. First, temporal patterns between therapist use of invited attention and all child behaviors were analyzed. Findings reveal that during sessions 1 and 2, when the actual stimulus was presented to Beth and elicited some avoidance (i.e., negative behaviors) as well as interest in the blender (i.e., positive behaviors), therapist invited attention often followed all child behaviors (negative, tolerant, and positive) (see Table 7). Differential use of invited attention was often tailored to the type of child behavior exhibited. Narration, exaggerated emotion using toys with play and humor, and redirection prompts were often used in response to Beth's simultaneous negative (e.g., verbal protest) and positive behavior (e.g., direct contact with the blender) to re-engage or maintain participation in the exposures. Additionally,

TABLE 7 Frequency of therapist invited attention immediately following child behaviors.

Session	Negative behavior				Tolerance of the feared stimulus				Positive behavior			
	Ashton	Beth	Colton	Danny	Ashton	Beth	Colton	Danny	Ashton	Beth	Colton	Danny
Session 1	0	**14**	2	9	**12**	7	6	**10**	**11**	**13**	7	6
Session 2	N/A	**27**	0	8	N/A	5	5	6	N/A	**13**	9	**12**
Session 3	1	9	4	6	6	5	8	7	**16**	4	**14**	**15**
Session 4	-	-	8		-	-	5		-	-	8	

N/A, session was not coded for this participant.
Lag sequential frequencies of 10 or more are bolded.

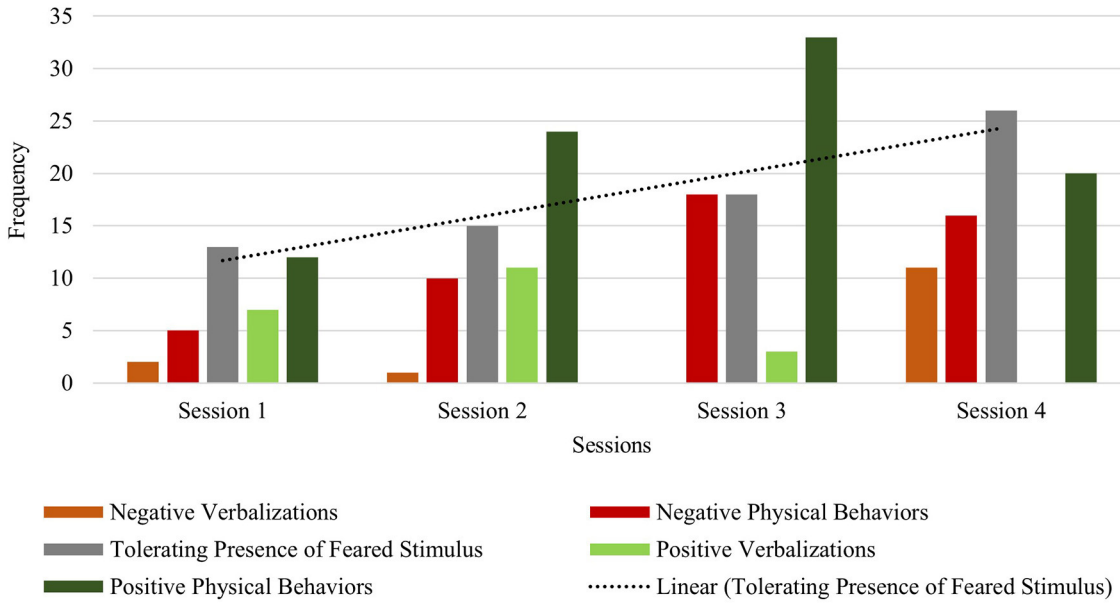


FIGURE 5
Colton: across session child behavior.

invited attention was also used as a priming technique to cue the child that an exposure activity was beginning to minimize upset or startle response with the transition. From sessions 1 and 2 to session 3, fewer negative child behaviors followed therapist invited attention. Rather, positive behaviors more often followed invited attention in session 3, suggesting effectiveness of the co-regulation efforts and encouraging the child to continue engagement in the therapy session—even as the stimulus type became increasingly challenging over the course of intervention sessions (see Table 6).

Colton

Colton's primary targeted fear was identified as blood pressure cuffs. Therapy sessions 1, 2, 3, and 4 were coded and analyzed as all sessions focused on this primary feared stimulus. Below child behavior (frequency coding) will be discussed first, followed by therapist behaviors (duration coding) and last, lag sequential findings (state event lag of 1).

Colton exhibited lower frequencies of within-session negative behaviors (verbalizations and physical avoidance) relative to within-session positive behaviors in sessions 1 and 2. In session 3, frequencies of negative behaviors were equal to his tolerating behaviors (frequency: 18), with twice the frequencies of positive behaviors (frequency: 36) in response to his feared stimulus. In session 4, although negative behaviors increased, Colton's observed tolerating of the feared stimulus increased as well (negative behaviors: frequency of 27; tolerating behavior: frequency of 26) (see Figure 5). Colton exhibited a relatively lower frequency of within-session negative verbalizations compared to positive verbalizations in sessions 1–3. In session 4, however, the frequency of his within-session positive verbalizations was relatively lower than his within-session negative verbalizations. Notably, Colton demonstrated a steady across-session increase of tolerating his feared stimulus (session one frequency: 13; session two frequency: 15; session three frequency: 18; session four frequency: 26). Furthermore, positive physical behaviors increased across sessions from the first (frequency of 12) to the final session (frequency of 20), indicating more frequent approach and direct contact with blood pressure cuffs, which most often included the actual stimulus type in session 4. Taken together, Colton's improved tolerance and positive physical behaviors are suggestive of improved regulation skills in response to his previously feared stimulus over the course of this brief therapy approach (Figure 5).

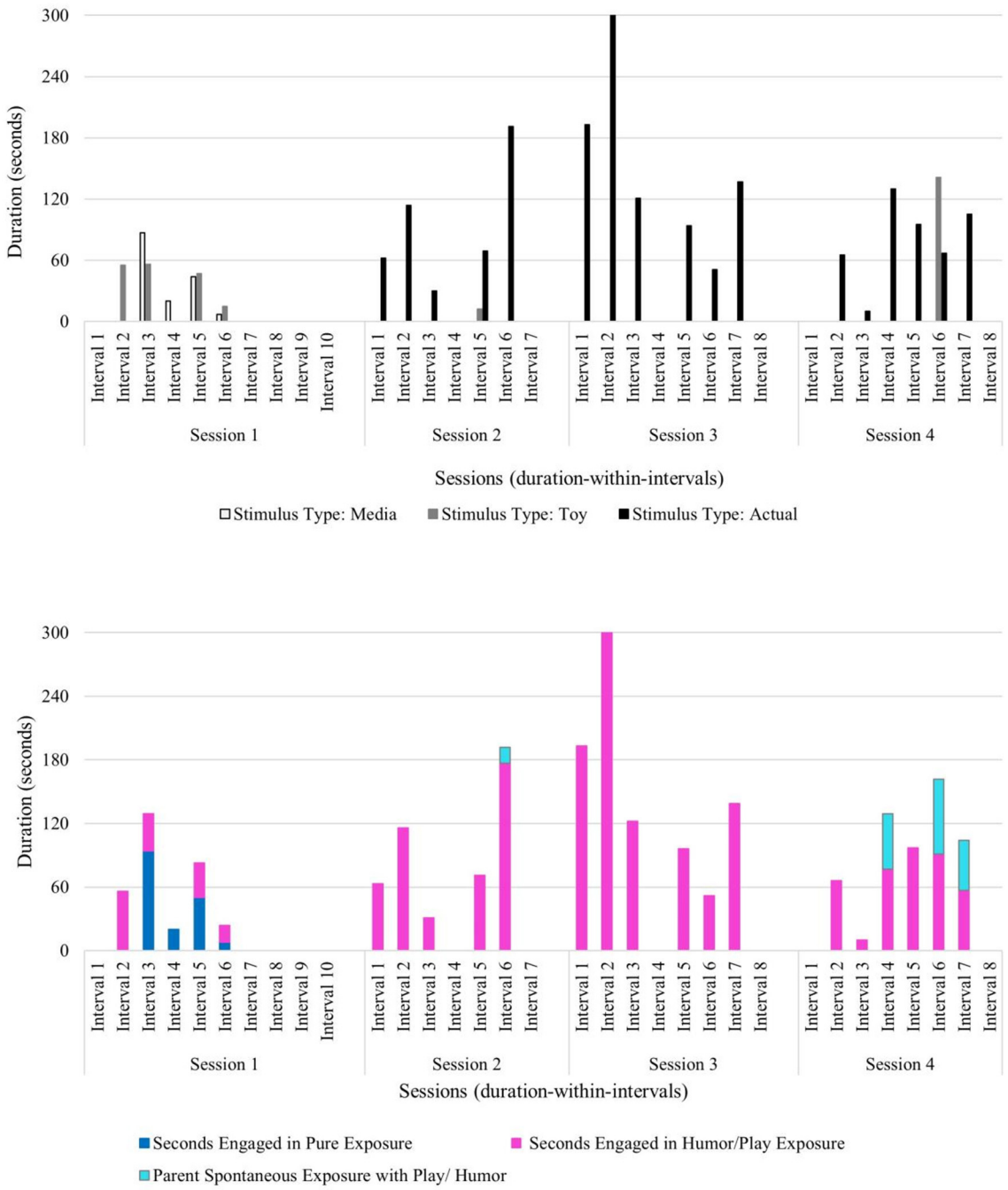


FIGURE 6
Colton: therapist stimulus type and exposure type across sessions.

The therapist attunement to Colton throughout treatment fostered the observed increase in his ability to tolerate his feared stimulus. Therapist use of pure exposure was brief and only used in session 1 with media examples of the feared stimulus. However, exposure with play and humor was used across all sessions with a toy version and the actual stimulus type (see Figure 6). Parent use of spontaneous play- and humor-infused exposure was also observed in session 2 (duration: 15 s) and session 4 (duration: 2 min, 50 s), suggesting that Colton's parent began acquiring and applying play- and humor-related skills with increasingly longer durations as therapy progressed (see Figure 6). The observed behavior pattern in Colton (who has relatively less developed verbal abilities compared to other children in this sample) highlights the benefit of this developmentally attuned intervention which incorporates adaptations to meet and respond to the individual child's needs and abilities during exposures. Colton's verbal score on the DAS-II was in the lower range (SS = 59), which speaks to the need for the therapist to gauge his behavior and non-verbal cues

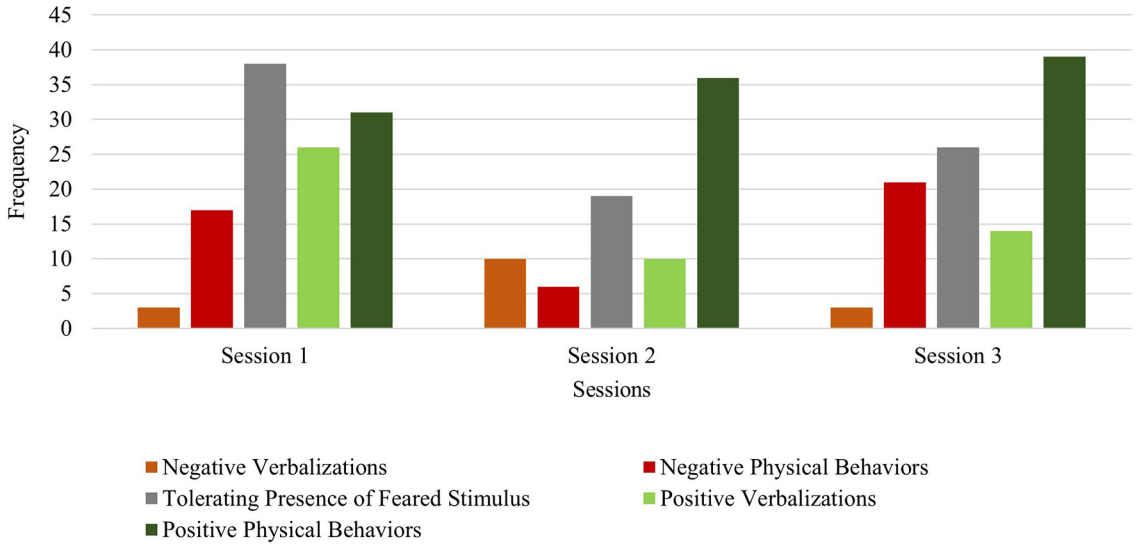


FIGURE 7
Danny: across session child behavior.

during exposures to sustain emotional regulation, avoid distress and upset, and minimize anxiety as they work up his fear hierarchy to the most challenging stimulus type: an actual blood pressure cuff. Taken together, the primary use of play and humor by the therapist and parent, along with shorter session durations, reflects the attunement to this child's individual needs.

Next, temporal patterns between child and therapist behavior codes were analyzed using lag sequential data analyses. First, temporal patterns between therapist use of invited attention and all child behaviors were analyzed. The results show that in response to varied child behavior (negative, tolerant and positive), the therapist frequently used invited attention techniques to either redirect the child to the activity (if negative child behavior was observed), use gesture or verbal prompts to gauge child's anxiety level (if child tolerating was observed), or narration in a playful and humorous way to continue child engagement in exposures (if positive child behavior was observed) (see Table 7). Following therapist invited attention, the most frequent observed child responses across sessions were positive physical behaviors (e.g., approaching the feared stimulus) and tolerance of the feared stimulus (i.e., blood pressure cuff) (see Table 6). These data provide some support for the effectiveness of the therapist's varied and tailored use of invited attention techniques with Colton considering he had limited verbal skills and was younger than the other participants.

Danny

Danny's primary targeted fear was identified as vacuum cleaners. Therapy sessions 1, 2, and 3 were coded and analyzed, but session 4 was not coded because the primary fear was not targeted during this session. Below, child behavior (frequency coding) will be discussed first, followed by therapist behaviors (duration coding) and last, lag sequential findings (state event lag of 1).

Danny exhibited lower frequencies of within-session negative behaviors (verbalizations and physical avoidance) relative to within-session tolerating and positive behaviors in session 1 (negative behavior frequency of 20; tolerating behavior frequency of 38; positive behavior frequency of 57), session 2 (negative behavior frequency of 16; tolerating behavior frequency of 19; positive behavior frequency of 46), and session 3 (negative behavior frequency of 24; tolerating behavior frequency of 26; positive behavior frequency of 53). Across session data show a slight increase in positive physical behaviors (i.e., approaching or making direct physical contact with the feared stimulus) from session 1 (frequency: 31) to session 3 (frequency: 39). Danny's profile of behavioral data trends shows his ability to tolerate and positively engage with vacuum cleaners within each session, even as the stimulus type became increasingly challenging (i.e., media to toy version to actual feared stimulus). It is notable that the child spent several minutes independently vacuuming the carpet in session 3 (see Figure 7).

Therapist use of pure exposure was primarily used with media versions of vacuum cleaners during sessions 1 and 2 and then used with a toy version and with an actual vacuum cleaner during session 3. Exposure with play and humor was used across all sessions primarily with a toy version and the actual stimulus type, and in shorter durations with media examples in session 1 (see Figure 8). Notably, Danny progressed through his fear hierarchy well, and by session 3, he was able to engage in independent vacuuming (i.e., pure exposure with the actual stimulus) (see Figure 8).

