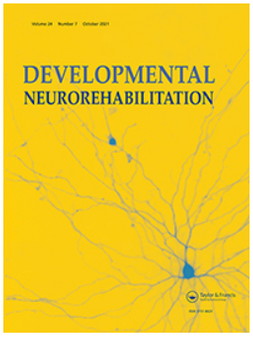

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Improvement of Planning Abilities in Adults with Prader-Willi Syndrome: A Randomized Controlled Trial

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ABSTRACT

Prader-Willi Syndrome (PWS) is a neurodevelopmental genetic disorder with executive deficits. Planning is one of the impaired executive functions implied in the regulation of behavior and everyday actions. We aimed to explore the feasibility and the effectiveness of a metacognitive strategy training designed to improve planning in adults with PWS using a double-blind between-group (training versus usual care) randomized controlled trial, with computerized tests and paper-pencil ecological outcome measures targeting planning, other executive functions, and achievement of personalized goal. Results showed better performances in several executive tasks and in achievement of personalized goals after both interventions, but better improvement for the experimental group (n = 27) compared to control (n = 26) only on the task assessing planning abilities. Interviews with occupational therapists demonstrated the feasibility of this training with this population. Despite a small number of sessions, the metacognitive strategy training showed encouraging results on planning abilities of patients.

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Cognitive rehabilitation; executive functions; intellectual disability; metacognitive strategy training; planning; prader-willi syndrome

Introduction

Prader-Willi Syndrome is a neurodevelopmental genetic disorder characterized by various expressions of endocrine, neurologic, cognitive and behavioral symptoms.[1] The disorder is caused by the loss of expression of the imprinted genes from the 15q11q13 region of the paternal chromosome 15.[2] Approximately 60% of cases are due to the deletion of the whole 15q11q13 region (type I deletion) or a part of it (type II deletion) and in 35% of cases, the entire 15th maternal chromosome is duplicated and the paternal chromosome is lost (uniparental maternal disomy).[3] PWS is characterized by infantile hypotonia, mental retardation, feeding difficulty in infancy that evolves to an extreme drive to eat in childhood, dysmorphic features, short stature, hypogonadism, sleep apnea, diabetes, and severe maladaptive behaviors including obsessive, compulsive, and oppositional behaviors.[4,5] Speech and language skills are also reported to be often impaired.[6] Intellectual disability and deficit in executive functions are well documented in PWS: deficits in inhibition, switching, updating, cognition estimation, planning[7–13] Executive functions (EF) are essential to allow a flexible and context-appropriate behavior when facing a new or complex situation.[14,15] Planning is conceived as a higher cognitive function that implies the effective inhibition, updating and shifting processes.[16]

Patients' daily life is impacted by executive deficits, and they have difficulties in managing social, professional, and familial aspects of their life. A key EF is planning which can be defined as the ability to anticipate and to organize a series of actions in an optimal sequence to achieve a goal.[17] Planning impairments have been found across various tests in PWS patients. Chevalère et al.[7] administered the Zoo Map from the Behavioral Assessment of the Dysexecutive Syndrome (BADS)[18] to 20 individuals with PWS to examine the ability to plan a route on the basis of several rules. Results showed poor planning abilities, moderately related to the indices of intelligence quotient (verbal IQ, performance IQ, full-scale IQ). In 2015, Chevalère and collaborators[8] assessed planning abilities with the Commission Test[19] and the Tower of London[20] revealing that PWS individuals performed worse than age-matched healthy controls. Estival et al.[21] compared 30 adults with PWS to two groups on an adapted version of the Zoo Map task. Results showed that PWS adults had difficulties in formulating and correctly achieving goals, due to deficits in planning. They also had difficulties in using effective strategies and in anticipating the steps needed for the task. In summary, planning difficulties in PW patients result in a failure to anticipate, to follow a series of steps, and to adjust behavior to unexpected events, resulting in a disorganized behavior. Translated into daily life challenges, difficulties in planning are particularly disabling for actions like being able to plan an appointment and be there on time, or being able to take the bus independently and adapt if an unexpected event changes the bus course.

Cognitive rehabilitation and particularly EF interventions are intended to improve the ability of individuals to participate in meaningful activities and to be functionally independent.[22] In current clinical practice, different approaches are used to help individuals with their EF difficulties: (1) providing environmental support and compensatory aids, (2) training component of EF, (3) training individuals on specific goals, (4) providing metacognitive strategies applicable to a variety of everyday situations.[23] Metacognitive strategy training is one of

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the most studied approaches to help patients with their executive difficulties.[24] Metacognitive strategies rely on a step-by-step approach to help individuals to better manage their behavior. Metacognitive programs often include self-awareness, self-monitoring, and self-control of cognition while performing an activity.[24] A well-known metacognitive approach is Goal Management Training (GMT). GMT is a metacognitive strategy training program which helps individuals to efficiently encode goals and sub-goals in order to achieve a task by learning a mental checklist routine.[25] GMT is based on Duncan et al.'s theory of "goal neglect"[26] which suggests that impairments in executive tasks are due to a poor construction and use of the "goal lists," necessary for goal-related behavior. In adults with acquired brain injury, metacognitive strategies are effective and recommended as standard practice.[27] However, in PWS, no research exploring metacognitive strategies have been done so far, revealing the lack of validated methods for EF rehabilitation in patients with intellectual disability.

The aim of this study was to assess the feasibility and effectiveness of a metacognitive strategy training on daily life planning difficulties in adults with PWS. Regarding feasibility, our purpose was to evaluate if adults with PWS were able to go through a new challenging training in group with occupational therapists (OTs). We also wanted to evaluate the feasibility of using Goal Attainment Scaling (GAS) methodology for the assessment of goal achievement related to planning difficulties in adults with PWS considering their intellectual disability. Regarding effectiveness, we hypothesized that individuals receiving the training would experience greater improvements in planning abilities compared with controls receiving care-as-usual. We also hypothesized that the training of a high-level EF such as planning can be generalized to other EFs (inhibition, updating and shifting) as planning is conceived as linked to these processes.

Materials and Methods

Study Design

The study used a double-blinded between-group randomized controlled trial design comparing performance pre- and post-intervention of patients undergoing the metacognitive strategy training to a control group receiving usual care. Both groups received occupational therapy, within a multidisciplinary rehabilitation program, in which only the content of occupational therapy session varied targeting either planning in the experimental group or other nonspecific goals excluding planning ability in the control group. The study design is detailed in a protocol paper published in the same journal.[28] During 17 months, participants were allocated to either the metacognitive strategy training (experimental group) or usual care (control group). Each month, the content of occupational therapy session (training or usual care) was randomly chosen by a researcher who was not involved in the assessment and intervention sessions. Participants were blind to the potential superiority of one of the interventions. The investigator conducting the assessment and patients' caregivers were blind to group assignment. The investigator conducting the assessment was a neuropsychologist trained in the administration of the assessment instruments. Prior to the inclusion of the first participant in the trial, the protocol was implemented in the rehabilitation center over one month with a test group consisting of four patients with PWS. Following the test group, the content of the intervention was adjusted and put into a manual.

Participants

All the participants were recruited from the PWS unit of the Hendaye (France) rehabilitation center belonging to the French Reference Center for PWS. Admissions to this unit are usually requested by either the patients or their caregivers for a period of one to two months. The purpose of the stay is to assess psychosocial and medical problems in order to define individual needs and to propose a personalized management strategy during their stay. To this purpose, the center employs a multidisciplinary approach to the syndrome. In general, for patients with PWS, the challenges are weight control, improvement of physical condition, medical care of complications (e.g., diabetes), promotion of psychological well-being and social life adaptation.[29] Furthermore, the stay allows a break from the family or from everyday residential routines. The admissions are never in response to an acute clinical situation. The study was approved by the French Reference Center board and conducted in accordance with the Declaration of Helsinki. Patients gave written informed consent prior to study enrollment. Only patients with genetically confirmed diagnosis were included. Exclusion criteria were: (1) inability to speak/understand French language; (2) severe psychiatric problems or severe mood symptoms; (3) IQ level under 50.