Next, temporal patterns between child and therapist behavior codes were analyzed using lag sequential data analyses. First, temporal patterns between therapist use of invited attention and all child behaviors were analyzed. The results show that the most frequently used therapist technique following child behaviors was invited attention (see Table 7). Specifically, invited attention in

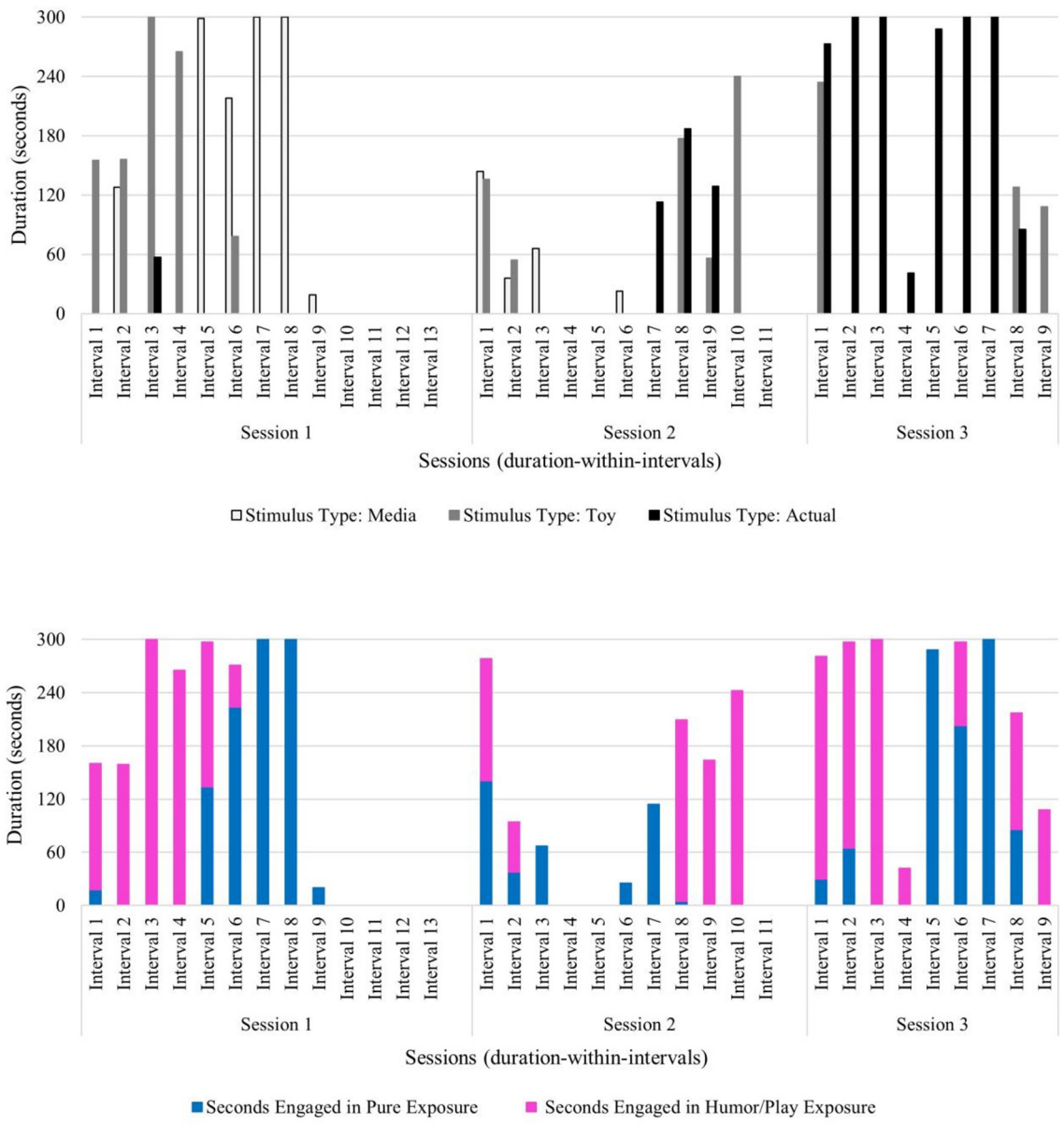


FIGURE 8
Danny: therapist stimulus type and exposure type across sessions.

session 1 most often followed child tolerating the feared stimulus and child negative behaviors. This demonstrates the therapist working to build and maintain rapport and encourage engagement early in therapy (e.g., redirection and narration) to prepare the child for increasingly challenging levels of the fear hierarchy. In sessions 2 and 3, the therapist most often used invited attention techniques following child tolerant behavior (session 2: 6; session 3: 7) and positive physical child behavior (session 2: 10; session 3: 13) (see Table 7). The pattern of invited attention used in sessions 2 and 3 suggests that as the child progressed through the fear hierarchy and was able to make direct physical contact with the feared stimulus more frequently, the therapist facilitated continued contact and positive engagement through narration, modeling of exaggerated emotion (e.g., "That vacuum is so loud! I'm going to cover my ears!"), and redirection prompts to the exposure. Lag sequential data of child behaviors following invited attention show that this approach yielded some success with Danny. Across sessions, Danny responded to the therapist's use of invited attention with increased frequency of positive physical behaviors (session 1: 5; session 2: 16; session 3: 18) (see Table 6).

Discussion

In our prior work demonstrating preliminary acceptability and utility of play- and humor-infused exposure therapy for children with Williams syndrome, parent and clinician ratings indicated improvements in fear for some children with Williams

syndrome, even after a very brief course of treatment. The current study validated these clinician and parent ratings by demonstrating that this observed improvement was evident based on behavioral coding. Increased tolerance of the feared stimulus across sessions and/or movement from engagement with videos or toys to engagement with real feared objects was observed over the course of this brief intervention for all of the participants. These findings add to our preliminary evidence of the promise of this play- and humor-infused exposure therapy for children with Williams syndrome to address intense emotional dysregulation in response to specific stimuli (i.e., fears and phobias). Additionally, the combination of therapist techniques, including use of invited attention, social play and humor, and relations of these techniques to child behavior, suggests that the therapist's attunement to the child's distress level encourages continued contact with the feared stimulus and results in adjustment of the intensity of the exposure through use of videos, toys, and real-world objects. Findings of the therapist techniques for co-regulation used within this play- and humor-infused approach show that attunement to the child, exaggerated emotion to join the child in their fear, and use of play and humor yield child behaviors suggestive of improved coping through co-regulation, ability to maintain contact with the feared stimulus without becoming overly distressed, ultimately leading to increase in the child's capacity to sustain emotional regulation, improved tolerance, and sometimes positive engagement with their feared stimulus.

The use of humor-infused therapy for children has been suggested to potentially be useful for treatment of anxiety (Consoli et al., 2018) in sustaining emotional regulation in the face of a moderate level of discomfort without overly distressing the child, thus minimizing disengagement from therapy. However, systematic examination of humor-infused therapy is lacking. One critique of this approach is that core to exposure is a need to avoid distractions to facilitate activation and subsequent extinction of the phobic association between the conditioned stimulus and the fear response (Rescorla, 1972). However, we propose that the mechanism of fear reduction here is the establishment and reinforcement of a *new* association of fear inhibition that goes against the typical activation of the amygdala during fear conditioning, rather than simply extinguishing the previously established fear association (Slifer et al., 2002; Craske et al., 2014). Furthermore, behavior therapy has recently been the subject of some critique based on clients' lived experiences due to its perceived aversiveness (Anderson, 2023). Importantly, while the intervention employed in this study is rooted in evidence-based, gold-standard therapies for fears and phobias (APA Division 12 Society of Clinical Psychology, 2022), the therapeutic approach of the current study focused on gradual incorporation of the feared stimulus into play, maintaining the comfort of the child in the context of the exposure to minimize its aversiveness. Essentially, we posit that this intervention fosters formation of an association between the feared stimulus and socially engaging play and humor, which runs counter to the fear response. This model fits well with what is known of the atypical amygdala-prefrontal connectivity thought to be responsible for more pronounced fear associations and subsequent responses (Meyer-Lindenberg et al., 2005).

This play- and humor-infused exposure intervention explicitly uses a therapist–child interactive play-based approach to deliver the therapy to children. The study therapist demonstrated some common patterns related to stimulus type (media, toy, or actual stimulus) and type of exposure (pure exposure or exposure with play and humor). Media examples of the feared stimulus were most often used within pure exposures (i.e., no play or humor). Toy versions of the feared stimulus and the (actual) feared stimulus were typically presented and incorporated within play- and humor-based exposures. This highlights the developmentally attuned nature of the therapeutic approach. By infusing more play and humor into the exposures as the child progresses up the fear hierarchy, the therapist is able to co-regulate with the child and thus the child is able to sustain emotional regulation, minimize distress and anxiety while simultaneously allowing for continued engagement and increasingly more direct contact with the feared stimulus. The feared stimulus is gradually incorporated into the sessions. As designed, this intervention is largely therapist-led, in the presence of the parent. Nevertheless, during this intervention, we observed some parent involvement in the exposures for one participant. It is likely that parental involvement would indeed facilitate generalization of the skills to the home environment. Therefore, future work developing this intervention approach may benefit from more explicit guidance of parents as play- and humor-infused exposure facilitators.

The combination of both behavioral (exposure, systematic desensitization, and counterconditioning) and play-based (age-appropriate social adult–child play development) approaches may be especially well-suited for children with Williams syndrome. The gregarious nature of children with Williams syndrome may mask the underlying emotion dysregulation stemming from anxiety. The focus of this intervention for children with Williams syndrome was to improve emotion regulation relating to specific phobias by building coping skills through social play-based approaches (Farrell et al., 2018) which leverages the social nature and motivation of children with Williams syndrome.

Patterns related to the use of invited attention were also observed. In the context of this study, invited attention was coded when one of the following techniques was observed: priming before an exposure began, narration during or immediately after an exposure, or gesture or verbal prompts during an exposure to direct or redirect the child's attention. Across participants in this sample, the therapist invited attention to the feared stimulus both before and after particular child behaviors were observed. Specifically, the therapist often primed the child before beginning an exposure. The use of narration and exaggerated emotion modeling (e.g., personifying a toy to cry when in the presence of the feared stimulus) was used with all children in this study to encourage continued engagement and promote co-regulation by joining the child in their fear. This approach leverages the empathic and social nature of children with Williams syndrome, by allowing them to experience discomfort and co-regulate with the therapist in a safe and supportive setting. By joining each child in their fear during exposures, the therapist helped them work through these more difficult

and uncomfortable emotions (e.g., fear) while gently encouraging their progress through increasingly challenging levels of the fear hierarchy. In some instances, over the course of the intervention, a child even comforted the personified toy (e.g., "it's okay, it's not that loud"). Additionally, when a child's behavior was not clearly positive or negative (i.e., tolerant), the therapist frequently used redirection to prevent escalated anxiety and to promote continued engagement in the exposure activity. The therapist-initiated techniques were often tailored to the individual needs of each child to facilitate progress through the fear hierarchies and ultimately reduce distress in the presence of the phobic stimulus.

Although children with Williams syndrome tend to have auditory-based phobias, the underlying etiology of these sound sensitivities is still undefined. Noise sensitivities may be associated with biological or cognitive behavioral mechanisms. Hyperacusis (i.e., hearing disorder of sensitivity) and phonophobia (i.e., anxiety disorder relating to fear of specific sounds) are two concerns that are commonly expressed by people with Williams syndrome (Silva et al., 2021). Rates as high as 95% (Klein et al., 1990; Nigam and Samuel, 1994) have been reported for people with Williams syndrome and co-occurring hyperacusis, with even more severe symptoms among children (Gothelf et al., 2006). The vulnerability to hyperacusis may be due to the genetic underpinnings of Williams syndrome. Two genes that are deleted on chromosome 7q11.23, one responsible for elastin (ELN) and the other (LIMK-1) dedicated to encoding a serine/threonine kinase, may provide a biologically based explanation for hyperacusis. Deficiencies in elastin may stiffen the stapedius tendon, which is responsible for sound regulation, and may therefore contribute to hyperacusis (Levitin et al., 2005; Prasad et al., 2019). Additionally, the LIMK-1 gene is responsible for the regulation of outer hair cell movement, and dysfunction in this gene has been suggested to increase sound amplification and subsequent startle response to auditory stimuli (Stanyon and Bernard, 1999; Meng et al., 2002; Matsumoto et al., 2011; Tyler et al., 2014). Further research is needed to provide clearer characterization of sound processing abilities and dysfunction among people with Williams syndrome, with particular attention toward the presentation of simultaneous aversion and keen interest in certain sounds (Krilčić and Petranović, 2017). A case study examined the effectiveness of a modified CBT intervention on anxiety and avoidance related to noise sensitivity in an adolescent with autism spectrum disorder and co-occurring intellectual disability (Fodstad et al., 2021). Treatment progress in this study was defined as improved tolerance to auditory input and reduced problem behaviors, with predefined coping skills available for participant use during exposure to such noise. Our current research study aligns with this procedure by allowing participants to engage in simultaneous negative and positive physical behaviors in the presence of the auditory-based feared stimulus (e.g., Beth covering her ears while also approaching a blender turned on), to ultimately increase emotional regulation in the presence of the feared stimulus in naturalistic settings. It is notable that evidence-based treatments for misophonia are largely exposure-based (Bernstein et al., 2013; McGuire et al., 2015; Reid et al., 2016). It may be that developmentally, when the children are young, they may be vulnerable to actual intense discomfort in response to some sounds. Physiologically, this may improve as they physically develop, yet the initial frightening experiences may evolve into an emotionally based phobia, with or without continued sensory sensitivity. Taking sound sensitivities into consideration, it is important to note that the brief play-and humor-infused exposure intervention used in this study demonstrated effectiveness for all four children, regardless of the etiology of such noise sensitivities.

Limitations and future directions

Limitations include a very small sample size ($n = 4$) and an A-B single-subject research design, which limit the generalizability of findings. This sample is made up of four participants who were White, hence lacked racial diversity. Recruitment strategies to diversify samples in future studies are needed. Furthermore, there were limitations to the behavior coding scheme. Specifically, the behavior coding scheme focused on coding the child's primary fear only (identified during the initial functional assessment interview) and did not capture the intensity of the stimulus sound or the intensity of the child's verbal responses (e.g., volume of protest). Therapist behavior patterns of exposure using sequenced feared stimuli across sessions that included secondary feared stimuli (identified during the functional assessment interview) were not coded (e.g., exposure with the use of a thunder video as a secondary feared stimulus, followed by exposure with the primary feared stimulus of a hand dryer). Furthermore, the behavior coding scheme did not explicitly capture child-initiated coping, which was on occasion observed. For example, Ashton was exhibiting increasing fearful behaviors while the hand dryer was turned on, and he then requested the use of a thunder tube to hold and shift attention toward; after holding and listening to the thunder tube while still in the presence of the hand dryer, his observable fear response reduced. Finally, we acknowledge that behavior coding introduces the possibility of rater bias (i.e., differences between rater identities and participant identities), which we aimed to minimize through independent behavior coding by respective coders.

Further examination of the effectiveness of this approach is needed using a more controlled, multiple baseline research design. Notably, the main aim of this initial round of this research was to gather video illustrations of this play- and humor-infused exposure therapy approach to disseminate to community clinicians; none of the participants completed what would be considered a full round of the intervention. Future research would benefit from collection of more systematic baseline and follow-up data as well as inclusion of community clinicians who can implement the appropriate length of intervention based on the specific needs of the child without the time constraints inherent in the design of the current study. Future studies may also explore associations between the auditory intensity of feared stimuli and the intensity of the child response, or physiological response to the feared stimuli, to better characterize the etiology of sound sensitivities among

children with Williams syndrome and provide recommendations on intervention modifications based on co-occurring conditions (e.g., hyperacusis, misophonia, phonophonia, and ASD) and degree of general cognitive and language difficulties. Additionally, the role of parent psychoeducation within this play- and humor-infused exposure therapy may be investigated in future research. Specifically, examination of the effects of parent psychoeducation about mechanisms for fear and anxiety and parental instruction in the use of play- and humor-infused approaches on the sustained impact of the intervention within the naturalistic environment is warranted.

Conclusion

This study adds to the sparse research on the utility and effectiveness of interventions for children with Williams syndrome and co-occurring fears and phobias. Specifically, the findings of the current study provide support for increased emotion regulation in the form of improved tolerance of the feared stimulus and/or the ability to progress through the fear hierarchy (i.e., media version to toy version to real-world stimulus) following a brief social play- and humor-infused exposure therapy approach with four children with Williams syndrome and co-occurring fears and phobias based on observational behavior coding. For two of the four children, improvements were evident in a move beyond tolerating the feared stimulus toward increased positive behaviors with the feared stimulus across sessions. All four children in this sample progressed through their fear hierarchy and were able to tolerate or even positively engage with the real-world previously feared stimulus by the final therapy session. This line of research provides evidentiary support for developmentally attuned approaches to exposure-based interventions for children with Williams syndrome, adding to the paucity of treatment development literature addressing anxiety and phobias in children with rare neurogenic conditions.

Data availability statement

The raw data supporting the conclusions of this article will be made available by the authors, without undue reservation.

Ethics statement

The studies involving human participants were reviewed and approved by University of Wisconsin-Milwaukee Institutional Review Board. Written informed consent to participate in this study was provided by the participants' legal guardian/next of kin. Written informed consent was obtained from the individual(s), and minor(s)' legal guardian/next of kin, for the publication of any potentially identifiable images or data included in this article.

Author contributions

BY contributed to the development of the coding scheme, served as the primary coder, analysis and interpretation of the data, drafting of the manuscript, and revising the manuscript based on co-author feedback. EM contributed to the refinement of the coding scheme, served as the secondary coder, contributed to data interpretation, and provided conceptual revision feedback on the manuscript. KL originated the humor- and play-infused exposure therapy approach, contributed to the development of the intervention manual, served as the study therapist, and provided conceptual feedback on the manuscript. BK-T collaborated with KL to operationalize the intervention approach into a treatment manual, recruited and enrolled participants, coordinated intervention appointments, assisted with intervention appointments, and supervised the development and implementation of the coding scheme, data interpretation, manuscript-writing, and including providing manuscript revisions. All authors contributed to the article and approved the submitted version.

Funding

This study was funded by Williams Syndrome Association (WSA #0110).

Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

Publisher's note

All claims expressed in this article are solely those of the authors and do not necessarily represent those of their affiliated organizations, or those of the publisher, the editors and the reviewers. Any product that may be evaluated in this article, or claim that may be made by its manufacturer, is not guaranteed or endorsed by the publisher.

References

Anderson, L. K. (2023). Autistic experiences of applied behavior analysis. *Autism*. 27, 737–50. doi: 10.1177/13623613221118216

APA Division 12 Society of Clinical Psychology (2022). *Exposure Therapies for Specific Phobias. Society of Clinical Psychology*. Available online at: https://div12.org/treatment/exposure-therapies-for-specific-phobias/

Bernstein, R. E., Angell, K. L., and Dehle, C. M. A. brief course of cognitive behavioural therapy for the treatment of misophonia: a case example. *Cogn. Behav. Therapist*. (2013) 6. doi: 10.1017/S1754470X13000172

Chorney, J. M., McMurtry, C. M., Chambers, C. T., and Bakeman, R. (2015). Developing and modifying behavioral coding schemes in pediatric

psychology: a practical guide. *J. Ped. Psychol.* 40, 154–64. doi: 10.1093/jpepsy/jsu099

Collins, I. I. R. T., Kaplan, P., Somes, G. W., and Rome, J. J. (2010). Long-term outcomes of patients with cardiovascular abnormalities and Williams syndrome. *Am J Cardiol*. 105, 874–8. doi: 10.1016/j.amjcard.2009.10.069

Conelea, C. A., and Klein-Tasman, B. P. (2013). Habit reversal therapy for body-focused repetitive behaviors in Williams syndrome: a case study. *J Develop Phys Disabil*. 25, 597–611. doi: 10.1007/s10882-013-9335-3

Consoli, A. J., Blears, K., Bunge, E. L., Mandil, J., Sharma, H., Whaling, K. M., et al. (2018). Integrating culture, pedagogy, and humor in CBT with anxious and depressed youth. *Pract Innovat*. 3, 138. doi: 10.1037/pri0000069

Costello, E. J., Copeland, W., and Angold, A. (2011). Trends in psychopathology across the adolescent years: what changes when children become adolescents, and when adolescents become adults? J. *Child Psychol. Psychiatry* 52, 1015–25. doi: 10.1111/j.1469-7610.2011.02446.x

Cowdrey, F. A., and Walz, L. (2015). Exposure therapy for fear of spiders in an adult with learning disabilities: a case report. *Br. J. Learn. Disabil*. 43, 75–82. doi: 10.1111/bld.12106

Craske, M. G., Treanor, M., Conway, C. C., Zbozinek, T., and Vervliet, B. (2014). Maximizing exposure therapy: an inhibitory learning approach. *Behav. Res. Therapy* 58, 10–23. doi: 10.1016/j.brat.2014.04.006

Dagnan, D., Jackson, I., and Eastlake, L. A. (2018). Systematic review of cognitive behavioural therapy for anxiety in adults with intellectual disabilities. *J. Intell. Disabil. Res.* 62, 974–91. doi: 10.1111/jir.12548

Davis, I. I. I. T. E., Kurtz, P. F., Gardner, A. W., and Carman, N. B. (2007). Cognitive-behavioral treatment for specific phobias with a child demonstrating severe problem behavior and developmental delays. *Res. Develop. Disabil*. 28, 546–58. doi: 10.1016/j.ridd.2006.07.003

Davis, I. I. I. T. E., Ollendick, T. H., and Öst, L. G. (2009). Intensive treatment of specific phobias in children and adolescents. *Cogn. Behav. Pract*. 16, 294–303. doi: 10.1016/j.cbpra.2008.12.008

Dovgan, K., Clay, C. J., and Tate, S. A. (2020). Dog phobia intervention: a case study in improvement of physiological and behavioral symptoms in a child with intellectual disability. *Develop. Neurorehabil*. 23, 121–32. doi: 10.1080/17518423.2019.1683909

Duncan, B. L., and Miller, S. D. (2000). *The Heroic Client: Doing Client-Directed, Outcome-Informed Therapy*. San Francisco, CA: Jossey-Bass.

Einfeld, S., Tonge, B., Turner, G., Parmenter, T., and Smith, A. (1999). Longitudinal course of behavioural and emotional problems of young persons with Prader-Willi, Fragile X, Williams and Down syndromes. *J. Intellect. Develop. Disabil*. 24, 349–54. doi: 10.1080/13668259900034111

Einfeld, S. L., Tonge, B. J., and Florio, T. (1997). Behavioral and emotional disturbance in individuals with Williams syndrome. *Am. J. Mental Retard*. 102, 45–53. doi: 10.1352/0895-8017(1997)102<0045:BAEDII>2.0.CO;2

Einfeld, S. L., Tonge, B. J., and Rees, V. W. (2001). Longitudinal course of behavioral and emotional problems in Williams syndrome. *Am. J. Mental Retard*. 106, 73–81. doi: 10.1352/0895-8017(2001)106<0073:LCOBAE>2.0.CO;2

Elliott (2007). *Differential Ability Scales: Second edition*. San Antonio, TX: Pearson. doi: 10.1037/t15074-000

Essau, C., and Longhi, E. C. B. T. "Intervention for anxiety in children and adolescents with Williams syndrome," *inThe Wiley-Blackwell Handbook of the Treatment of Childhood Anxiety*. (2013) (pp. 559-574). Wiley-Blackwell. doi: 10.1002/9781118315088.ch24

Farrell, L. J., Kershaw, H., and Ollendick, T. (2018). Play-modified one-session treatment for young children with a specific phobia of dogs: a multiple baseline case series. *Child Psych. Human Develop*. 49, 317–29. doi: 10.1007/s10578-017-0752-x

Fodstad, J. C., Kerswill, S. A., Kirsch, A. C., Lagges, A., and Schmidt, J. (2021). Assessment and Treatment of noise hypersensitivity in a teenager with autism spectrum disorder: a case study. *J. Autism Develop. Disord*. 51, 1811–22. doi: 10.1007/s10803-020-04650-w

Gallo, F. J., Klein-Tasman, B. P., Gaffrey, M. S., and Curran, P. (2008). Expecting the worst: observations of reactivity to sound in young children with Williams syndrome. *Res. Develop. Disabil*. 29, 567–81. doi: 10.1016/j.ridd.2007.09.003

Gothelf, D., Farber, N., Raveh, E., Apter, A., and Attias, J. (2006). Hyperacusis in Williams syndrome: characteristics and associated neuroaudiologic abnormalities. *Neurology* 66, 390–5. doi: 10.1212/01.wnl.0000196643.35395.5f

Hagopian, L. P., Crockett, J. L., and Keeney, K. M. (2001). Multicomponent treatment for blood-injury-injection phobia in a young man with mental retardation. *Res. Develop. Disabil*. 22, 141–9. doi: 10.1016/S0891-4222(01)00063-4

Hirshfeld-Becker, D. R., Masek, B., Henin, A., Blakely, L. R., Pollock-Wurman, R. A., McQuade, J., et al. (2010). Cognitive behavioral therapy for 4-to 7-year-old children with anxiety disorders: a randomized clinical trial. *J. Consult. Clin Psychol*. 78, 498. doi: 10.1037/a0019055

Hurley, A. D. (2004). Treatment of multiple phobias and agoraphobia in a man with Down syndrome. *Mental Health Aspects Develop Disabil*. 7, 142–8.

Järvinen, A., Korenberg, J. R., and Bellugi, U. (2013). The social phenotype of Williams syndrome. *Curr. Opin. Neurobiol*. 23, 414–22. doi: 10.1016/j.conb.2012.12.006

Kershaw, H., Farrell, L. J., Donovan, C., and Ollendick, T. (2017). Cognitive behavioral therapy in a one-session treatment for a preschooler with specific phobias. *J. Cogn. Psychoth*. 31, 7–22. doi: 10.1891/0889-8391.31.1.7

Klein, A. J., Armstrong, B. L., Greer, M. K., and Brown, I. I. I. F. R. (1990). Hyperacusis and otitis media in individuals with Williams syndrome. *J. Speech Hearing Disords*. 55, 339–44. doi: 10.1044/jshd.5502.339

Klein-Tasman, B. P., and Albano, A. M. (2007). Intensive, short-term cognitive-behavioral treatment of OCD-like behavior with a young adult with Williams syndrome. *Clin. Case Stud*. 6, 483–92. doi: 10.1177/1534650106296370

Klein-Tasman, B. P., Young, B. N., Levine, K., Rivera, K., Miecielica, E. J., Yund, B. D., et al. (2022). Acceptability and effectiveness of humor-and play-Infused exposure therapy for fears in Williams syndrome. *Evid. Based Pract. Child Adoles. Mental Health*. 7, 94–111. doi: 10.1080/23794925.2021.1950080

Koegel, R. L., Openden, D., and Koegel, L. K. A. (2004). systematic desensitization paradigm to treat hypersensitivity to auditory stimuli in children with autism in family contexts. *Res. and Pract. Persons with Severe Disabil*. 29, 122–34. doi: 10.2511/rpsd.29.2.122

Kreslins, A., Robertson, A. E., and Melville, C. (2015). The effectiveness of psychosocial interventions for anxiety in children and adolescents with autism spectrum disorder: a systematic review and meta-analysis. *Child Adoles. Mental Health*. 9, 1–2. doi: 10.1186/s13034-015-0054-7

Krilčić, P., and Petranović, A. (2017). New findings on central auditory processing in Williams syndrome. *Funda Neurosci*. 4, 79–85.

Levitin, D. J., Cole, K., Lincoln, A., and Bellugi, U. (2005). Aversion, awareness, and attraction: investigating claims of hyperacusis in the Williams syndrome phenotype. J. *Child Psychol. Psychiatry* 46, 514–23. doi: 10.1111/j.1469-7610.2004.00376.x

Leyfer, O. T., Woodruff-Borden, J., Klein-Tasman, B. P., Fricke, J. S., and Mervis, C. B. (2006). Prevalence of psychiatric disorders in 4 to 16-year-olds with Williams syndrome. Am. *J. Med. Gen. Part B Neuropsych. Gen*. 141, 615–22. doi: 10.1002/ajmg.b.30344

Maiano, C., Coutu, S., Tracey, D., Bouchard, S., Lepage, G., Morin, A. J., et al. (2018). Prevalence of anxiety and depressive disorders among youth with intellectual disabilities: a systematic review and meta-analysis. *J. Affect. Disord*. 236, 230–42. doi: 10.1016/j.jad.2018.04.029

Matsumoto, N., Kitani, R., and Kalinec, F. (2011). Linking LIMK1 deficiency to hyperacusis and progressive hearing loss in individuals with Williams syndrome. *Communicat. Integrat. Biol*. 4, 208–10. doi: 10.4161/cib.4.2.14491

McGuire, J. F., Wu, M. S., and Storch, E. A. (2015). Cognitive-behavioral therapy for 2 youths with misophonia. *J. Clin. Psychiatry* 76, 3143. doi: 10.4088/JCP.14cr09343

Meng, Y., Zhang, Y., Tregoubov, V., Janus, C., Cruz, L., Jackson, M., et al. (2002). Abnormal spine morphology and enhanced LTP in LIMK-1 knockout mice. *Neuron* 35, 121–33. doi: 10.1016/S0896-6273(02)00758-4

Mervis, C. B. (2000). Klein-Tasman BP. Williams syndrome: cognition, personality, and adaptive behavior. *Mental Retardation Develop. Disabil. Res. Rev*. 6, 148–58. doi: 10.1002/1098-2779(2000)6:2<148::AID-MRDD10>3.0.CO;2-T