Procedure

Identifying planning difficulties in participants' ecological context

Prior to the intervention, participants' primary caregiver filled in two questionnaires: the Dysexecutive Questionnaire (DEX) from the BADS[18] and the 6-Item version of the Disability Assessment for Dementia (DAD-6).[30] The DEX questionnaire is a 20-item checklist in which cognitive, behavioral, and emotional aspects of executive difficulties are assessed. Each item is scored on a five-point Likert scale ranging from 0 (never) to 4 (very often) with a maximum score of 80. A higher score indicates a greater severity of executive functioning problems. The DAD-6 questionnaire is an informant-based questionnaire assessing six instrumental activities. Each activity includes three questions corresponding to three executive factors: initiation, organization-planning, realization (i.e., effective performance). Each response "yes" or "no" is scored 1 point and the maximum score is 18 (corresponding to 18 answers "yes"). Even if the DAD-6 questionnaire was originally designed to assess early dementia, this questionnaire highlights the EF required to perform the instrumental activities of daily life and allowed us to evaluate participants' executive difficulties in ecological context. These questionnaires provided a basis for a phone-interview with caregivers regarding planning difficulties of participants and potential personalized goals relating to planning. Principal rehabilitation goals given by parents were

extracted. A face-to-face interview of the participants was then conducted by an OT using the report of the phone-interview with caregivers to lead the discussion. Rehabilitation goals related to planning were selected based on participants' interest. Comparison of those two interviews (caregivers versus participants) provided information about perceived difficulties and intervention priorities.

Intervention

Two intervention conditions were employed: (1) metacognitive strategy training of planning abilities with the ETAPP (Evaluation of a Therapeutic Aid of the Planning function in Prader-Willi Syndrome) program; (2) usual care by the same OTs, not focusing specifically on planning. Each intervention (training or usual care) was delivered by two trained OTs who usually work with PW patients in this center (6 to 12 years of experience).

Metacognitive strategy training. The ETAPP program is a composite metacognitive strategy training method designed to address planning difficulties in adults with PWS. This program is based on the GMT,[25] as well as Attention and Problem Solving[31] with the use of a clarified schema describing the several steps of the metacognitive strategy. Principles of Ylvisaker's self-regulation scripts were added to facilitate the development of self-regulation: for example, deciding before the execution if the target task is hard/easy, if the participant is ready/not ready, if the task is a big deal/little deal.[32] Finally, "problem-orientation" notions were added to highlight participants' reactions when confronted to the target task: for example, asking about participants' beliefs, assumptions and expectations concerning the task and its execution.[33]

The training was conducted in a small group format (three or four participants) with the two OTs. Each session began with OTs introducing the step to work on during the session. Explanations and examples were provided to participants then OTs encouraged them to discuss about the step. Then, the new step was implemented in the task chosen by the group. Group intervention was chosen over individual intervention to encourage interactions between the participants but the fact of having two OTs for a group of maximum four people also made it possible to notice specific difficulties of the participants during the realization of the task. OTs provided guidance, but allowed participants to experience planning errors on-task, in order to promote awareness and adjustment to experienced difficulties. For this purpose, OTs would let patients think things through until they saw a problem or asked question to highlight the problems.

The ETAPP program comprised six 1-h group-sessions which aimed at learning and applying the strategy "Pause – Define the task – List – Do it – Evaluate" in ecological and relevant tasks chosen by the group. The first session focused on increasing participants' awareness of planning difficulties in daily life. Discussion in this first session was triggered through illustrated stories presented to participants by the OTs, where characters were experiencing difficulties in daily life because of inadequate planning. Stories were chosen to echo the situations experienced by the participants: "I had to pick up my sister from the station. I had about 10 minutes before I left and so I turned on the TV. It was one of my favorite movies and it was pretty close to the end. It was only about 25 minutes later that I checked the time. Watching the TV, I forgot what I needed to do. The consequence was that my sister was worried about not knowing where I was and when she found out why I was late, she felt sad." In the following sessions (2–5) the group performed a task they choose (e.g., set alarms on the phone to avoid missing appointments, organize a bowling day out) following the strategy "Pause – Define the task – List – Do it – Evaluate" with guidance from OTs. The aim of the first step named "Pause" was to define the main objective of the task: what am I doing? What am I supposed to do? What is the main goal of my task? The second step was about defining precisely the task: Is this going to be hard or easy? How much time will this take? What materials will I need? What difficulties may I experience? What are my strengths and weaknesses? Can I ask for help? In the third step, the group listed the steps and established a plan, taking into account potential problems or obstacles. The next step "Do it" focused on execution while monitoring the steps: are there mistakes in the steps? Do I follow the steps correctly? In the final session, participants evaluated if they had followed all the steps as planned and evaluated their overall performance on the task: what worked? What did not work?

Control intervention. Usual care for the control group was individually determined by the same OTs and included intervention about dressing, morning activities like washing or getting up after a fall. Interventions in usual care insisted on motor training rather than action planning.

Measures

Measures included monitoring of procedural fidelity of the training, neuropsychological assessment, questionnaires, GAS and ecological repeated measures of both experimental and control groups.

Monitoring of Procedural Fidelity and Feasibility Assessment

To ensure that the training was implemented in the same throughout the study, an independent evaluator assessed one of the six session in each group, chosen at random, on a procedural fidelity check list to ensure that the protocol was implemented in the same way throughout the study.[34,35] The independent evaluator provided feedback to the OTs, as needed, to maximize adherence to the program. Scores were averaged for each item at the end of the study to verify if the intervention was delivered in the same way over the 17 months of the study. Details of rating can be found in Appendix A.

Interventions were regularly followed by unstructured interviews with the OTs to collect qualitative information about the feasibility of the program, participants' motivation and satisfaction of OTs regarding elements of rehabilitation included in the intervention. Unstructured interviews allowed us to collect OTs' point of view in a non-formal way to deepen some answers and assess the overall feasibility of implementing such a challenging program in a PWS unit. Additional items were included in the fidelity checklist to assess the feasibility of the program.

Neuropsychological Assessment

Neuropsychological assessment was conducted in pre-intervention (T0) and post-intervention (T1) sessions by an investigator blinded to group allocation. The pre-intervention assessment was distributed in two 1-h sessions over two successive days. The post-intervention assessment was a 1-h session and was conducted one to two days after the last intervention session.

The main outcome was the performance on the Modified Six Elements task (MSET) from the BADS[18] to measure planning abilities. The BADS was originally developed to predict everyday problems arising from the dysexecutive syndrome with ecological tasks.[36] Those tasks were designed to be analogous to those required in everyday life activities involving executive functioning. The MSET emphasizes on a person's ability to plan, organize and monitor behavior[37] and requires a considerable amount of planning and monitoring behavior because of its highly unstructured character.[38] The task consisted of three open-ended tasks (dictation, written picture naming, and simple arithmetic) divided into two parts (subtasks A and B). Participants were instructed to execute at least a part of each of these six subtasks within 10 minutes. However, it was not allowed to switch between two subtasks of the same type (e.g., after completing the first subtask of the picture naming task, participants had to switch to dictation or arithmetic). An unusual aspect of the MSET is that the subject's performance in each task is not important. The measures were the number of executed subtasks within the 10 minutes as well as the amount of time spent on each subtask and the number of time where the rule of alternation between the subtasks was broken (rule breaks). The effectiveness of planning process was evaluated by a raw score, obtained by subtracting the rule breaks from the number of executed tasks (range 0–6). Each rule break (e.g., switching from subtask A of the arithmetic task directly to subtask B of arithmetic) was considered as a cue of planning difficulty. In agreement with the manual of the BADS, this score was converted into profile scores (range 0–4). An adapted scoring method was also calculated to estimate time distribution over the subtasks and avoid ceiling effects.[38] This adapted scoring takes into account the distribution of time spent on the six subtasks by subtracting the shortest time spent on one of the six subtasks (i.e., the total time spent on the subtask) from the longest time spent on one of the six subtasks (i.e., the total time spent on the subtask). As a more homogeneous distribution of time between the tasks indicates better planning abilities, a lower score indicates better planning performance. This adapted scoring is thought to have a better sensitivity compared with the poor sensitivity of the traditional score.[38] The MSET have been proved to be a reliable and valid measure for French population of individuals with PWS.[7]