Mervis, C. B., and John, A. E. (2010). "Cognitive and behavioral characteristics of children with Williams syndrome: implications for intervention approaches," in *American Journal of Medical Genetics Part C: Seminars in Medical Genetics 2010 May 15*. Vol. 154, No. 2, pp. (229-248). Hoboken: Wiley Subscription Services, Inc., A Wiley Company. doi: 10.1002/ajmg.c.30263

Mervis, C. B., Klein-Tasman, B. P., and Mastin, M. E. (2001). Adaptive behavior of 4-through 8-year-old children with Williams syndrome. *Am. J. Mental Retard*. 106, 82–93. doi: 10.1352/0895-8017(2001)106<0082:ABOTYO>2.0.CO;2

Meyer-Lindenberg, A., Hariri, A. R., Munoz, K. E., Mervis, C. B., Mattay, V. S., Morris, C. A., et al. (2005). Neural correlates of genetically abnormal social cognition in Williams syndrome. *Nature Neurosci* 8, 991–3. doi: 10.1038/nn1494

Moskowitz, L. J., Braconnier, M., and Jeffay, M. (2019). "Anxiety and phobias in individuals with intellectual disabilities," in *Handbook of Intellectual Disabilities*. (Springer, Cham) (pp. 813-842). doi: 10.1007/978-3-030-20843-1_44

Ng-Cordell, E., Hanley, M., Kelly, A., and Riby, D. M. (2018). Anxiety in Williams syndrome: the role of social behaviour, executive functions and change over time. J. *Autism Develop. Disord*. 48, 796–808. doi: 10.1007/s10803-017-3357-0

Nigam, A., and Samuel, P. R. (1994). Hyperacusis and Williams syndrome. *J. Laryngol. Otol*. 108, 494–6. doi: 10.1017/S0022215100127203

Oar, E. L., Farrell, L. J., Waters, A. M., Conlon, E. G., and Ollendick, T. H. (2015). One session treatment for pediatric blood-injection-injury phobia: a controlled multiple baseline trial. *Behav. Res. Therapy* 73, 131–42. doi: 10.1016/j.brat.2015.08.002

Ollendick, T., Muskett, A., Radtke, S. R., and Smith, I. (2021). Adaptation of one-session treatment for specific phobias for children with autism spectrum disorder using a non-concurrent multiple baseline design: a preliminary investigation. *J. Autism Develop. Disor*. 51, 1015–27. doi: 10.1007/s10803-020-04582-5

Ollendick, T. H., and Davis, I. I. I. T. E. (2013). One-session treatment for specific phobias: a review of Ost's single-session exposure with children and adolescents. *Cognitive Behav. Therapy* 42, 275–83. doi: 10.1080/16506073.2013.773062

Phillips, K. D., and Klein-Tasman, B. P. (2009). Mental health concerns in Williams syndrome: intervention considerations and illustrations from case examples. *J. Res. Intellectual Disabil.* 2, 110–33. doi: 10.1080/19315860802627601

Prasad, K. C., Azeem Mohiyuddin, S. M., Anjali, P. K., Harshita, T. R., Indu Varsha, G., Brindha, H. S., et al. (2019). Microsurgical anatomy of stapedius muscle: anatomy revisited, redefined with potential impact in surgeries. *Indian J. Otolaryngol. Head Neck Surg*. 71, 14–8. doi: 10.1007/s12070-018-1510-5

Reid, A. M., Guzick, A. G., Gernand, A., and Olsen, B. (2016). Intensive cognitive-behavioral therapy for comorbid misophonic and obsessive-compulsive symptoms: a systematic case study. *J. Obsess. Compul. Relat. Disord*. 10, 1–9. doi: 10.1016/j.jocrd.2016.04.009

Rescorla, R. A. A. (1972). Theory of Pavlovian conditioning: variations in the effectiveness of reinforcement and non-reinforcement. *Curr. Res. Theory*. 3, 64–99.

Rosen, T. E., Connell, J. E., and Kerns, C. M. A. (2016). review of behavioral interventions for anxiety-related behaviors in lower-functioning individuals with autism. *Behav. Intervent*. 31, 120–43. doi: 10.1002/bin.1442

Royston, R., Howlin, P., Waite, J., and Oliver, C. (2017). Anxiety disorders in Williams syndrome contrasted with intellectual disability and the general population: a systematic review and meta-analysis. *J. Autism Develop. Disord*. 47, 3765–77. doi: 10.1007/s10803-016-2909-z

Silva, L. A., Kawahira, R. S., Kim, C. A., and Matas, C. G. (2021). Abnormal auditory event-related potentials in Williams syndrome. *Eur. J. Med. Gen*. 64, 104163. doi: 10.1016/j.ejmg.2021.104163

Slifer, K. J., Koontz, K. L., and Cataldo, M. F. (2002). Operant-contingency-based preparation of children for functional magnetic resonance imaging. *J. App Behav. Anal.* 35, 191–4. doi: 10.1901/jaba.2002.35-191

Stanyon, C. A., and Bernard, O. (1999). LIM-kinase1. *J. Biochem. Cell Biol.* 31, 389–94. doi: 10.1016/S1357-2725(98)00116-2

Stinton, C., Tomlinson, K., and Estes, Z. (2012). Examining reports of mental health in adults with Williams syndrome. *Res. Develop. Disabil.* 33, 144–52. doi: 10.1016/j.ridd.2011.09.002

Strømme, P., Bjømstad, P. G., and Ramstad, K. (2002). Prevalence estimation of Williams syndrome. *J. Child Neurol.* 17, 269–71. doi: 10.1177/088307380201700406

Thom, R. P., Pineda, J., Nowinski, L., Birtwell, K., Hooker, J. M., McGuire, J. F., et al. Virtually delivered adapted cognitive-behavioral therapy group for adults with williams syndrome and anxiety. *Cogn. Behav. Practice*. (2022). doi: 10.1016/j.cbpra.2022.02.025

Tyler, R. S., Pienkowski, M., Roncancio, E. R., Jun, H. J., Brozoski, T., Dauman, N., et al. (2014). review of hyperacusis and future directions: part I. Definitions and manifestations. *Am. J. Audiol.* 23, 402–19. doi: 10.1044/2014_AJA-14-0010

Ung, D., Selles, R., Small, B. J., and Storch, E. A. A. (2015). systematic review and meta-analysis of cognitive-behavioral therapy for anxiety in youth with high-functioning autism spectrum disorders. *Child Psychiatry Human Develop*. 46, 533–47. doi: 10.1007/s10578-014-0494-y

Whiteside, S. P., Sim, L. A., Morrow, A. S., Farah, W. H., Hilliker, D. R., Murad, M. H., et al. (2020). meta-analysis to guide the enhancement of CBT for childhood anxiety: exposure over anxiety management. *Clin. Child Family Psychol. Rev*. 23, 102–21. doi: 10.1007/s10567-019-00303-2

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," in *American Journal of Medical Genetics Part C: Seminars in Medical Genetics* 2010 May 15 Vol. 154, No. 2, pp. (277-290). Hoboken: Wiley Subscription Services, Inc., A Wiley Company. (2010)

frontiers | Frontiers in Psychiatry

TYPE Original Research
PUBLISHED 24 August 2023
DOI 10.3389/fpsyt.2023.1066286

OPEN ACCESS

EDITED BY
Mirko Uljarevic,
The University of Melbourne, Australia

REVIEWED BY
Donald A. Wilson,
New York University, United States
Michele Malaguarnera,
Principe Felipe Research Center (CIPF), Spain
Benoist Schaal,
Université de Bourgogne, France

*CORRESPONDENCE
Geneviève Petitpierre
✉ genevieve.petitpierre@unifr.ch

RECEIVED 10 October 2022
ACCEPTED 21 July 2023
PUBLISHED 24 August 2023

CITATION
De Blasio C, Dind J and Petitpierre G (2023) Odor hedonic responses in children and young people with profound intellectual and multiple disabilities.
Front. Psychiatry 14:1066286.
doi: 10.3389/fpsyt.2023.1066286

Odor hedonic responses in children and young people with profound intellectual and multiple disabilities

Catherine De Blasio, Juliane Dind and Geneviève Petitpierre*

Department of Special Education, University of Fribourg, Fribourg, Switzerland

Introduction: Odors are closely linked to emotions, play an important role in the well-being of individuals and can influence mood. Despite these crucial properties, the hedonic responses to odors of people with profound intellectual and multiple disabilities (PIMD) remain little explored.

Aim: This within-subjects study aims to examine whether children and young people with PIMD react in a differentiated way to odors evaluated as pleasant or unpleasant by neurotypical adults and, if so, with which behaviors. The influence of their global mood on their emotional responses to odors is also examined.

Method: Twenty children and young people (7–18 years old) with PIMD were exposed to four pairs of hedonically contrasted odors. A control stimulus was presented before each odorant. Five emotional responses, one physiological reaction (nausea reactions), and three responses reflecting approach toward or avoidance of the stimulus were recorded throughout the duration of the stimulus exposure. The participants' global mood status was measured before the start of the research with the French version of the Mood, Interest and Pleasure Questionnaire (Ross and Oliver, 2003).

Results: The results show that when exposed to pleasant odorants, participants kept their heads aligned with the odorant source longer, smiled longer, and produced more positive vocalizations. In contrast, unpleasant odorants elicit more pouts and grimaces. Nausea reactions occurred in the presence of unpleasant odorants. The hedonic responses were more marked during the second presentation of the stimuli. Participants with a higher MIPQ score showed significantly more emotional reactions to odors.

Conclusion: The results confirm the presence of olfactory preferences in participants with PIMD and the existence of a link between their mood, emotions and olfactory hedonic processing. They prompt the use of odors to support not only the cognitive development of this population, but also their mood and their emotional regulation abilities.

KEYWORDS

profound intellectual and multiple disabilities, olfactory hedonic processing, olfaction, mood, emotion

10.3389/fpsyt.2023.1066286

1. Introduction

Profound intellectual and multiple disabilities (PIMD) refer to profound cognitive and motor impairments, as well as health problems and/or sensory limitations, mostly due to pre- or peri-natal causes (1, 2). For caregivers concerned about this population's well-being and mental health, it is important to know how positive or negative affective messages are conveyed by individuals with very limited or non-existent expressive language (3). However, little is known about how people with PIMD express themselves, how differentiated their responses are, and whether their emotions are congruent with their internal states or the characteristics of the stimulus. This study aimed at exploring odor hedonic responses in children and young people with PIMD and to explore the link with their mood.

Odor hedonics refers to the ability to assign valences to perceived odors and express olfactory preferences. Olfactive processing includes detection, habituation, identification, discrimination and preference abilities, that can be studied separately, even if they operate closely together and interact most frequently (4). Experiencing pleasure and displeasure is a fundamental part of life and a core process of motivational and bodily state (5). Odors inducing pleasure are perceived as beneficial by the organism, which tries to approach and/or explore them. In contrast, the organism tries to move away from or reject odors causing displeasure and are perceived as negative. Emotions or feelings of positive value, characterized by enjoyment, happiness or satisfaction, often accompany odors perceived as positive, while emotions of negative value usually induce discomfort or dissatisfaction (6). However, literature in typical population shows that hedonic evaluation differs depending on age, the averages of the pleasantness ratings in children under 12 years old being usually higher than in adolescents or adults (7, 8).

A few exceptions aside, sensory studies in PIMD mainly focused on vision and audition and research on hedonic olfactory processing is still in its infancy (9). However, Vlaskamp and Cuppen-Fonteine (10) found that the reactions of children with PIMD to pleasant and potentially comforting odors were more reliably reported by their teachers than their reactions to other stimuli. An explanation might be that the reactions of children with PIMD to olfactory stimuli are less ambiguous than their reactions to other sensory stimuli, that these children rely more on the sense of smell in comparison to other senses, and/or that the emotional component, which is known to be strongly linked to odor processing, induces more explicit reactions in odorous conditions. Although, Soussignan et al. (11) found that odors elicit spontaneous highly legible reactions in a clinical population made up of children with severe developmental disorders and no verbal language, Lima et al. (12), who presented stimuli from five different sensory modalities (visual, auditory, olfactory, gustatory, and tactile) to three children with PIMD, reported heart rate and electrodermal reactions, but no behavioral responses to two odors (vanilla, orange) and one trigeminal stimulus (vinegar) supposed to induce contrasted emotional reactions. In a second study involving a 3-year-old boy with PIMD, the same authors (13) observed no responses at all for vanilla, physiological responses to orange, and both behavioral and physiological responses to vinegar. They also reported that their participants did not exhibit observable distress or defensive responses to vinegar, a potentially aversive stimulus, but a consistent increase in motor activity, occurring simultaneously with acceleration in heart rate, that nevertheless could suggest some degree of discomfort. The authors concluded that, at least under their exposure conditions in which the stimulus was presented for 5 s only, the behavioral responses of people with PIMD to odorous, respectively trigeminal stimuli, were either limited and hardly visible or absent.

The lack of scientific knowledge about the hedonic olfaction of people with PIMD is problematic because odors are particularly powerful, both in humans and animals. They may relieve pain, reduce stress and promote well-being thanks to the unique characteristics of odor processing and anatomical and psychological ties that it shares with emotions (14–16). First, the chemical senses project onto regions of the brain that also process basic affects and reward, cortical and subcortical cerebral areas involved in the perception of emotion are therefore related to the olfactory structures (17). Second, similarity judgment studies have shown that the affective dimension of smells is one of the most salient, and odors are appropriate stimuli to generate affects (18). Literature reports that they can act on mood (15) as well as on perceived emotions (19–21), the latter being linked to the activation of the autonomous nervous system. Odors can influence the way individuals interact with others and perceive their environment. They can particularly regulate positive or negative emotional states and mood. They also can modulate behavior (toward food for example), cognition, learning processes, memories and reactions to certain events, consciously or not. Finally, beyond its strong emotional tone, olfaction is also characterized by its distal component, enabling a non-intrusive approach (e.g., compared to other emotional distal senses such as gustation or texture sensitivity).

Olfactory experience begins during the third trimester of gestation thanks to the amniotic fluid and the placenta that allow the baby to be in contact with various external odorous substances (e.g., food, smoke, perfume). At the time of birth, life becomes air-dependent and the little human consolidates and diversifies their olfactory experience. Rooted in both prenatal and postnatal experiences, encounters with odors elicit the individual's various emotions and reactions, including appetence and rejection (22). Most human emotional responses to odors are triggered by association learning and encounters with certain odors in their daily lives play a big role in individuals' olfactory preferences (14).

Odors elicit behavioral and/or psycho-physiological manifestations, which often occur simultaneously (23, 24) and are observed both in typical children and adults (4). Three types of factors can modulate or alter a response: namely the characteristics of the odorant itself, those of the perceiver, or both of them together (6). The individual's internal affective state, particularly their mood is a well-known modulator of hedonic response, and vice versa (25). Because they interact closely, overlap heavily and connect in a bidirectional way, emotions and mood are often mistaken for each other (26). However, emotions refer to short-lived, event- or stimulus-driven immediate responses to the anticipation or occurrence of rewarding or punishing stimuli or events, while mood is depicted as a long-lasting affective state which is internally generated and not necessarily induced by a specific situation (27). People with PIMD, children as well as adults, are not spared from low mood or even depression. Families and professionals who describe signs of deep psychological distress in some of them, manifested by agitation, screaming, crying, but also more passive signs, such as the absence of communication, withdrawal, lack of facial expression, attribute their causes to loneliness, regression of abilities, physical pain and/or epilepsy (28). Affective disorders, which are very difficult to assess in this population,

who do not communicate verbally (3), are suspected to reach a prevalence of 30% (29). Because they both have limited access to symbolic communication, people with PIMD, like typical young children, rely heavily on their caregivers to modulate their emotions. Self-control is very challenging for them and extremely understudied, even if a few authors however report the presence of simple non-verbal strategies such as, for instance, gaze shifting, behaviorally driven situation modification or response modulation (30, 31).

Considering the need to maximize the well-being of people with PIMD and to promote support to them, and in light of the identified gaps in the current knowledge about how individuals with PIMD process pleasant or unpleasant odors and the fact that, in the general population, the hedonic processing of odors is related to mood, this study raised the three following research questions: (1) What kind of behavioral manifestations do children and young adults with PIMD express when presented with a pleasant vs. an unpleasant odorant? (2) Are these behavioral manifestations hedonically differentiated? (3) Does the global mood of children and young adults with PIMD influence their behavioral responses to odors? Do age (puberty), or feeding mode, also influence these responses?

2. Participants, materials and methods

2.1. Ethics and general design

Three specialized schools participated in the study. Ethical approval was given by the Geneva and Vaud Ethics Committees (ID: 2019-00234). Written consent was obtained from parents or legal guardians before the start of the study. To respect the well-being of the highly vulnerable participants, the session was postponed if the participant's state of alertness, fatigue or health required it. Participant's participation was permanently discontinued if they showed discomfort after two sessions.

A within-subjects experimental design was chosen to explore the research questions. The experimental manipulation of the odors appeared to be the better option to study odor processing in this population. Previous literature has indeed noticed that smelling behaviors are difficult to observe in ecological conditions and that, in such contexts, the researchers themselves are not always aware of the presence of the odors (9). We decided against creating a control group. The first reason is the insoluble problem of finding matching criteria with people with PIMD (32); the second lies in the limited constructive scope of comparative studies that focus on the difficulties of people with disabilities compared to typical people, and not on the functioning of people with disabilities *per se*.

2.2. Participants

2.2.1. Selection criteria

Direct support workers from the three participating schools were asked to select all participants meeting the following inclusion criteria: (1) presenting the key characteristics of PIMD (1); (2) being aged between 7 and 18 years and (3) attending the school for at least 6 months at the time of recruitment. Children and young people presenting food allergies or chronic airway problems with indication of daily respiratory therapy were excluded.

2.2.2. Experimental sample—demographic characteristics

Thirty-one children and young people were identified. However, four were removed from the list after a thorough check between the researchers and the schools due to uncertain key characteristics of PIMD; two families declined the participation call; three participants left the study at the very beginning of the data collection: two because of severe health problems and a third because he could not stand the experimental setting. A further two left due to organizational constraints at the time of data collection, including a hospitalization. The final sample contained 20 participants aged between 7 and 18 years ($M = 13.2$ years; SD = 3.5), comprising two 7-year-olds, one 8-year-old, two 10-year-olds, two 11-year-olds, one 12-year-old, two 13-year-olds, two 14-year-olds, one 15-year-old, two 16-year-olds, three 17-year-olds and two 18-year-olds. *Clinical characteristics*: Most participants were impacted by pre- or perinatal causes (i.e., maternal hemorrhage during pregnancy; neonatal anoxia; epileptic encephalopathy of genetic origin; neonatal herpes simplex virus infection; adrenoleukodystrophy; Christianson syndrome; Wolf-Hirschhorn syndrome; startle syndrome; partial trisomy 9; cerebral palsy; multicystic encephalomalacia). PIMD was acquired in two participants, one as a result of an acute encephalopathy when he was 2 years old, the other because of a head injury at 18 months. Etiology was unknown for two and the onset of epileptic encephalopathies uncertain for the last two (Table 1).

2.3. Odor stimuli

2.3.1. Selection of odorants

A set of 18 stimuli was preselected by the researchers in collaboration with one of the schools that initiated the research. Odors that were common or likely to be encountered by participants in their daily life were given priority. The hedonicity of the stimuli was evaluated by neurotypical adults and not by the participants on a 3-level categorical scale: U=Unpleasant; N=Neither pleasant nor unpleasant; P=Pleasant. The adult group comprised 8 men and 21 women: three 18–29 years old, twelve 30–39 years old, eight 40–49 years old, four 50–59 years old and one 60–69 years old. Odor perceived intensity was determined by the same individuals on a 9-level Likert scale (1 = no odor at all to 9 = extremely strong odor). The concentration of non-commercial odorants was manipulated to select those of medium intensity according to neurotypical judgments (33, 34). Eight hedonically contrasted odorants were selected. Two odors came from the standardized "Sniffin' Sticks Test" manufactured by Burghart Messtechnik GmbH,[1] five were produced by the Swiss manufacturer of flavors and fragrances Givaudan, Geneva, one, hand sanitizer [Sterilium®], comes from a commercially available product and was chosen because it was used by school staff for their own hand hygiene. An odorless stimulus—dipropylene glycol—was used as control.

1 www.burghart-mt.de

10.3389/fpsyt.2023.1066286

TABLE 1 Participants' characteristics.

Demographics	(%)
Gender	
Male	14 (70%)
Female	6 (30%)
Etiology	
Known or strongly suspected within	18 (90%)
Pre- and perinatal[a]	14 (70%)
Postnatal	2 (10%)
Time of onset not available	2 (10%)
Unknown	2 (10%)
Epilepsy	
Seizures controlled by medication	9 (45%)
Seizures once a month	2 (10%)
Seizures once a week	3 (15%)
Seizures once a day	1 (5%)
No epilepsy	5 (25%)
Visual impairment	
Confirmed	16 (80%)
No visual impairment	4 (20%)
Auditive impairment	
Confirmed	1 (5%)
Presumed	2 (10%)
No auditive impairment	16 (80%)
Missing	1 (5%)
Tactile impairment	
Confirmed	5 (25%)
Presumed	6 (30%)
No tactile impairment	8 (40%)
Missing	1 (5%)
Medication	
Medicated	19 (95%)
Non medicated	1 (5%)
Feeding mode	
Blended food	7 (35%)
Chunky food	3 (15%)
Enteral feeding (partial)	9 (45%)
Enteral feeding (complete)	1 (5%)

[a]Onset through seven days after birth.

2.3.2. Stimulus properties

Four of the eight stimuli were considered pleasant by typical adult raters, and four unpleasant. Intensity ranged between 3.59 and 6.45, with an average of 5.01 for pleasant and 5.59 for unpleasant stimuli on a scale of 9. Participants' stimuli familiarity, checked by their parents or legal guardians on a binary scale (0 = unfamiliar or not familiar at all; 1 = somewhat familiar or very familiar), ranged between 0.13 (lily of the valley) and 0.81 (cheese), with an average of 0.34 for pleasant and 0.51 for unpleasant stimuli on a scale of 1.

2.3.3. Stimulus delivery device

The odors were presented at ambient temperature in a 14 cm long and 1.3 cm diameter Burghart pen-like device (Burghart Medical Technology) using a tripod with an articulated arm and a clip. The device was made from an adjustable microphone stand with a telescopic 70 cm long swivel arm to prevent the researcher from bringing his or her hand too close to the participant's face and hiding their face. During the odor presentation, the tip of the stick was protected by a rubber tube to prevent direct contact with the mouth or any other part of the participants' face (Figure 1). Each stick was stored in a hermetically sealed glass container to avoid contamination and to minimize loss due to solvent evaporation between sessions.

2.3.4. Stimuli delivery sequences

Stimuli were combined to form food and non-food pairs, respectively. The first, which are usually processed retronasally, may elicit a different hedonic experience in comparison to the second, which have a high orthonasal but low retronasal familiarity (35). Food pairs were: orange-garlic and cinnamon-cheese. Non-food were: lily of the valley-sweat and summer rain-hand sanitizer odors. Each pair teamed a pleasant (O_P) with an unpleasant stimulus (O_U). See Table 2 for the characteristics of the pairs.

2.4. Experimental setting

2.4.1. Physical environment

The experiment took place in the participants' schools. Quiet, easily ventilated rooms, far from the kitchen and/or places that may generate odors (laundry), were used. To limit competing visual or auditory stimuli during the sessions, adjustments were made such as drawing the curtains, partially lowering the blinds, hiding potentially attractive objects, etc. The experimenters and the teacher were careful not to wear perfume and not to bring food into the rooms. The researcher presenting the stimuli wore cotton gloves to prevent contamination of the odorants when handling the sticks. The rooms were aired before and after each session, as well as before and after each participant. A white 12.5 cm x 12.5 cm grid was used to harmonize the flat surface behind the participant and provide marks for adjusting the zoom. It also facilitated the coding of some indicators, such as head displacements. The rest of the material consisted of a transparent pouch containing numbered cards corresponding to the steps of the study and the odorants presented. This pouch was fixed on the black screen behind the participant and the experimenter changed the cards as the stimuli were presented. Finally, the equipment included a poster informing that the experiment was in progress to avoid inopportune interruptions, photographs of the participant and experimenters, as well as pictograms to present the stages of the session to the participant and a small bell to announce the beginning and end of the test to the participant.

2.4.2. Participants' condition

The test procedure took place during school time (9–11:30 or 13:45–16:00). Sessions and lunchtime were a minimum half hour apart to limit the influence of the participants' satiety or hunger state on their hedonic olfactory responsiveness. The interval was respected independently of the participants' feeding mode (blended, chunky, partial or full enteral), the influence of which on hedonic responses is

FIGURE 1
Clip holding the delivery device with its protective cylindrical ring.

TABLE 2 Identity, concentration, origin and average values (±standard deviations) of odor properties as reported by a panel of neurotypical adults.

	Pair 1		Pair 2		Pair 3		Pair 4	
	Orange (Burghart)	Garlic (Burghart)	Cinnamon	Cheese	Lily of the valley	Sweat	Summer rain	Hand sanitizer
Hedonicity	Pleasant	Unpleasant	Pleasant	Unpleasant	Pleasant	Unpleasant	Pleasant	Unpleasant
Intensity								
M	5.00	6.45	6.10	5.07	5.36	5.66	3.59	5.21
SD	(0.92)	(0.91)	(0.93)	(1.33)	(1.02)	(1.44)	(0.93)	(1.17)
Familiarity								
M	0.48	0.38	0.29	0.81	0.13	0.33	0.48	0.52
SD	(0.48)	(0.49)	(0.46)	(0.40)	(0.35)	(0.48)	(0.51)	(0.51)
Concentration								
%	NA	NA	10	1	20	1	50	NA

NA, Non available.

unknown. As epilepsy is a high comorbidity in people with PIMD and can cause loss of consciousness and altered perception of the environment, monitoring it throughout the session was of great importance. The presence of a direct support worker who knew the participant well was systematically organized during the data collection to alert the researchers to possible seizures and ensure the participant's well-being and safety. Before each session, the researchers asked about the participant's current state of health and alertness and decided with the teacher if the session could take place or would have to be postponed. Finally, determining the positioning of the participant during the presentation of the odors was essential so that, to the extent of their possibilities, the participant had as much freedom of movement as possible. For this reason, a member of the research team met every participant a few weeks before the data collection and asked about the type of personal seat used by the participant to be sure that this seat allows enough freedom of movement and minimizes tonic and motor fluctuations. If necessary, a better postural adaptation was sought with the participant's physio- and/or occupational therapists, i.e., introduction of a headrest or support behind the neck for the sessions. The pilot study confirmed that the sitting position in the participant's own seat was favorable to present the odors.

2.4.3. Social environment

The experiment was attended by the participant, a direct support worker who knew the participant well, and two researchers. At the time the odors were presented, the experimenters and the teacher refrained from speaking to the participant so that they could focus their full attention on the olfactory stimuli. Nonverbal interactions with the participant were also kept to a minimum (without appearing totally indifferent) so as not to bias the observations.

2.5. Experimental procedure

2.5.1. Experimental design

To avoid a possible fatigue and olfactory saturation effect, data were collected in four sessions –one pair of odors per session– spaced about 1 week apart (one session/week for 4 weeks). The total assessment time for each session was 4 min per participant. Of the 20 participants, 19 completed the four sessions, i.e., were exposed to the eight odors, while one was only exposed to six odors as he was absent at a session. The dataset results in a total of 158 records ($19 \times 8 + 1 \times 6$). Each session involved the odor pairs and the control stimulus being presented twice (T1 and T2) alternately for 30 s each. Thirty-second exposure was chosen to be enough long to observe how the participants react and cope emotionally with the odor. For the interstimulus interval, 30 s are usually considered enough time for the olfactory recovery between two odorous stimuli (36). We decided to not counterbalance the order of the stimuli across the sessions to (1) avoid exposing participants to two successive unpleasant stimuli at the start or end of the session and (2) because in contrast to typical participants, participants with severe neurodevelopmental disorders do not exert control over their spontaneous facial configuration (11). Their responses are therefore unlikely to be biased by social expectations. Table 3 shows the sequence of stimuli presentation for each pair.

2.5.2. Procedure

The experiment was preceded by (1) a pilot study and (2) a preliminary contact with the participant. The pilot study was conducted with five children and young adults with PIMD who were not part of the sample. The aim was to decide how the stimuli should be presented (sticks or bottle), to test the procedure (session duration, tolerance of participants toward the setting, experimenters' and participants' locations) and to confirm the predicted indicators. The pilot study showed the advantage of sticks over bottles in terms of ease of handling. It also showed the need to add the protective rubber tube at the end of the stick. It confirmed that the participants were tolerant toward the setting and the duration of the sessions. Finally, it confirmed the selection of the panel of indicators.

The purpose of the preliminary meeting was to allow the experimenter to meet each participant for half a day, around 2 weeks before the experiment, to get to know them and discuss adaptations and organizational points with their teachers (i.e., when, morning or afternoon, the participant's state of awareness was more favorable, determining the positioning of the participant during the presentation etc.).

2.5.3. Experimenters' operational roles

Two researchers and the participant's teacher or educator were present during the data collection. The researcher, who presented the stimuli to the participant, stood either to the right or left of the participant, alternating from one session to another to avoid participant orientation bias. The second researcher oversaw time indications and made sure that both cameras were working properly. The participant's teacher or educator attended the session for the safety reasons described above. The researcher who presented the stimuli to the participant opened the stick, fixed it on the clip and presented it about 2 cm in front of the participant's nose, between their nose and chin (12). Sessions were videotaped by 2 digital camcorders located 2 meters in front of the participant, one focusing on the participant's face, the other on his or her body. For close-up framing, the camera focus on the participant's face was 3 background black screen squares high by 5 wide (37.5×62.5 cm). For wide frame, the participant had to be seen from head to toe. Neither the researchers nor the teacher or educator spoke to the participant between the beginning and the end of the presentation of the odorants. The researchers were not blind to the kind of stimulus presented.