As secondary outcome measures, executive functioning was assessed using a battery of computerized neuropsychological tests previously used and validated for people with PWS.[39] These tests were chosen based on reduced test–retest effects and sensibility to executive dysfunction in patients with PWS.[39]

Updating was measured with the N-Back task adapted from Li et al.[40] The aim of the task was to constantly recall the previous position of a stimulus evolving in a grid of 16 boxes. The participant had to press the "enter" key on the keyboard if the current position stimulus is the same as two trials before ("Match" trials). He had to not press any key if the position of the stimulus is different ("No Match" trials). Measures were the reaction time (RT) i.e., the time measured between the moment of appearance of the stimulus and the participant's response, the percentage of errors and the type of errors (omission when participant failed to press the key when he should have or false-alarm when he pressed the key when he should not have). In this task, the more efficient the update process, the faster the RTs and the lower the number of errors for "Match" and "No-Match" trials. The N-Back task has been proven to be a reliable and valid measure for French population of individuals with PWS.[39]

Shifting was measured with the Voluntary Task Switching[41] which measures shifting efficiency when it is controlled by the participant himself. The participant saw a point that changed position constantly in a grid of four boxes and had to perform two tasks randomly: define the point position either as left or right (task A: horizontal position) or as up or down (task B: vertical position). Measures were the RT, the percentage of errors, and the percentage of the type of task achieved. In this task, switching between two tasks results in longer RTs and a higher number of errors than repeating the same two tasks. This difference is called "switching cost" which corresponds to alternated trials (when the participant has switched between vertical and horizontal position) minus repeated trials (when the participant has repeated the same position). This task allowed also to measure the proportion of repeated trials versus alternated trials. A "repetition bias" is usually observed, which corresponds to a higher percentage of repeated trials than alternated trials. The Voluntary Task Switching have been proved to be a reliable and valid measure for French population of individuals with PWS.[8]

Inhibition was measured with the Hayling task, which measures the capacity to inhibit verbal automatic response.[42] In this study, we used an adapted version of this task.[43] The task consisted of two blocks: initiation and suppression. The participant had to read a sentence in French where the last word is missing. In the initiation block, the participant had to choose quickly which of the two words proposed on the screen completes correctly the sentence. In the suppression block, the participant had to choose the word that does not complete correctly the sentence. Two measures were collected: the RT and the percentage of errors. The Hayling task has been proven to be a reliable and valid measure for French population of adults and elderly patients.[43]

Questionnaires

We measured self-awareness of executive difficulties of participants with the self-administered version of the DEX questionnaire in pre-(T0) and post-intervention (T1) sessions which gave indication of the participants' insight of difficulties. The DEX and DAD-6 questionnaires were also filled in again by caregivers for a follow-up 6 months after the end of the training. Since participants were thought to rate themselves differently than their caregivers, the total scores in T0 and T1 were compared between raters.[18]

Goal Attainment Scaling

Following the face-to-face interview lead by the OT, personalized goals regarding planning abilities were selected by the participant and the OT and transformed into Goal Attainment Scales (GAS) using Kiresuk & Sherman original method[44] but adapted to follow recently published guidelines.[45] GAS is a method for writing personalized evaluation scales to quantify progress toward goals relevant for the participant. GAS was used in this study to assess functional and participation changes after the intervention and was rated during an individual session by (1) the OT that conducted the intervention; (2) the participant. In the final session, the participant and the OT assessed the participant's performance with the GAS scale via self-report by providing evidence for their GAS rating. Success of the intervention was quantified on a 5-point ordinal scale, ranging from −2 to +2. The middle point "0" corresponded to a goal attained as expected, while +2 was to a goal attained much better than expected. Example of a full GAS scale can be found in Table 1. In the follow-up 6 months later, caregivers gave subjective feedback on the attainment of the general GAS themes identified using four categories: a lot better, a little better, identical, worse, and cannot answer. This method was not the usual GAS method as for the participant and the OT to simplify the collection of caregivers' data.

Ecological Repeated Measures

Several studies have shown that the relationship between executive tasks and measures of everyday functioning are moderate to weak.[46,47] Because evidence regarding the correlation of the MSET with everyday executive functioning is controversial,[38,48] we also used two other ecological measures of planning in daily life: (1) between-session assignments; (2) a punctuality score. Between-session assignments consisted of ecological tasks involving planning (e.g., "which bus line should you take and what time to be at the train station at 12 h tomorrow?") and required participants to search for information. The list of between-session assignments can be found in table. All the tasks involved initiation (no reminder provided), planning and flexibility if information could not be found as planned. The difficulty of the tasks varied but their order was randomized to overcome potential effect due to unequal difficulty. A punctuality score was used to assess the ability to come on time to the different rehabilitation sessions. This ability was thought to be linked to the ability to anticipate and plan ahead which was expected to improve in the experimental group.

Feasibility of the ecological repeated measures. We evaluated the feasibility of the assignments with the percentage of success of each assignment. This percentage was compared between the experimental group and the control group. The feasibility of obtaining a punctuality score was assessed with the number of actually collected data and the subjective feedback of rehabilitation staff.

Monitoring of progress on ecological repeated measures. Each between-session assignment was scored as succeeded, not attempted or attempted but failed. The progress of each group was monitored by their percentage of success of between-session assignments through the study. The punctuality score took into account the number of times the participant arrived on time and/or participant's delay each week to provide a weekly punctuality score. The progress of each group was followed by a punctuality percentage calculated by assigning points to the number of times the participant arrived on time out of the number of activities actually scheduled.

Statistical Analysis

For statistical analysis, IBM SPSS version 20 (SPSS Inc, Chicago, IL, USA) was used with alpha set at 0.05 for all analysis. To compare the demographic characteristics between the groups, *t*-tests were conducted. Multivariate analyses of variance were performed to compare the performance on the MSET and EF tasks between the two groups (experimental and control) in pre- and post-intervention sessions. Because data loss due to missing values was undesirable, all test-measures were analyzed separately. To control for possible effects of demographic differences, multivariate analyses of covariance were performed when applicable. Regarding GAS scales, five levels were written. Each GAS scale resulted in a raw score with the possible values −2, −1, 0, +1, and +2 and each level was considered as a category. GAS scores were analyzed using non-parametric statistics to preserve the ordinal nature of GAS scales. The distribution of raw scores by the five levels in the two groups was analyzed visually and by chi-squared test.