2.6. Behavioral variables and coding

2.6.1. Definition of the variables

Five hedonic indicators, i.e., smiles, positive vocalizations, pouts, negative vocalizations, emotional outburst, plus nausea reactions as physical response, were recorded throughout the duration of exposure to the stimuli. The selection of these indicators was based on two rationales: (1) that they have been reported in the developmental literature [(37) for a systematic review] and that they (2) belonged to the register of behaviors of people with PIMD (3, 9, 38, 39). As a positive hedonic attribution usually induces approach or avoidance behavior (5), three more indicators were used, namely: head alignment on the stick, nose/hand coordination on the stick and attempts to reject the stick, resulting in nine indicators in total. Depending on the relevance for the target behavior, five behaviors were coded on duration (head alignment on the stick, nose/hand/stick coordination, smile, positive vocalizations, negative vocalizations) and four on occurrences (pouts, attempts to reject the odor, emotional outbursts and nausea reactions). This choice was validated by the practicality of the measurement, methodological recommendations (40), coding procedure used in olfaction research on newborns and young children (41), and the results of the pilot study.

TABLE 3 Sequence of presentation of pairs of odors.

	T1				T2			
Stimulus	Control stimulus	Pleasant odorant	Control stimulus	Unpleasant odorant	Control stimulus	Pleasant odorant	Control stimulus	Unpleasant odorant
Duration in seconds	30	30	30	30	30	30	30	30
Time flow in seconds	0–30	30–60	60–90	90–120	120–150	150–180	180–210	210–240

2.6.2. Approach or avoidance responses

The head alignment on the stick was coded when the participant was holding their head up (immobile or with very slight movements) and aligned on the axis of the stick (the projected straight line as a continuation of the stick reaching the nostrils). With regard to the stick rejection, the behavior had to appear within 2 s after the insertion of the stick, the participant pushing either the holder or the stick away from under their nose or genuinely turning their head away while it was aligned with the odorant at the moment of presentation, i.e., the angle of the head in relation to the stick ends up at about 45° (perpendicular). Nose/hand coordination on the stick was coded when the participant grasped the stick under their nose without moving it maintaining the nose motionless aligned with the stick, or making small lateral head movements the nose on the stick or when they applied lateral or vertical movements to the stick in the proximity of their nose.

2.6.3. Hedonic responses

Smiling was defined as the elevation of the two corners of the mouth (42), with the mouth open or closed (43); rictus due to epileptic manifestations or paradoxical smiles and social smiles directed toward persons were excluded. Pouting referred to momentary deformations of the face. i.e., frowning the nose to express displeasure (pouting) or disgust (grimacing). Positive vocalizations referred to sounds produced with the vocal cords in a "positive" (e.g., cooing with pleasure, babbling) mode (44), or to vocal sounds made while smiling. Plaintive vocalizations, such as whining, moaning, screaming and/or irritated/aggrieved/enervated vocal sounds, were defined as negative vocalizations. The sounds could be single, regardless of their duration, or linked, i.e., a series of close vocal sounds, each one less than 1 s apart from the one preceding or following it. Emotional outburst refers to massive emotional reaction (e.g., cries of distress, crying, trembling, paradoxical laughter).

2.6.4. Physiological response

Nausea reactions refer to rapid and intense contraction of the pharynx, causing the opening of the mouth and usually the protrusion of the tongue (heave).

2.6.5. Coding

The coding was carried out by a team of three coders experienced in video analysis, two of them with the target group. Two had a doctoral degree in special education, the third a MA degree. Only one coder was fully blind to the odor being tested. The coding was done both in continuous and real-time measurement. The direct support workers were consulted when the coders had a doubt about certain behaviors. The coding procedure was set up in the EUDICO Linguistic Annotator (ELAN) Software version 5.9 (Max Planck Institute for Psycholinguistics, Nijmegen, The Netherlands). A refined reliability assessment procedure using EasyDIAg algorithm (45) was used to check both the observers' agreement about presence/absence of a behavior and the temporal overlap of their coding. The reliability value (kappa) was calculated on 20% of the data. The accuracy of the grid makes it possible to reach a satisfactory interobserver agreement for 5 out 9 indicators: head alignment on the stick ($k = 0.83$); smiling ($k = 0.72$); pouting ($k = 0.77$); rejecting the stick ($k = 0.85$); positive vocalizations ($k = 0.80$). The interjudge of four indicators could not be calculated due to too few data. Full details on the coding procedure are available in a previous publication (33).

2.7. Measurement of mood

Nonverbal communication prevents people with PIMD from self-reporting on their emotional state (46, 47). The Mood, Interest and Pleasure Questionnaire (MIPQ) (48) was used to identify the participants' affects. This 23-item Likert hetero-reported questionnaire is based on definitions of low mood and anhedonia outlined under the criteria for major depressive episode in DSM-IV (49), as well as on studies validating behavioral indices of the measured constructs in the population with PIMD (50). The total score is a maximum of 92 points, high scores denoting high mood and interest and pleasure levels. Both the internal consistency and interrater reliability of the French version are good [Cronbach's *alpha* = 0.97 for the total scale; Pearson $r = 0.90$, (51)]. The study required the participant's teacher or a member of the educational team to complete the MIPQ with reference to the two preceding weeks.

2.8. Statistical analyses

As the data was not normally distributed, nonparametric statistics were used to perform the analyses. A Wilcoxon signed ranks test was used to compare the responses between pleasant and unpleasant modalities as well as at time 1 and time 2. Effect sizes for the Wilcoxon signed ranks test were calculated according to Pallant (52) that is by dividing the Z value of the test by the square root of the number of observations [$r = Z/\sqrt{(n1 + n2)}$]. Confirmatory analyses comparing control, pleasant and unpleasant modalities, were performed with Friedman tests (χ^2), alpha level at 0.05 (unilateral) with Dunn-Bonferroni *post hoc* tests with a correction for multiple testing being used for pairwise comparisons. Effect sizes were carried out with Kendall W statistics. The role of the mood, age (puberty), and feeding mode, on the olfactory responses was investigated thanks to generalized linear mixed models (t). The participants were divided into two groups based on their MIPQ scores for this analysis. Group 1 includes participants who scored less than 60 in total and/or obtained low scores on 2 out of the 3 MIPQ subscales (i.e., positive mood score ≤ 23, interest score ≤ 17 or negative mood score ≤ 17) and were therefore considered to have a low general mood. Group 2 includes participants who scored above these cutoffs. Two groups were also formed to study the influence of puberty ("up to 11 years" vs. "12 years old and over"), and two others to study feeding mode ("blendy and chunky" vs. "complete or partial enteral feeding mode"). All analyses were performed using SPSS software version 26.0.0 (IBM Corp.). No analysis was carried out for the indicator "emotional outburst, which did not obtain any occurrences.

3. Results

3.1. Participants' responses to pleasant vs. unpleasant odors and control stick

The first analysis aimed to answer the first two research questions, namely, to explore if the participants exhibited differentiated behaviors in the presence of pleasant vs. unpleasant odors and, if so, how? For this analysis, the participants' ($n = 20$) responses to all pleasant odorants (orange, cinnamon, lily of the valley, summer rain) were compared to those to all unpleasant odorants (garlic, sweat, cheese,

10.3389/fpsyt.2023.1066286

TABLE 4 Responses per odor (times 1 and 2 aggregated).

Responses (in seconds)	Orange $N = 20$		Lily of the valley $N = 20$		Summer rain $N = 19$		Cinnamon $N = 20$		Garlic $N = 20$		Sweat $N = 20$		Hand sanitizer $N = 19$		Cheese $N = 20$	
	M	SD	*M*	SD	*M*	SD	*M*	SD	*M*	SD	*M*	SD	*M*	SD	*M*	SD
Positive																
Smiling	2.93	5.60	5.44	7.78	3.75	6.72	5.17	10.33	2.36	6.41	2.01	5.37	4.00	6.19	3.33	6.24
Positive vocal.	0.52	1.00	0.56	1.16	1.37	3.46	2.08	5.79	0.21	0.47	0.20	0.63	0.36	0.85	0.84	2.88
Negative																
Pouting	0.70	1.38	1.25	3.04	0.11	0.32	0.60	1.10	2.10	2.57	1.70	2.66	1.37	1.80	1.65	1.93
Negative vocal.	0.37	1.67	0.16	0.55	0.09	0.39	1.13	4.12	0.14	0.35	0.17	0.42	0.20	0.77	0.72	2.00
Physiol. react.																
Nausea react.	0	0	0	0	0	0	0	0	0.25	0.91	0.15	0.49	0.05	0.23	0	0
Approach/escape																
Head alignment	23.84	13.32	26.45	15.17	31.74	16.97	26.12	17.03	19.76	13.66	22.23	14.83	27.81	15.08	25.84	16.46
Nose/hand stick	1.72	4.66	1.28	3.95	0.72	2.17	1.52	3.71	1.27	2.55	0.67	1.99	0.48	1.51	1.04	3.08
Reject. the stick	0.25	0.55	0.20	0.52	0.21	0.42	0.10	0.31	0.40	0.68	0.40	0.68	0.21	0.54	0.20	0.41

hand sanitizer) on time 1 (T1) and 2 (T2) aggregated (Table 4). For greater clarity of the results, we replaced the medians (Mdn), which were often small, by the means (*M*) and the standard deviations (*SD*) in the Wilcoxon statistics. The results show that the participants responded significantly differently when presented with pleasant vs. unpleasant odors for five of the nine responses studied. They emitted more positive vocalizations ($M_{Pleasant} = 1.09$ s, SD = 3.51, $M_{Unpleasant} = 0.36$ s, SD = 1.56, $Z = -3.05$, $p = 0.001$, $r = 0.25$) and smiled longer when presented with pleasant odors than with unpleasant odors ($M_{Pleasant} = 4.11$ s, SD = 7.75, $M_{Unpleasant} = 2.81$ s, SD = 6.08, $Z = -1.77$, $p = 0.039$, $r = 0.14$). Faced with unpleasant odors, the participants exhibited significantly more pouting in comparison to pleasant odors ($M_{Unpleasant} = 1.78$, SD = 2.27, $M_{Pleasant} = 0.70$, SD = 1.82, $Z = -4.60$, $p < 0.001$, $r = 0.37$). It was also noticed that nausea reactions were manifested only in the presence of unpleasant stimuli ($M_{Unpleasant} = 0.12$, SD = 0.54, $M_{Pleasant} = 0$, SD = 0, $Z = -2.06$, $p = 0.020$). Because of the scarcity of the nausea reaction manifested by four participants with three unpleasant odors out of four, i.e., garlic, sweat and hand sanitizer triggered such a response, but not cheese, the effect size was small ($r = 0.17$). Finally, about approach or avoidance behaviors, participants aligned their heads in line with the source significantly longer when they were exposed to the pleasant odors compared to the unpleasant ($M_{Pleasant} = 27.24$ s, SD = 15.76, $M_{Unpleasant} = 24.32$ s, SD = 15.18, $Z = -2.11$, $p = 0.017$, $r = 0.17$).

A verification of the results using a Friedman test, which is a more conservative test allowing more than two conditions to be compared, and thus enabling the previous comparison to be extended to the control modality, confirms the significant influence of the hedonic modality on two indicators. The results confirm the significant effect of the hedonic modality on the duration of participants' head alignment on the stick [X^2 (2, $n = 20$) = 6.30, $p = 0.021$], with an effect size that could be described as small ($W = 0.178$) and pairwise comparisons with Dunn-Bonferroni corrected for multiple testing showing that the head alignment is significantly longer when participants are exposed to pleasant compared to unpleasant odors ($p = 0.026$). The comparison of the three modalities (un/pleasant and control) also showed a significant effect of the hedonic modality on pouting [X^2 (2, $n = 20$) = 18.29, $p = 0.000$], with an effect size that could be described as moderate to large ($W = 0.457$) and pairwise comparisons with Dunn-Bonferroni correction revealing that the participants exhibited significantly more pouting when faced with unpleasant odors compared to pleasant ($p = 0.002$), as well as when they faced unpleasant odors compared to control ($p = 0.002$).

3.2. Participants' responses during the first exposure to odors vs. the second

The second analysis aimed to study whether the differences in hedonic responses were already present during the first exposure to the odor (T1) in comparison to the second (T2). The analyses were performed on all participants ($n = 20$). The results show that differentiated responses were already present during the first exposure. At the first exposure, participants vocalized positively significantly longer in the presence of pleasant odors ($M_{Pleasant} = 0.53$ s, SD = 1.67, $M_{Unpleasant} = 0.13$ s, SD = 0.35, $Z = -2.56$, $p = 0.006$) and made more pouts in the presence of unpleasant ones ($M_{Unpleasant} = 0.76$, SD = 1.12, $M_{Pleasant} = 0.33$, SD = 0.96), $Z = -3.41$, $p < 0.001$, with mean effect sizes ($r = 0.20$ and $r = 0.27$ respectively). With a smaller effect size ($r = 0.14$), participants smiled significantly more in the presence of pleasant odors than in the presence of unpleasant odors ($M_{Pleasant} = 2.19$ s, SD = 4.17, $M_{Unpleasant} = 1.43$ s, SD = 3.05, $Z = -1.72$, $p = 0.043$).

At the second exposure, participants vocalized positively for significantly longer in the presence of pleasant odors ($M_{Pleasant} = 0.59$ s, SD = 2.19, $M_{Unpleasant} = 0.27$ s, SD = 1.42, $Z = -2.39$, $p = 0.009$, $r = 0.19$). In contrast, they made significantly more pouting and faces ($M_{Unpleasant} = 0.95$, SD = 1.54, $M_{Pleasant} = 0.33$, SD = 0.97, $Z = -3.89$, $p < 0.001$) and longer negative vocalizations ($M_{Unpleasant} = 0.195$ s, SD = 0.71, $M_{Pleasant} = 0.12$, SD = 0.86, $Z = -2.04$, $p = 0.021$), in the presence of unpleasant stimuli, with moderate ($r = 0.31$) and small ($r = 0.16$) effect sizes, respectively. They exhibited nausea reactions only with unpleasant odorants ($M_{Unpleasant} = 0.08$, SD = 0.31, $M_{Pleasant} = 0$,

10.3389/fpsyt.2023.1066286

TABLE 5 Summary of the significant results for the different measurement times.

Responses	Sign. asymp *p*/effect size *r*		
	T1 $N = 20^a$	T2 $N = 20^a$	T1 + T2 $N = 20^a$
Positive			
Smiling	*/0.14	*/0.13	*/0.14
Positive vocalizations	**/0.20	**/0.19	**/0.25
Negative			
Emotional outburst			
Pouting, making a face	**/0.27	***/0.31	***/0.37
Negative vocalizations		*/0.16	
Physiological reaction			
Nausea reactions		*/0.17	*/0.17
Approach/escape behavior			
Head alignment on the stick		*/0.15	*/0.17
Nose/hand on the stick			
Rejecting the stick			

*$p < 0.05$, **$p < 0.01$, ***$p < 0.001$, Pleasant odors > Unpleasant odors; Pleasant odors < Unpleasant odors.
[a]19 participants completed 4 sessions while one participant completed only 3 sessions.