Table 1. Example of a full GAS scale

Difficulties reported by caregivers: "can't remember his appointments by himself"	
Difficulties reported by the participant: "I don't remember activities and appointments; I forget to update appointments when a change happens. I can't use the calendar my mother bought me."	
Objective: Learning to use properly his calendar	
Target behaviour: Write down in calendar everyday activities with updating and out-of-ordinary events, from Monday 15th April to Sunday 4th May	
- 2	Doesn't write down in his calendar any activities and appointments
- 1	Starts to use his calendar by putting some everyday, regular activities without updating
0	Uses his calendar by putting everyday activities with updating
+1	Uses his calendar by putting everyday activities with updating and out-of-ordinary events
+2	Uses his calendar by putting everyday activities with updating and out-of-ordinary events beyond the requested date
Subjective rating of global progress toward participant identified goal, viewed by caregivers: In the objective of acquiring more independence in the management of his activities and appointments with the help of his calendar, following his stay in Hendaye in April, the situation seems to be (a lot better, a little better, identical, worse and cannot answer)	

Results

Participants

The experimental group comprised 27 adults with PWS, 16 women and 11 men. Eighteen patients had a paternal deletion, three had uniparental maternal disomy and six had other forms of genetic defect (imprinting center mutations or translocations). The control group comprised 26 adults with PWS, 16 women and 10 men. Twenty-three patients had a paternal deletion, one patient had uniparental maternal disomy and two other forms of genetic defect (translocations). Characteristics of the two groups relative to age, gender, FSIQ, VIQ and PIQ can be found in Table 2. Intergroup difference was found for age, t (51) = 2.10, p = .03 and was later controlled for in between-group comparisons. No differences were found between the groups for FSIQ, t (39) = −.23, p = .82, VIQ, t (39) = −.15, p = .88, or PIQ, t (39) = .07, p = .94. An assessment of the overall cognitive efficiency of the participants was done with the MoCA (Montreal Cognitive Assessment).[49] The MoCA is an assessment tool that allows to quickly evaluate several cognitive functions such as visuospatial skills, attention, concentration, EF, memory, language, abstraction, calculation and orientation. The maximum score is 30 points with a threshold of detection of mild cognitive impairment at 26/30. There was no difference between the groups for the MoCA score, t (51) = .81, p = .42.

Identifying Planning Difficulties in Participants' Ecological Context

The DEX questionnaire and the DAD-6 were used to highlight executive difficulties, rated by both participants and caregivers. Individual interviews with participants showed that the goals most important to them (e.g., arrange appointments at his girlfriend's care home to see her more regularly) were often different from goals proposed by their caregivers after the phone-interview (e.g., be able to manage a budget monthly).

Monitoring of Procedural Fidelity and Feasibility Assessment

Intervention proved to be feasible with low attrition rate: of the 60 participants included in the study, four withdrew after the face-to-face interview with OTs (two in each group) and three withdrew during pre-intervention assessment session (two in the experimental group and one in the control group).

Procedural fidelity checklist revealed high fidelity (75.8%) and an overall feasibility of the intervention in adults with PWS (see Appendix A). Regarding the fidelity of the intervention, scores showed that the intervention was consistent over the 17 months: content of sessions was successfully administered in the same way for all the participants (between 87.5% and 100%). A weak score on only one item showed that OTs tended to forget to recall the step "PAUSE" during the execution of the task. Feedbacks on assignments and punctuality were also consistent (between 94.4% and 100%). Regarding the feasibility of the ETAPP program, most of the items scored at 100% meaning that the procedure of the intervention was successfully implemented. Elements of the training were adapted to patients with PWS (understanding of the content, stories and the metacognitive strategy training) with adequate attitude from the OTs (encouragements and keeping on track). However, one item of the feasibility checklist highlighted participants' difficulties to keep referring to the list of steps during the execution of the task.

Unstructured interviews with OTs revealed that the ETAPP program was very well received by participants in the experimental group. OTs usually used individual session with patients in their previous work but group occupational therapy sessions in the ETAPP program helped to enhance communication between patients. OTs were satisfied with the intervention and with the procedure of identifying patients' difficulties and choosing personalized rehabilitation goals with the patient, in both experimental and control group. OTs also reported the patients' comments during the final session: patients found the intervention feasible, enjoyed working in group, and being able to choose the task to work on.

Main Outcome Measure: Performance on the MSET

As age differed significantly between the groups, multivariate analyses of covariance were conducted with group as intergroup factor, time of evaluation (pre-intervention vs. post-intervention) as intragroup factor and age as covariate. Results can be found in Table 3. An effect of the time of evaluation was found regarding the total of executed subtasks,

Table 2. Characteristics of the experimental group and the control group

	Experimental group (n = 27)		Control group (n = 26)		
		(*SD*)		(*SD*)	Difference
Mean age	36.00	(*6.63*)	31.42	(*9.06*)	< .05
Age range	24-54		19-57		
WAIS-III	n = 23		n = 19		
FSIQ	60.27	(*8.95*)	61.00	(*11.31*)	n.s
VIQ	61.59	(*11.04*)	62.10	(*10.59*)	n.s.
PIQ	62.68	(*11.91*)	62.42	(*11.47*)	n.s.
MoCA	20.44	(*4.99*)	19.42	(*4.09*)	n.s.

Note. WAIS-III = Wechsler Adult Intelligence Scale-III; FSIQ = Full Scale Intellectual Quotient; VIQ = Verbal Intellectual Quotient; PIQ = Performance Intellectual Quotient, MoCA = Montreal Cognitive Assessment; n.s. = non-significant

Table 3. Means and SD of scores the neuropsychological assessment for the experimental and the control group in T0 and T1

	Experimental group (n = 27)		**Control group** (n = 26)		**ANCOVA**		
	M T0 (*SD*)	**M T1 (*SD*)**	**M T0 (*SD*)**	**M T1 (*SD*)**	**Group**	**Time**	**Time x group**
MSET total of subtasks	4.41 (*1.39*)	5.26 (*1.05*)	4.73 (*1.31*)	5.00 (*1.38*)	n.s.	*	*
MSET rule breaks	0.85 (*0.90*)	0.85 (*1.06*)	0.88 (*0.95*)	0.65 (*0.84*)	n.s.	n.s.	n.s.
MSET raw score	3.56 (*1.62*)	4.41 (*1.69*)	3.85 (*1.28*)	4.35 (*1.35*)	n.s.	*	n.s.
MSET profile score	2.59 (*0.79*)	3.07 (*0.91*)	2.65 (*0.62*)	2.92 (*0.74*)	n.s.	*	n.s.
MSET adapted score	104.18 (*132.28*)	59.29 (*48.67*)	74.00 (*71.57*)	78.11 (*137.69*)	n.s.	n.s.	n.s.
N-Back 'match' RT	939 (*267*)	965 (*332*)	968 (*287*)	930 (*267*)	n.s.	n.s.	n.s.
N-Back 'no match' RT	1041 (*293*)	1130 (*401*)	1095 (*301*)	1036 (*248*)			
N-Back 'match' ER	9.59 (*3.51*)	9.43 (*3.91*)	9.85 (*2.95*)	8.71 (*3.66*)	n.s.	*	n.s.
N-Back 'no-match' ER	1.91 (*2.53*)	1.40 (*1.58*)	2.73 (*3.55*)	2.98 (*3.63*)			
VTS 'alternate' RT	1438 (*493*)	1442 (*549*)	1293 (*460*)	1245 (*453*)	n.s.	*	n.s.
VTS 'repeated' RT	1204 (*466*)	1207 (*413*)	1120 (*336*)	1084 (*328*)			
VTS 'alternate' ER	15.47 (*15.17*)	22.50 (*23.86*)	19.87 (*16.42*)	21.48 (*18.67*)	n.s.	n.s.	n.s.
VTS 'repeated' ER	16.29 (*16.22*)	23.12 (*22.15*)	21.58 (*17.17*)	21.33 (*17.39*)			
VTS % of 'alternate' trials	42.88 (*16.74*)	43.37 (*19.16*)	43.85 (*18.68*)	42.67 (*18.58*)	n.s.	n.s.	n.s.
VTS % of 'repeated' trials	57.12 (*16.74*)	56.63 (*19.16*)	56.15 (*18.68*)	57.33 (*18.58*)			
Hayling activation RT	2228 (*874*)	1911 (*722*)	2312 (*936*)	1881 (*653*)	n.s.	*	n.s.
Hayling inhibition RT	2475 (*1120*)	2213 (*888*)	2843 (*1202*)	2271 (*1300*)			
Hayling activation ER	3.73 (*4.29*)	1.97 (*4.45*)	4.05 (*6.73*)	2.68 (*4.32*)	n.s.	*	n.s.
Hayling inhibition ER	10.23 (*19.42*)	3.50 (*6.89*)	6.77 (*11.91*)	4.26 (*6.90*)			

Note. ANCOVA = analysis of covariance; T0 = pre-intervention; T1 = post-intervention; RT = reaction time (ms); ER = percentage of errors; VTS = Voluntary Task Switching; n.s. = not significant; *significant *p*-value
Effects of the type of trial (VTS), type of trial (N-Back task) and type of block (Hayling task) are described in the text and do not appear in the table.

F (1, 50) = 6.77, p = .012, η_p^2 = 0.11, with a greater number of executed subtasks in the post-intervention session compared to pre-intervention (5.13 vs 4.56). No effect of the group was found, F (1, 50) < 1, p = .50. An interaction effect between the group and the time of evaluation was found, F (1, 50) = 4.15, p = .04, η_p^2 = 0.07. The number of executed subtasks was higher in post-intervention session for both groups, with a better improvement for the experimental group (0.96) compared to the control group (0.16). No effects were found regarding the rule breaks, Fs < 1. An effect of the time of evaluation was found on the raw score, F (1, 50) = 10.12, p = .003, η_p^2 = 0.16, with a higher raw score in the post-intervention session compared to pre-intervention (4.37 vs 3.70). There was also an effect of the time of evaluation on the profile score, F (1, 50) = 6.38, p = .015, η_p^2 = 0.11, with a higher profile score in the post-intervention session compared to pre-intervention (2.99 vs. 2.62). No group effects were found for the raw score or the profile score, Fs < 1. And no interaction effect was found for the raw score, F (1, 50) = 2.38, p = .12, or the profile score, F (1, 50) = 1.94, p = .17 Analysis of covariance was also conducted on the adapted scores.[31] No group effect was found, F (1, 32) = 1.02, p = .32. No other effects were found Fs < 1.

To take into account specific aspects of planning in a qualitative manner, strategies of participants were observed.[50] The main strategy was to spend some time on each task until completing the six tasks. The second strategy was to do an item per task and move on to the following one, successively, until time was up. The latter strategy was used by 25.92% of participants in the experimental group and 28.84% of participants in the control group, with no significant difference regarding the time of evaluation (p = .23).

Assessment of Executive Functioning

N-Back test – updating

Analyses of covariance with group as intergroup factor, type of trial ("Match" trials vs. "No Match" trials) and time of evaluation (pre-intervention vs. post-intervention) as intragroup factors and age as covariant were conducted on the RTs. An effect of the type of trial was found, F (1, 30) = 15.77, p < .001, η_p^2 = 0.33, participants were longer for the "No-Match" trials than "Match" trials (1069 ms vs 950 ms). There was no effect of the time of evaluation, F (1, 30) = 1.04, p = .31. There was no interaction effect between the time of evaluation and the group, F (1, 30) = 2.03, p = .16, and no interaction effect between the time of evaluation and the type of trial, F (1, 30) = 1.08, p = .30. No other effects were found, Fs < 1. Regarding the percentage of errors, an effect of the type of trial was found, F (1, 43) = 17.28, p < .001, η_p^2 = 0.28, with a higher percentage of errors for "Match" trials than "No-Match" trials (9.39% vs. 2.26%). No effect of the time of evaluation was found, F (1, 43) = 1.46, p = .23. An interaction effect was found between the time of evaluation and the type of trial, F (1, 43) = 4.79, p = .03, η_p^2 = 0.10. The difference in the percentage of errors between the "Match" trials and the "No-Match" trials was higher in the pre-intervention session (7.4) compared to post-intervention (6.88). No interaction effect between the group and the type of trial was found, F (1, 43) = 2.17, p = .14. No effect of double-interaction between the group, the type of trial, and the time of evaluation were found, F (1, 43) = 2.82, p = .10. No other effects were found, Fs < 1.

Voluntary Task Switching – Shifting

Analyses of covariance with group as intergroup factor, type of trial (alternated vs. repeated) and time of evaluation (pre-intervention vs. post-intervention) as intragroup factors, and age as covariant were conducted on the RTs. No group effect was found, F (1, 44) = 1.13, p = .29. A marginal effect of the time of evaluation was found, F (1, 44) = 3.09, p = .08, η_p^2= 0.06, with a longer RT in the pre-intervention than in the post-intervention (1263 ms vs. 1245 ms). An effect of the type of trial was found, F (1, 44) = 37.51, p < .001, η_p^2= 0.45, with a longer RT for the alternated trials (1355 ms) than for the repeated trials (1154 ms). This effect corresponds to switch cost. An interaction effect was found between the time of evaluation and the type of trial, F (1, 44) = 7.48, p = .009, η_p^2= 0.14. Alternated trials were longer than repeated trials and this difference was higher in the pre-intervention session (203 ms) compared to the post-intervention session (199 ms). This switch cost was statistically significant in the pre-intervention session, F (1, 44) = 6.84, p = .007, η_p^2= 0.13, but disappeared in the post-intervention session, F (1, 45) < 1, p = .34. No other effects were found, Fs < 1. Regarding the percentage of errors, there was no effect of the type of trial, F (1, 45) = 1.61, p = .21 and no interaction effect between the time of evaluation and the group, F (1, 45) = 1.82, p = .18. No other effects were found, Fs < 1. Analysis of covariance with group as intergroup factor, type of trial (alternated vs. repeated) and time of evaluation (pre-intervention vs. post-intervention) as intragroup factors and age as covariant were conducted on the percentage of type of task achieved. A marginal effect of the type of trial was found, F (1, 45) = 3.29, p = .07, η_p^2 = 0.06, with a greater number of repetition (56.81%) than alternation (43.18%). This effect is called repetition bias. There was no other effect, Fs < 1.

Hayling Task – Inhibition

Analyses of covariance with group as intergroup factor, type of block (activation vs. suppression) and time of evaluation (pre-intervention vs. post-intervention) as intragroup factors and age as covariant were conducted on the RTs. An effect of the type of block (activation vs. suppression) was found, F (1, 49) = 9.11, p = .004, η_p^2 = 0.15, participants were faster in the activation block than in the suppression block (2083 ms vs. 2450 ms). An effect of the time of evaluation was found, F (1, 49) = 22.76, p < .001, η_p^2 = 0.31, with RTs longer in the pre-intervention session compared to post-intervention (2465 ms vs 2069 ms). No interaction effect was found between the time of evaluation and the group, F (1, 49) = 2.37, p = .13. No other effects were found, Fs < 1. Regarding the percentage of errors, an effect of the time of evaluation was found, F (1, 49) = 6.42, p = .014, η_p^2 = 0.11, with a higher percentage of errors in the pre-intervention session compared to post-intervention (6.20% vs. 3.10%). An effect of the type of block was found, F (1, 49) = 6.19, p = .016, η_p^2 = 0.11, with a higher percentage of errors in

the suppression block than in the activation block (6.19% vs. 3.11%). No other effects were found, *F*s < 1.

Questionnaires

DEX

Analysis of variance with group (experimental vs. control) and informant (participant vs. caregiver) as intergroup factors and time of evaluation (pre-intervention vs. post-intervention) as intragroup factor was conducted regarding the total score (ranging from 0 to 80). Results can be found in Table 4. An effect of the informant was found, F (1, 88) = 4.91, p = .02, η_p^2 = 0.05, with a higher score from the caregivers (30.64) compared to the participants (23.92). An interaction effect between the informant and the time of evaluation was found, F (1, 88) = 9.75, p = .002, η_p^2 = 0.10. Participants evaluated themselves better in post-intervention (25.61 vs. 22.23) whereas caregivers evaluated them worst in post-intervention (29.67 vs. 31.62). Finally, a marginal effect of double-interaction was found, F (1, 88) = 3.27, p = .07, η_p^2 = 0.03, with caregivers in the control group evaluating participants with higher scores (32.94) than in the experimental group (28.34). No other effects were found, *F*s < 1.