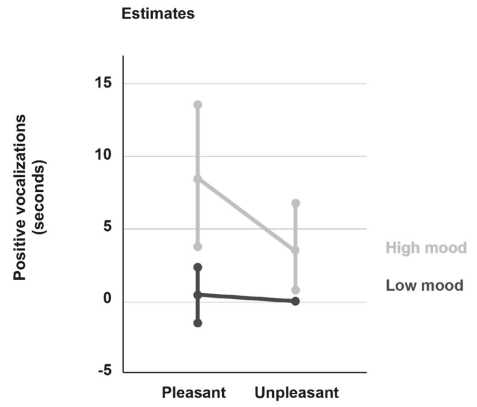


FIGURE 2
Effect of mood and odor pleasantness on average (±standard deviation) duration of positive vocalizations.

SD = 0, $Z = -2.12$, $p = 0.034$, $r = 17$). With smaller effect sizes, participants held their heads aligned on the stick significantly longer ($M_{Pleasant} = 14.02$ s, SD = 9.62, $M_{Unpleasant} = 11.92$ s, $SD = 8.34$, $Z = -1.94$, $p = 0.026$, $r = 0.15$) and smiled more in the presence of pleasant odors ($M_{Pleasant} = 2.14$ s, SD = 4.29, $M_{Unpleasant} = 1.48$ s, SD = 3.75, $Z = -1.65$, $p = 0.049$, $r = 0.13$).

Emotional reactions at Time 1 and Time 2 considered separately are consistent with the previous analysis on T1 and T2 aggregated, with participants showing, on average, more positive emotional reactions when presented with pleasant odors and more negative emotional reactions when presented with unpleasant odors. However, we note only 3 significant indicators out of 9, at T1, but 6 out of 9, at T2. Table 5 shows the significant emotional reactions in each of the odorant presentation times.

3.3. Impact of the mood on the participants' olfactory hedonic responses

The purpose of this analysis was to answer the research question about the influence of the participants' general mood on their behavioral responses to odors. For this analysis, the participants were divided into two groups based on their MIPQ scores. The first group includes seven participants considered to have a low general mood (score total < 60); the other 13 participants were considered to have a high general mood (score total > 60). The mean overall MIPQ score is $M = 46.12$ (SD = 6.64) for the low mood group and $M = 74.54$ (SD = 6.57) for the high mood group. A generalized linear mixed model analysis ($\alpha = 0.05$) was performed with group membership (high vs. low mood), the hedonic value of the stimulus (pleasant vs. unpleasant) and their interaction as independent variables.

The results show an interaction effect of mood and hedonic value of the stimuli on positive vocalizations. On average, participants vocalized positively longer when presented with pleasant odors ($M = 0.47$ s, SD = 0.14) than when presented with unpleasant odors [$M = 0.18$ s, SD = 0.07, $F(1, 154) = 8.56$, $p = 0.004$]. In addition, participants with a high mood vocalized longer ($M = 0.62$ s, SD = 0.19) than those with a low mood [$M = 0.03$ s, SD = 0.03, $F(1,154) = 8.82$, $p = 0.003$]. The significant interaction effect between the two factors indicates that the response of participants with high mood to pleasant vs. unpleasant odors differs significantly from those of participants with low mood (see Figure 2).

About simple interactions, the results show a significant main effect of hedonic valence of odors on the head alignment duration on the stimulus [$F(1, 154) = 11.19$, $p = 0.001$] with participants remaining aligned significantly longer on pleasant ($M = 28.53$, SD = 2.2) than on unpleasant odorants ($M = 25.39$, SD = 2.26). This result is consistent with that of the first analysis. A second main effect was noted for this indicator [$F(1, 154) = 8.01$, $p = 0.005$], participants with low mood aligning their head on the stick for significantly longer ($M = 33.13$, SD = 3.91) than participants

with high mood ($M = 20.79$, SD = 1.93). The third effect is that of participants' mood on smile duration [$F(1, 154) = 6.69$, $p = 0.011$], participants with high mood smiling significantly longer ($M = 4.86$, SD = 1.51) than those with low mood ($M = 0.76$, SD = 0.47). With respect to positive vocalizations, two main effects were found: one with mood [$F(1, 154) = 8.82$, $p = 0.003$], the other with the hedonic valence of odors [$F(1, 154) = 8.56$, $p = 0.004$]. Participants with high mood vocalized positively significantly longer ($M = 0.62$, SD = 0.19) than those with low mood ($M = 0.03$, SD = 0.03), and pleasant odors triggered significantly longer positive vocalizations ($M = 0.47$, SD = 0.14) compared to unpleasant ($M = 0.18$, SD = 0.07). Finally, a significant main effect was found with respect to negative vocalization duration and mood [$F(1, 154) = 4.84$, $p = 0.029$], with participants with high mood vocalizing negatively significantly longer ($M = 0.23$ s, SD = 0.10) than those with low mood ($M = 0.001$, SD = 0.001).

3.4. Impact of the age and the feeding mode on the participants' olfactory hedonic responses

No interactions effect were found between the hedonic value of the stimuli and the participants' age, or their feeding mode, on the behavioral responses to odors.

4. Discussion

To date, the abilities of individuals with PIMD to express emotions have mainly been extrapolated from reporting by proxies (11). This research offers a systematic analysis of the responses of participants with PIMD to pleasant and unpleasant odorants. Three results stand out: first, the participants expressed hedonically differentiated and congruent responses to odorous stimuli of contrasted valence; second, their expressive and vocal repertoires toward odor-borne hedonic modalities seem basically the same as those observed in typical pre-verbal populations; third, the results show that their general mood has a significant influence on their emotional reactions.

Emotions are adaptive and serve as a basis for reacting to contextual events and forming representations. They also enable organisms to signal what contributes to their well-being (53). In this sense, confirming the existence of emotional discrimination abilities in individuals with PIMD is an important step and suggests a first lever for understanding their emotional balance. To our knowledge, it is the first time that an interaction effect of both mood and stimuli valence is found in this population. In a study involving 27 participants with severe and profound intellectual disability, Vos et al. (54) found single effects between stimuli valence, mood and participants' behavioral emotion score, but they did not find any links between the three parameters. In our research, participants with low mood expressed significantly fewer contrasted emotional reactions both to pleasant and unpleasant odors, in comparison to peers with higher mood. Three models studied the link between mood and emotions (54): the Positive Attenuation model suggests that depressed people show attenuated physiological responses to stimuli with a positive valence, but that they do not differ from nondepressed individuals in their reaction to unpleasant, negatively valenced, stimuli (55); the Negative Potentiation hypothesis states that depression increases reactivity to negative stimuli that are felt even more negatively in comparison to nondepressed people, positive stimuli reactivity remaining preserved (56); finally the Emotion Context Insensitivity theory (57), with which our results are best aligned, states that low mood, depression risk, and especially diagnosed depression, lead to a general disengagement with the environment and a lack of contextually appropriate reactions to both *positive and negative* stimuli. Of course, emotion reactivity and emotional behaviors are two different sides of the same coin, but the comparison with these theories may draw attention to the influence of mood in the responses of people with PIMD to hedonic stimuli. It also highlights the importance of assessing emotional multiaxial indices across these individuals' development and contexts, as for typical ones (58). Finally, our findings raise the question of the gradation of the emotions expressed, as pleasant odors trigger both significant emotional and appetitive responses, and unpleasant only emotional ones. This result can be interpreted in different ways, either people with PIMD only have limited experience of opportunities to disengage, which seems to be contradicted by the statistically significative presence of rejection reactions to odorous stimuli of any valence vs. controls; or unpleasant stimuli are sufficient to elicit negative hedonic facial reaction, but not gestural responses, which might suggest a gradation of reactions. A greater variety of responses, which was not part of the research questions, was observed during the second stimulus exposure. With regard to this result, two points come to mind. Either the repetition of the stimulus presentation offers additional time for odor exploration and helps the participants to confirm and enrich their first impressions and reactions. Or their responses are functionally correlated, and graded in terms of intensity of response between first and second presentation. From an applied perspective, and in terms of information processing and pedagogical implications, a confirmation of these interpretations might clarify the fact that the hedonic response is not fully expressed during the first contact with the stimulus and that a second exposure is necessary to trigger a fuller and more marked expression of preferences.

4.1. Limitations

Despite various precautions, this study has several limitations. The perceptual properties that is intensity, familiarity, and the trigeminal properties, were only partially or not controlled at all while this may play a salient role on odor perception and especially valence attribution (59). A second limitation is that only one of the three coders was fully blind to the experimental conditions. Another weakness is that the sample, which was heterogeneous in terms of disabilities and syndromes, can question the integrity of the participants' olfactory system. First because, as in the typical population, they are exposed to acquired factors, such as sinonasal and upper respiratory tract infections that can alter olfaction (60). Second, because individuals with developmental delay or epilepsy are overrepresented in pediatric population with radiological anomalies of the olfactory system (61). Although the literature did not report any relationship between the participants' disabilities and syndromes and olfaction, olfactory impairment has been reported in mesial temporal lobe epilepsy due to unilateral

hippocampal sclerosis (62), and in prenatal hypoxia or herpes simplex virus in rare animal research (63, 64). It is also well known that medication can modify olfactory perception and people with PIMD are heavy consumers of medication. In our study, a medical student and a professor of pharmacology helped us to analyze the participants' medications and to calculate the percentage of those known to interfere with the sense of smell. The analysis was based on the literature as well as the monographs about Swissmedic-approved drugs.[2] Thirty-six active constituents (see Supplement 1) were identified corresponding to nearly 50 brand names, among which four were known for a potential side effect on olfaction, i.e., Topiramate (an antiepileptic), Risperidone (an antipsychotic), morphine, and Levothyroxine (hormones). Two participants were taking Topiramate, two Risperidone, one morphine and one hormonal medication; we cannot rule out that side effects on olfaction could have occurred in one or more of them. The wide participants' age range may explain why no age effect on olfactory preferences was found, in contrasts to the typical population (22). A control group would have been welcome. Finally, asking typical children, and not typical adults, to characterize the intensity as well as the hedonic dimension of the stimuli would have made it possible to take better account of the developmental factor.

5. Conclusion and perspectives

In spite of these limitations, this study provides a more in-depth knowledge of the olfactory hedonic functioning of children and adolescents with PIMD. It also shows that despite the severity of the participants' impairment, odors trigger clearly contrasted emotions in most participants, emphasizing the relevance of using olfactory simulation for cognitive, emotional, regulation, and well-being purposes. In this approach, given their strong interconnections, the use of odors to support emotional expression and mood regulation opens particularly interesting intervention perspectives, including the use of pleasant potentially comforting odors for therapeutic interventions. The findings should invite caregivers to pay systematized attention to somatic and whole-body responses, i.e., nauseous reactions, in daily and research settings when people are nonverbal or have few means of action on the world. Alternatives have to be found to substances inducing aversive reactions, e.g., the hand sanitizer that is used daily by educational personnel working with the participants. Finally, the findings remind us that emotional competence supposes access to the full range of emotional experiences, and that the presence of negative emotions, because of their adaptive function, is not worrying in itself, contrary to a blunted or overly vivid emotional expression (65).

Data availability statement

The consolidated data supporting the conclusions of this article is available on request.

2 https://www.swissmedicinfo.ch/?Lang=EN

Ethics statement

The studies involving human participants were reviewed and approved by Geneva and Vaud ethics committees (ID: 2019-0023 4), which are part of the national organisation Swissethics. Written informed consent to participate in this study was provided by the participants' legal guardian/next of kin.

Author contributions

GP and JD: concept and design. CB, JD, and GP: acquisition, analysis, and interpretation of data. GP and CB: drafting of the manuscript. GP: critical revision of the manuscript. CB and GP: statistical analysis. All authors contributed to the article and approved the submitted version.

Funding

This work was supported by Givaudan Foundation, Stiftung "Heilpädagogisches Zentrum" (Special Education Foundation), and University of Fribourg.

Acknowledgments

The authors express our gratitude to all participants, their families, and their schools, in particular the Clair Bois Foundation which initiated the study, as well as to Dr Germaine Gremaud for her help with coding and to Ms Graziela Leite Costa (medical student) and Dr med Jelena Vukovic who helped us to discriminate which drugs could affect the olfactory system.

Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

Publisher's note

Supplementary material

The Supplementary material for this article can be found online at: https://www.frontiersin.org/articles/10.3389/fpsyt.2023.1066286/full#supplementary-material

References

1. Nakken H, Vlaskamp C. A need for a taxonomy for profound intellectual and multiple disabilities. *J Policy Pract Intellect Disabil*. (2007) 4:83–7. doi: 10.1111/j.1741-1130.2007.00104.x

2. Ponsot G, Boutin A. Le polyhandicap, Une situation particulière de handicap [profound intellectual and multiple disabilities, a particular situation within disabilities] In: P Camberlein and G Ponsot, editors. *La personne polyhandicapée: La connaître, l'accompagner, la soigner [people with profound intellectual and multiple disabilities: Knowing them, accompanying them, caring for them]*. Paris: Dunod (2021, 2021).