DAD-6

Regarding the DAD-6 questionnaire, analysis of variance with group as intergroup factor and time of evaluation (pre-intervention vs. post-intervention) as intragroup factor were conducted on the scores rated by caregivers. Considering the total number of "yes" in response to the activities proposed in the questionnaire, results showed no group effect, F (1, 36) = 2.10, p = .15 and no other effects, *F*s < 1. Results can be found in Table 5. Detailing dimensions, no difference was found between the groups, F (1, 36) = 1.78, p = .19 and no other effects (*F*s < 1) were found for the dimension "initiation" or the dimension "organization-planning". For the dimension "realization", a marginal difference between the two groups was found, F (1, 36) = 2.91, p = .09, η_p^2 = 0.07 with a higher number of "yes" for the experimental group compared to the control group (4.67 vs. 4.00). No other effects (*F*s < 1) were found.

Goal Attainment Scaling

In order to evaluate the ability of the GAS-setting team to generate valid, reliable and meaningful scales, quality of GAS scales was assessed to respect recommended guidelines.[45] Relevance, attainability and pre-intervention performance were assessed by rehabilitation center caregivers and staff, knowing the participant but not participating in the study, prior to the intervention. The overall score for those three criterions scored by the rehabilitation team during staff meeting was 2.68/3, revealing a good quality appraisal. Classification of goals types (according to the ICF), specificity, equidistance of levels, time-specificity, measurability, unidimensionality and precise definition of the context of measurement were assessed in a posteriori analysis by external judges. 41.5% of the 53 GAS got the maximum quality score of 3/3 regarding those 7 criterions. 15 GAS filled 94.44% of the criterions and 13 GAS filled between 77.77% and 88.88% of the criterions. Those scores revealed a consistency in GAS use, with adequate reliability and validity of the GAS scales. GAS quality did not differ between groups, χ^2 (1) = 0.13, p = .72. Examples of the most frequent goal types can be found in Appendix B.

GAS data were not available for six participants because they refused to score themselves on a level during the post-intervention individual interview with OT. Therefore, 98 GAS were analyzed. Distribution of post-intervention GAS scores rated by participants is shown in Figure 1a. Distribution of post-intervention GAS scores rated by participants did not differ significantly between the experimental group and the control group, χ^2 (4) = 2.12, p = .73. Most GAS scores attained the expected level of "0" in each group. Distribution of post-intervention GAS scores rated by OT is shown in Figure 1b. Distribution of post-intervention GAS scores rated by OT did not differ significantly between the experimental group and the control group, χ^2 (4) = 1.95, p = .77. GAS scores of "-2" and "-1" were more frequent than intermediate results ("0" and "+1"). Rating by participants was highly correlated to the rating by OT, r (46) = 0.80, p < .001. Distribution of follow-up GAS category rated by caregivers can be found in Figure 2. Distribution of follow-up GAS category rated by caregivers did not differ significantly between the experimental group and the control group, χ^2 (5) = 6.29, p = .27. "Identical" was the most chosen category by caregivers.

Ecological Repeated Measures

Feasibility of the Ecological Repeated Measures

A large proportion of punctuality measures was missing due to organizational issues in both groups (11.3% of 53 participants). Therefore, punctuality data could not be interpreted. Regarding between-session assignments, description of the assignments and the percentage of success of each assignment can be found in Table 6. Between-session assignments were globally succeeded by both groups; however, some assignments were more failed (i.e., percentage of success under 40%). There

Table 4. Means, SD and ranges scores of the DEX questionnaire for the experimental and the control group rated by participants and caregivers

	Experimental group		Control group	
	T0 Mean (*SD*) [min-max]	T1 Mean (*SD*) [min-max]	T0 Mean (*SD*) [min-max]	T1 Mean (*SD*) [min-max]
Participants	25.70 (*13.00*) [4-49]	22.78 (*13.37*) [2-49]	25.52 (*16.51*) [0-63]	21.68 (*15.64*) [0-61]
Caregivers	28.68 (*14.73*) [4-66]	28.00 (*15.53*) [3-58]	30.67 (*15.49*) [5-59]	35.24 (*15.95*) [13-67]

Note. A higher score indicates a greater severity of executive functioning problems.

Table 5. Mean (and SD) of the DAD-6 questionnaire for the experimental and the control group rated by caregivers

	Experimental group (n = 16)		Control group (n = 19)	
	T0 M (*SD*)	T1 M (*SD*)	T0 M (*SD*)	T1 M (*SD*)
Initiation	4.56 (*0.96*)	4.28 (*1.32*)	3.80 (*1.36*)	4.05 (*1.60*)
Organisation-planning	3.72 (*1.01*)	3.89 (*1.13*)	3.50 (*1.23*)	3.35 (*1.69*)
Realisation	4.61 (*1.14*)	4.72 (*1.01*)	4.20 (*1.39*)	3.80 (*1.67*)
Total 'yes'	12.89 (*2.82*)	12.89 (*3.06*)	11.50 (3.54)	11.20 (*4.46*)

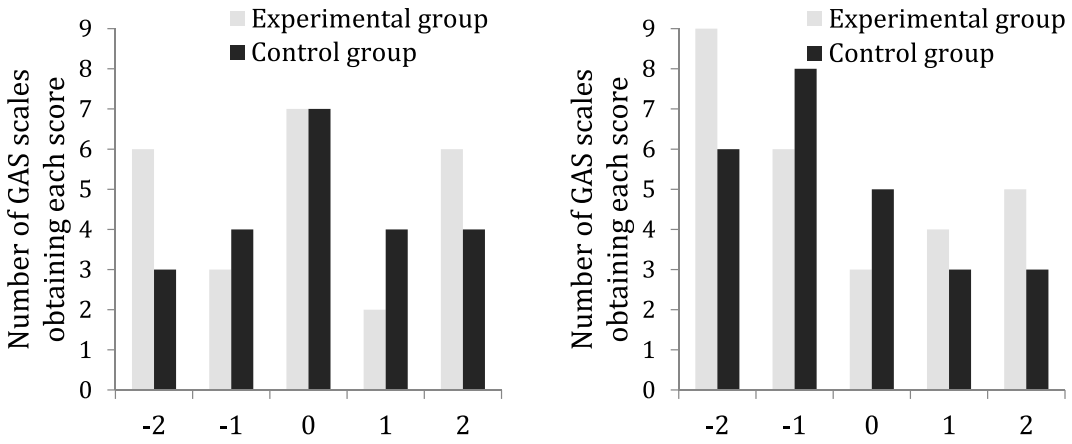

Figure 1. 1a) Distribution of all post-intervention GAS scores rated by participants; 1b) Distribution of all post-intervention GAS scores rated by OT.

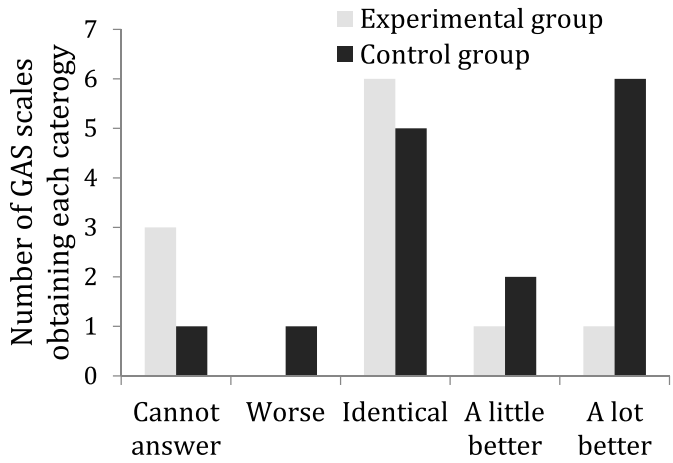

Figure 2. Distribution of all post-intervention GAS categories rated by caregivers.

was a group difference only for the assignment "Bring back a paper that interested you", t (51) = 2.62, p = .02, with the experimental group who performed this assignment better than the control group, independently of when (before the intervention start or during the intervention) this task was performed.

Monitoring of Progress on Ecological Repeated Measures

The aim was to evaluate if the experimental group performed better in performing the assignments throughout the study compared to the control group, regardless of the assignment. The experimental group seemed to perform better (see Figure 3) however t-tests showed that there is no significant difference between the two groups on none of the assignment (p > .05).

Discussion

The aim of this study was to assess the feasibility and effectiveness of a metacognitive strategy training focusing on planning difficulties in adults with PWS. ETAPP intervention proved to be feasible and very well received by the participants. On the main outcome measure, that is, planning

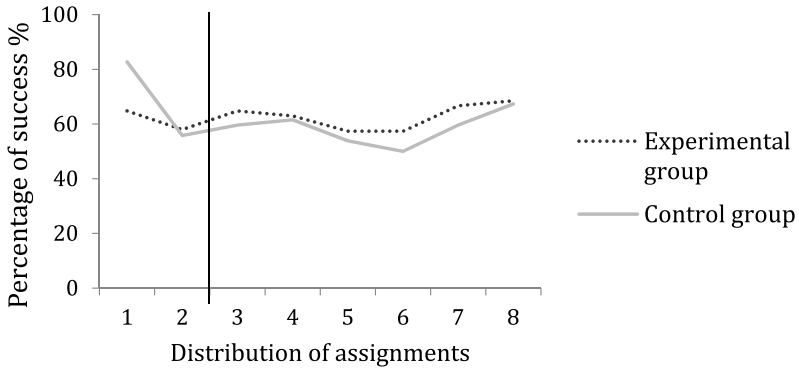

Figure 3. Progress of participants through the intervention. The vertical line represents the beginning of the intervention.

assessed with the MSET, the experimental group (following the metacognitive strategy training) performed better than the control group, which suggests an effect of our program on planning abilities. However, both the experimental and the control groups performed better in the post-intervention session compared to the pre-intervention session in other executive tasks, which does not allow us to conclude on the links between planning and other EF. Finally, the use of GAS methodology to assess goals related to planning difficulties in adults with PWS was feasible in this study. OTs have found this to be an effective way to formalize highly personalized and motivating goals for patients in their regular practice. Results of GAS showed that all participants progressed toward their goals but no difference was found between the two groups.

Assessing planning difficulties of patients can be challenging. As reported by Bennett et al.[51] on the DEX questionnaire, family members and patients tend to provide less accurate information than professionals because there are less trained to identify executive deficits. To obtain more accurate information the DEX should also be rated by healthcare staff according to the authors. Using the DAD-6 was also difficult for caregivers because some examples of activities included two different aspects, one easily achieved by the patient (managing things relative to leisure activities) and the other completely abandoned (cleaning the house). Phone-interview with caregivers after the completion of questionnaires showed the necessity of characterizing patient's difficulties more precisely than with questionnaires only. Individual interviews with participants often showed that difficulties and related goals proposed by caregivers were different from relevant goals for patients. Data from the DEX and the DAD-6 before and after the intervention provided information about evolution of participants' insight of difficulties and evolution of executive difficulties perceived by caregivers. DEX scores were different between caregivers and patients after the intervention with patients evaluating themselves better (lower scores) whereas caregivers evaluated them worst (higher scores). Those data can reflect that caregivers tend to search more information about the protocol and have higher expectations when the patient is engaging in the training, regardless of patient's allocation in the protocol. It can be noted that the DEX questionnaire assesses executive functioning on larger dimensions than what is worked on in the sessions; therefore, changes may be difficult for caregivers to perceive. It can also reflect the gap between patients' interests and caregivers' focus which can contribute to a lack of motivation if the rehabilitation does not focus on what is important for them. For this reason, using GAS for goal related to planning difficulties in patients with PWS appeared to be relevant. After the first individual interview with the OT, patients were able to understand the aim of the intervention which became more personalized.

Additional study parameters like between-session assignments served the intervention as they engaged the patients in the rehabilitation process and gave them feedback on their planning ability at each rehabilitation session. In the same logic, using GAS was a way to assess planning abilities in an ecological context and allowed involving patients in their rehabilitation. According to Benarroch et al.[5] this is particularly important for people with PWS as we need to approach each patient as an individual, to be sensitive to traits and strengths (related to and especially unrelated to PWS) and choose among most appropriate therapeutic approaches. This is why the involvement and motivation of patients in their care is an important point to take into account in the construction of a metacognitive strategy training program. Data regarding punctuality reflect the difficulty to implement this type of measure with PW patients during their stay at the hospital (e.g., activities canceled), to poor involvement of participants regarding this measure (e.g., lost sheets) and to planning difficulties (e.g., forgotten sheets not allowing to follow-up punctuality). Between-session assignments were also a good way to monitor the motivation and the implication of the participants even if no difference was found between the two groups in performance.

Regarding other outcomes measures, results suggest that the improvement was globally the same for the two groups, which may mean that patients with PWS can improve their executive tests performance. This may be due either to a test–retest effect or to the overall effect of the multidisciplinary rehabilitation stay, regardless of whether training of planning abilities was planned or not. There are number of reasons why a multidisciplinary rehabilitation may improve cognitive functioning nonspecifically: the time spend with the therapist, feeling supported ad enthusiastic by a novel protocol, patient's expectations, other factors taken care of

Table 6. Description of between-session assignments and percentage of success in each group of each assignment

Assignments	Experimental group	Control group	*t-test*
Get a copy of your rehabilitation activities timetable and give it to your occupational therapist	82.69 %	84.61	n.s.
Write down what is your weight this week	68.51	82.69	n.s
Check who is the doctor on call tomorrow	66.66	71.15	n.s
Choose one movie at the St Jean cinema for tomorrow and give the movie schedule	55.55	61.53	n.s
Find what is the next staff meeting to be attended by your occupational therapist	46.15	38.46	n.s
Find the address of the nearest post office	66.66	65.38	n.s.
Bring back a paper that interested you	79.62	50	< .05
Give the number or the colour of the bus line you should take to go to the train station tomorrow and the schedule to be there at 12 am	38.88	36.53	n.s

during the rehabilitation stay (i.e., sleep, depression, new treatment). The other activities proposed during the multidisciplinary rehabilitation (music-therapy, riding therapy, social activities) probably stimulated EF as well, and may explain the improvement of both groups in executive measures, as reported by many authors.[52] The specific impact of the ETAPP intervention can be difficult to extract from those. However as reported by Diamond and Ling,[53] including exercises that solicit EF in rehabilitation is important because it also improves feeling of self-confidence and self-efficacy, which in turn leads to improvement in executive functioning.

There is also a possible bias by using the same tasks in post-intervention if participants remember strategies of the previous tasks. On the other hand, limited effects of the training of EF in the experimental group can prove a lack of sensitivity to change of the outcome measures that were used: repeating a measure that focuses on an executive deficit is typically insensitive to the effectiveness of an intervention that aims to compensate for deficits.[53] Finally, expecting a transfer of increased planning ability does not seem realistic given the short duration of the training (six 1-h sessions): previous researches showed that the transfer of the executive training appears to be narrow and require a lot of practice, which was not the case here.[52]

The ETAPP program was designed to be a short intervention to correspond to the usual length of stay of PWS adults who cannot spend months in the rehabilitation center because they work in specialized establishments and have regular leisure activities, which made this intervention different from post-stroke rehabilitation where daily life "stops" because of the new disability. Moreover, in a neurodevelopmental disorder like the PWS, adults can be less motivated by a long rehabilitation. That is why the ETAPP program comprised six sessions only while others studies on metacognitive strategy training of EF recommend between 20 and 24 sessions.[31,54,55] Considering the low incidence of the PWS, we included a large number of patients. But the sample size may still be too small to detect small effects on others tests. One more limitation regarding the neuropsychological assessment was the use of neuropsychological tools already used on PW population in previous studies but not formally designed and validated for this population.