3. Adam D, Oliver C. The expression and assessment of emotions and internal states in individuals with severe or profound intellectual disabilities. *Clin Psychol Rev*. (2011) 31:293–306. doi: 10.1016/j.cpr.2011.01.003

4. Cameron EL. Olfactory perception in children. *World J Otorhinolaryngol Head Neck Surg*. (2018) 4:57–66. doi: 10.1016/j.wjorl.2018.02.002

5. Becker S, Bräscher AK, Bannister S, Bensafi M, Calma-Birling D, Chan RCK, et al. The role of hedonics in the human affectome. *Neurosci Biobehav Rev*. (2019) 102:221–41. doi: 10.1016/j.neubiorev.2019.05.003

6. Rouby C, Pouliot S, Bensafi M. Odor hedonics and their modulators. *Food Qual Prefer*. (2009) 20:545–9. doi: 10.1016/j.foodqual.2009.05.004

7. Bensafi M, Rinck F, Schaal B, Rouby C. Verbal cues modulate hedonic perception of odors in 5-year-old children as well as in adults. *Chem Senses*. (2007) 32:855–62. doi: 10.1093/chemse/bjm055

8. Brothánková P, Vodička J, Bažant J. The influence of a child's age on the evaluation of smells and their hedonistic assessment. *Eur J Pediatr*. (2021) 180:745–9. doi: 10.1007/s00431-020-03778-2

9. Roemer M, Verheul E, Velthausz F. Identifying perception behaviours in people with profound intellectual and multiple disabilities. *J Appl Res Intellect Disabil*. (2017) 31:820–32. doi: 10.1111/jar.12436

10. Vlaskamp C, Cuppen-Fonteine H. Reliability of assessing the sensory perception of children with profound intellectual and multiple disabilities: a case study. *Child Care Health Dev*. (2007) 33:547–51. doi: 10.1111/j.1365-2214.2007.00776.x

11. Soussignan R, Schaal B, Schmit G, Nadel J. Facial responsiveness to odours in normal and pervasively developmental disordered children. *Chem Senses*. (1995) 20:47–59. doi: 10.1093/chemse/20.1.47

12. Lima M, Silva K, Amaral I, Magalhães A, de Sousa L. Beyond behavioural observations: a deeper view through the sensory reactions of children with profound intellectual and multiple disabilities. *Child Care Health Dev*. (2011) 39:422–31. doi: 10.1111/j.1365-2214.2011.01334.x

13. Lima M, Silva K, Magalhães A, Amaral I, Pestana H, de Sousa L. Can you know me better? An exploratory study combining behavioural and physiological measurements for an objective assessment of sensory responsiveness in a child with profound intellectual and multiple disabilities. *J Appl Res Intellect Disabil*. (2012) 25:522–30. doi: 10.1111/j.1468-3148.2012.00698.x

14. Kadohisa M. Effects of odor on emotion, with implications. *Front Syst Neurosci*. (2013) 7:66. doi: 10.3389/fnsys.2013.00066

15. Richardson JT, Zucco GM. Cognition and olfaction: a review. *Psychol Bull*. (1989) 105:352–60. doi: 10.1037/0033-2909.105.3.352

16. Yeshurun Y, Sobel N. An odor is not worth a thousand words: from multidimensional odors to unidimensional odor objects. *Annu Rev Psychol*. (2010) 61:219–41. doi: 10.1146/annurev.psych.60.110707.163639

17. Berridge KC, Robinson TE. Parsing reward. *Trends Neurosci*. (2003) 26:507–13. doi: 10.1016/S0166-2236(03)00233-9

18. Schiffman SS. Physicochemical correlates of olfactory quality. *Science*. (1974) 185:112–7. doi: 10.1126/science.185.4146.112

19. Schiffman SS, Sattely-Miller EA, Suggs MS, Graham BG. The effect of pleasant odors and hormone status on mood of women at midlife. *Brain Res Bull*. (1995) 36:19–29. doi: 10.1016/0361-9230(94)00133-L

20. Alaoui-Ismaïli O, Robin O, Rada H, Dittmar A, Vernet-Maury E. Basic emotions evoked by odorants: comparison between autonomic responses and self-evaluation. *Physiol Behav*. (1997) 62:713–20. doi: 10.1016/S0031-9384(97)90016-0

21. Krusemark EA, Novak LR, Gitelman DR, Li W. When the sense of smell meets emotion: anxiety-state-dependent olfactory processing and neural circuitry adaptation. *J Neurosci*. (2013) 33:15324–32. doi: 10.1523/JNEUROSCI.1835-13.2013

22. Schaal B, Saxton TK, Loos H, Soussignan R, Durand K. Olfaction scaffolds the developing human from neonate to adolescent and beyond. *Philos Trans R Soc B Biol Sci*. (2020) 375:20190261. doi: 10.1098/rstb.2019.0261

23. Soussignan R. Olfaction, réactivité hédonique et expressivité faciale chez l'enfant [olfaction, hedonic responsivity and facial expresssivity in children]. *Enfance*. (1997) 50:65–83. doi: 10.3406/enfan.1997.3047

24. Soussignan R, Schaal B. Les systèmes émotionnels chez le nouveau-né humain: Invariance et malléabilité des réponses aux odeurs [Emotional systems in the human neonate: Invariance and malleability of odor responses]. *Enfance*. (2001) 53:236–46. doi: 10.3917/enf.533.0236

25. Flohr ELR, Erwin E, Croy I, Hummel T. Sad man's nose: emotion induction and olfactory perception. *Emotion*. (2017) 17:369–78. doi: 10.1037/emo0000224

26. Beedie C, Terry P, Lane A. Distinctions between emotion and mood. *Cogn Emot*. (2005) 19:847–78. doi: 10.1080/02699930541000057

27. Kontaris I, East BS, Wilson DA. Behavioral and neurobiological convergence of odor, mood and emotion: a review. *Front Behav Neurosci*. (2020) 14:35. doi: 10.3389/fnbeh.2020.00035

28. Perifano A, Scelles R. Regards croisés Sur la souffrance psychique des enfants polyhandicapés atteints de maladies évolutives [psychological distress of children with progressive diseases and multiple disabilities: a crossed analysis]. *Arch Pediatr*. (2015) 22:916–23. doi: 10.1016/j.arcped.2015.06.009

29. Gabbaï P. Les troubles psychiatriques de la personne polyhandicapée [psychiatric disorders in persons with profound intellectual and multiple disabilities] In: P Camberlein and G Ponsot, editors. *La personne polyhandicapée: La connaître, l'accompagner, la soigner [people with profound intellectual and multiple disabilities: Knowing them, accompanying them, caring for them]*. Paris: Dunod (2021)

30. Poppes P, van der Putten A, Vlaskamp C. Frequency and severity of challenging behaviour in people with profound intellectual and multiple disabilities. *Res Dev Disabil*. (2010) 31:1269–75. doi: 10.1016/j.ridd.2010.07.017

31. Vos P, De Cock P, Petry K, Van Den Noortgate W, Maes B. Do you know what I feel? A first step towards a physiological measure of the subjective well-being of persons with profound intellectual and multiple disabilities. *J Appl Res Intellect Disabil*. (2010) 33:366–78. doi: 10.1111/j.1468-3148.2010.00553.x

32. Maes B, Nijs S, Vandesande S, van keer I, Arthur-Kelly M, Dind J, et al. Looking back, looking forward: methodological challenges and future directions in research on persons with profound intellectual and multiple disabilities. *J Appl Res Intellect Disabil*. (2021) 34:250–62. doi: 10.1111/jar.12803

33. Petitpierre G, Dind J, De Blasio C, Gremaud G. Odour detection in children and young people with profound intellectual and multiple disabilities. *J Appl Res Intellect Disabil*. (2022) 35:519–30. doi: 10.1111/jar.12963

34. Petitpierre G, Dind J. Measuring olfaction in children and young people with profound intellectual and multiple disabilities In: M Bensafi, editor. *Basic protocols on emotions, senses, and foods*. New York: Springer (2023).

35. Bojanowski V, Hummel T. Retronasal perception of odors. *Physiol Behav*. (2012) 107:484–7. doi: 10.1016/j.physbeh.2012.03.001

36. Hummel T, Livermore A. Intranasal chemosensory function of the trigeminal nerve and aspects of its relation to olfaction. *Int Arch Occup Environ Health*. (2002) 75:305–13. doi: 10.1007/s00420-002-0315-7

37. Tristão RM, Lauand L, Costa KSF, Brant LA, Fernandes GM, Costa KN, et al. Olfactory sensory and perceptual evaluation in newborn infants: a systematic review. *Dev Psychobiol*. (2021) 63:e22201. doi: 10.1002/dev.22201

38. Petry K, Maes B. Identifying expressions of pleasure and displeasure by persons with profound and multiple disabilities. *Intellect Dev Disabil*. (2006) 31:28–38. doi: 10.1080/13668250500488678

39. Green CW, Reid DH. Defining, validating, and increasing indices of happiness among people with profound multiple disabilities. *J Appl Behav Anal*. (1996) 29:67–78. doi: 10.1901/jaba.1996.29-67

40. Thompson T, Felce D, Symons F. *Behavioral observation: Technology and applications in developmental disabilities*. Baltimore: Paul H. Brookes (2000).

41. Schaal B, Soussignan R, Marlier L. Olfactory cognition at the start of life: the perinatal shaping of selective odor responsiveness In: C Rouby, B Schaal, D Dubois, R Gervais and A Holley, editors. *Olfaction, taste, and cognition*. Cambridge: Cambridge University Press (2002)

42. Legerstee M, Anderson D, Schaffer A. Five- and eight-month-old infants recognize their faces and voices as familiar and social stimuli. *Child Dev*. (1998) 69:37–50. doi: 10.1111/j.1467-8624.1998.tb06131.x

43. Marsch H, Stavropoulos J, Nienhuis T, Legerstee M. Six- and 9-month-old infants discriminate between goals despite similar action patterns. *Infancy*. (2010) 15:94–106. doi: 10.1111/j.1532-7078.2009.00002.x

44. Rochat P, Striano T. Who's in the mirror? Self-other discrimination in specular images by four- and nine-month-old infants. *Child Dev*. (2002) 73:35–46. doi: 10.1111/1467-8624.00390

45. Holle H, Rein R. EasyDIAg: a tool for easy determination of interrater agreement. *Behav Res Methods*. (2015) 47:837–47. doi: 10.3758/s13428-014-0506-7

46. Flynn S, Vereenooghe L, Hastings R, Adams D, Cooper SA, Gore N, et al. Measurement tools for mental health problems and mental well-being in people with severe or profound intellectual disabilities: a systematic review. *Clin Psychol Rev*. (2017) 57:32–44. doi: 10.1016/j.cpr.2017.08.006

47. Berg K, Arron K, Burbidge C, Moss J, Oliver C. Carer-reported contemporary health problems in people with severe and profound intellectual disability and genetic syndromes. *J Policy Pract Intellect Disabil*. (2007) 2007:120–8. doi: 10.1111/j.1741-1130.2007.00109.x

48. Ross E, Oliver C. Preliminary analysis of the psychometric properties of the mood, interest and pleasure questionnaire (MIPQ) for adults with severe and profound learning disabilities. *Br J Clin Psychol*. (2003) 42:81–93. doi: 10.1348/014466503762842039

49. American Psychiatric Association. *Diagnostic and statistical manual of mental disorders*. *4th* ed. Washington: APA (1994).

50. Oliver C, Arron K, Burbidge C, Hayley M, Moss J, Ross E, et al. *Manual for the repetitive behaviour questionnaire (RBQ), Activity questionnaire (TAQ), mood interest and pleasure questionnaire (MIPQ) and food related problems questionnaire (FRPQ)*. Birmingham: University of Birmingham (2010).

51. Verbel Sierra P. (2013). Validation psychométrique du questionnaire humeur, intérêt et plaisir (QHIP), version francophone du Mood, Interest and Pleasure Questionnaire (MIPQ) [Psychometric validation of the Mood, Interest and Pleasure Questionnaire (MIPQ), French version]. Geneva: Unpublished Master thesis. University of Geneva.

52. Pallant J. *SPSS Survival Manual*. *7th* ed. London: Routledge (2020).

53. Scherer K. Appraisal considered as a process of multilevel sequential checking In: KR Scherer, A Schorr and T Johnstone, editors. *Appraisal processes in emotion: Theory, methods, research*. New York: Oxford University Press (2001).

54. Vos P, De Cock P, Petry K, Van Den Noortgate W, Maes B. Investigating the relationship between observed mood and emotions in people with severe and profound intellectual disabilities. *J Intellect Disabil Res*. (2013) 57:440–51. doi: 10.1111/jir.12021

55. Clark LA, Watson D. Tripartite model of anxiety and depression: psychometric evidence and taxonomic implications. *J Abnorm Psychol*. (1991) 100:316–36. doi: 10.1037/0021-843X.100.3.316

56. Browning M, Holmes EA, Harmer CJ. The modification of attentional bias to emotional information: a review of the techniques, mechanisms, and relevance to emotional disorders. *Cogn Affect Behav Neurosci*. (2010) 10:8–20. doi: 10.3758/CABN.10.1.8

57. Rottenberg J, Gross JJ, Gotlib IH. Emotion context insensitivity in major depressive disorder. *J Abnorm Psychol*. (2005) 114:627–39. doi: 10.1037/0021-843X.114.4.627

58. Bylsma L. Emotion context insensitivity in depression: toward an integrated and contextualized approach. *Psychophysiology*. (2021) 58:e13715. doi: 10.1111/psyp.13715

59. Licon CC, Manesse C, Dantec M, Fournel A, Bensafi M. Pleasantness and trigeminal sensations as salient dimensions in organizing the semantic and physiological spaces of odors. *Sci Rep*. (2018) 8:8444. doi: 10.1038/s41598-018-26510-5

60. Schriever VA, Gellrich J, von der Hagen M, Hummel T. Acquired olfactory dysfunction in children and adolescents: a systematic review of the literature. *Chem Senses*. (2018) 43:571–81. doi: 10.1093/chemse/bjy046

61. Booth TN, Rollins NK. Spectrum of clinical and associated MR imaging findings in children with olfactory anomalies. *Am J Neuroradiol*. (2016) 37:1541–8. doi: 10.3174/ajnr.A4738

62. Espinosa-Jovel C, Toledano R, Jiménez-Huete A, Aledo-Serrano Á, García-Morales I, Campo P, et al. Olfactory function in focal epilepsies: understanding mesial temporal lobe epilepsy beyond the hippocampus. *Epilepsia Open*. (2019) 4:487–92. doi: 10.1002/epi4.12343

63. Dubrovskaya NM, Vasilev DS, Tumanova NL, Alekseeva OS, Nalivaeva NN. Prenatal hypoxia impairs olfactory function in postnatal ontogeny in rats. *Neurosci Behav Physiol*. (2022) 52:262–70. doi: 10.1007/s11055-022-01233-3

64. Shivkumar M, Milho R, May JS, Nicoll MP, Efstathiou S, Stevenson PG. Herpes simplex virus 1 targets the murine olfactory neuroepithelium for host entry. *J Virol*. (2013) 87:10477–88. doi: 10.1128/JVI.01748-13

65. Cole P, Jacobs A. From children's expressive control to emotion regulation: looking back, looking ahead. *Eur J Dev Psychol*. (2018) 15:658–77. doi: 10.1080/17405629.2018.1438888

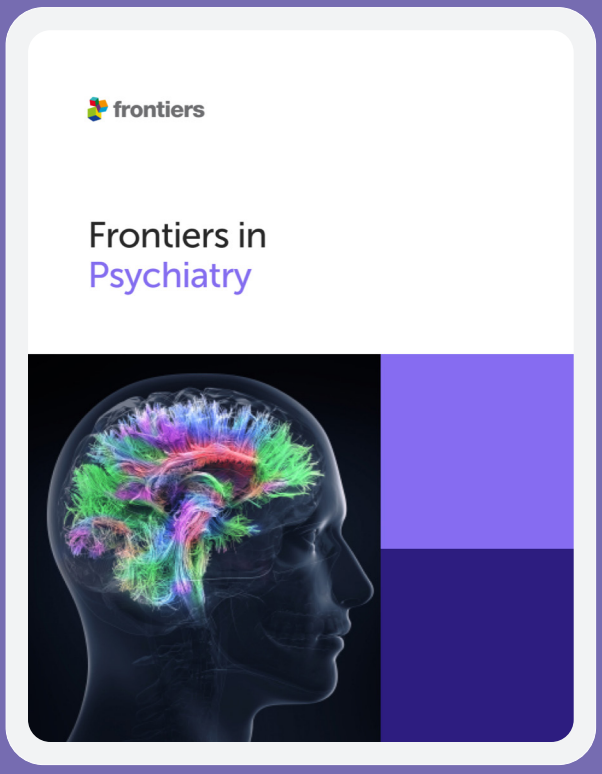
Frontiers in Psychiatry

Explores and communicates innovation in the field of psychiatry to improve patient outcomes

The third most-cited journal in its field, using translational approaches to improve therapeutic options for mental illness, communicate progress to clinicians and researchers, and consequently to improve patient treatment outcomes.

Discover the latest Research Topics

See more →



Frontiers

Avenue du Tribunal-Fédéral 34
1005 Lausanne, Switzerland
frontiersin.org

Contact us

+41 (0)21 510 17 00
frontiersin.org/about/contact