This study emphasizes the importance of personalized goal in rehabilitation, in order to improve engagement from participants in the rehabilitation. PWS patients perform better when there are stimulated and supported. Encouraging results about feasibility and planning performances showed that this type of rehabilitation program could be offered as part of treatment by OTs. However, for clinical use, this intervention may need further adaptation. For example, items on the procedural fidelity checklist with a low percentage give indication on possible limits that should be corrected in the future (e.g., working more precisely on some steps). Considering the small number of sessions imposed by the one-month stay, a second study of the ETAPP program should be considered to highlight the benefits of a more intensive support (e.g., intervention 4 times a week during 3 weeks). A longer or more intensive program would allow patients to integrate the metacognitive strategy and generalize it to various tasks requiring planning. Intervention for caregivers should also be considered to implement the training at home and to adapt the metacognitive strategy training on daily activities at home. The impact of such an intervention on patients' and caregivers' well-being should also be monitored in futures studies.

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Disclosure Statement

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References

1. Holm VA, Cassidy SB, Butler MG, Hanchett JM, Greenswag LR, Whitman BY, Greenberg F. Prader-willi syndrome: consensus diagnostic criteria. Pediatrics. 1993;91(2):398–402.
2. Cassidy SB, Driscoll DJ. Prader-Willi syndrome. Eur J Hum Genet. 2009;17(1):3–13. doi:10.1038/ejhg.2008.165.
3. Butler MG, Miller JL, Forster JL. Prader-Willi syndrome-clinical genetics, diagnosis and treatment approaches: an update. Curr Pediatr Rev. 2019;15(4):207–44. doi:10.2174/1573396315666190716120925.
4. Gunay-Aygun M, Schwartz S, Heeger S, O'Riordan MA, Cassidy SB. The changing purpose of Prader-Willi syndrome clinical diagnostic criteria and proposed revised criteria. Pediatrics. 2001;108(5):E92. doi:10.1542/peds.108.5.e92.
5. Benarroch F, Hirsch HJ, Genstil L,Landau YE, Gross-Tsur V. Prader-Willi syndrome: medical prevention and behavioral challenges. Child Adolesc Psychiatr Clin N Am. 2007;16(3):695–708. doi:10.1016/j.chc.2007.03.007.
6. Lewis BA, Freebairn L, Heeger S, Cassidy SB. Speech and language skills of individuals with prader-willi syndrome. Am J Speech-Lang Pat. 2002;11(3):285–94. doi:10.1044/1058-0360(2002/033).
7. Chevalère J, Postal V, Jauregi J,Copet P, Laurier V, Thuilleaux D. Assessment of executive functions and relationship with intellectual level. J Appl Res Intellect Disabil. 2013;26(4):309–18. doi:10.1111/jar.12044.
8. Chevalère J, Postal V, Jauregi J,Copet P, Laurier V, Thuilleaux D. Executive functions and Prader-Willi syndrome: Global deficit linked with intellectual level and syndrome-specific associations. Am J Intellect Dev Disabil. 2015;120(3):215–29. doi:10.1352/1944-7558-120.3.215.
9. Copet P, Jauregi J, Laurier V,Ehlinger V, Arnaud C, Cobo A-M, Molinas C, Tauber M, Thuilleaux D. Cognitive profile in a large French cohort of adults with Prader-Willi syndrome: differences between genotypes. J Intellect Disabil Res. 2010;54(3):204–15. doi:10.1111/j.1365-2788.2010.01251.x.

10. Jauregi J, Arias C, Vegas O,Alén F, Martinez S, Copet P, Thuilleaux D. A neuropsychological assessment of frontal cognitive functions in Prader-Willi syndrome. J Intellect Disabil Res. 2007;51(Pt 5):350–65. doi:10.1111/j.1365-2788.2006.00883.x.
11. Whittington J, Holland T, Webb T,Butler J, Clarke D, Boer H. Cognitive abilities and genotype in a population-based sample people with Prader-Willi syndrome. J Intellect Disabil Res. 2004;48(Pt 2):172–87. doi:10.1111/j.1365-2788.2004.00556.x.
12. Woodcock KA, Oliver C, Humphreys GW. Task-switching deficits and repetitive behaviour in genetic neurodevelopmental disorders: data from children with Prader–Willi syndrome chromosome 15 q11–q13 deletion and boys with Fragile X syndrome. Cogn Neuropsychol. 2009;26(2):172–94. doi:10.1080/02643290802685921.
13. Whittington J, Holland A. Cognition in people with Prader-Willi syndrome: insights into genetic influences on cognitive and social development. Neurosci Biobehav Rev. 2017;72:153-167. doi:10.1016/j.neubiorev.2016.09.013.
14. 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(1):49–100. doi:10.1006/cogp.1999.0734.
15. Shallice T, Burgess PW, Robertson IH. The domain of supervisory processes and temporal organization of behaviour. Philos Trans R Soc Lond B. 1996;351:1405–12.
16. Diamond A. Executive functions. Annu Rev Psychol. 2013;64 (1):135–68. doi:10.1146/annurev-psych-113011-143750.
17. Luria AR. Higher cortical functions in man. New York (NY): Basic Books Inc, 1996.
18. Wilson BA, Alderman N, Burgess PW,Wilson B, Evans JJ. Behavioural assessment of the dysexecutive syndrome. Bury St Edmunds (UK): Thames Valley Test Company; 1996
19. Martin R. Le test des commissions. Épreuve d'adaptation méthodique. 2e. Editest: Bruxelles, Belgique; 1972.
20. Rainville C, Amieva H, Lafont S,Dartigues J-F, Orgogozo M, Fabrigoule C. Executive function deficits in patients with dementia of the Alzheimer's type: a study with a tower of London task. Arch Clin Neuropsychol. 2002;17(6):513–30. doi:10.1093/arclin/17.6.513.
21. Estival S, Chevalère J, Laurier V,Mourre F, Postal V. Study of the deficit in planning abilities of adults with Prader-Willi Syndrome. 2021. 18p.
22. Krasny-Pacini A, Chevignard M, Evans J. Goal Management Training for rehabilitation of executive functions: a systematic review of effectiveness in patients with acquired brain injury. Disabil Rehabil. 2014;36(2):105–16. doi:10.3109/09638288.2013.777807.
23. Krasny-Pacini A, Limond J, Evans J,Hiebel J, Bendjelida K, Chevignard M. Context-sensitive goal management training for everyday executive dysfunction in children after severe traumatic brain injury. J Head Trauma Rehabil. 2013;29(5):E49–E64. doi:10.1097/HTR.0000000000000015.
24. Kennedy MRT, Coelho C, Turkstra L,Ylvisaker M, Moore Sohlberg M, Yorkston K, Chiou H-H, Kan P-F. Intervention for executive functions after traumatic brain injury: a systematic review, meta-analysis and clinical recommendations. Neuropsychol Rehabil. 2008;18(3):257–99. doi:10.1080/09602010701748644.
25. Levine B, Robertson IH, Clare L,Carter G, Hong J, Wilson BA, Duncan J, Stuss DT. Rehabilitation of executive functioning: an experimental clinical validation of goal management training. J Int Neuropsychol Soc. 2000;6(3):299–312. doi:10.1017/S1355617700633052.
26. Duncan J, Emslie H, Williams P,Johnson R, Freer C. Intelligence and the frontal lobe: the organization of goaldirected behavior. Cogn Psychol. 1996;30(3):257–303. doi:10.1006/cogp.1996.0008.
27. Cicerone KD, Langenbahn DM, Braden C, Malec JF, Kalmar K, Fraas M, Felicetti T, Laatsch L, Harley JP, Bergquist T, et al. Evidence-based cognitive rehabilitation: updated review of the literature from 2003 through 2008. Arch Phys Med Rehabil. 2011;92(4):519–30. doi:10.1016/j.apmr.2010.11.015.
28. Estival S, Krasny-Pacini A, Laurier V,Maugard C, Thuilleaux D, Postal V. Cognitive training targeting planning dysfunction in adults with Prader-Willi Syndrome: brief report of a study protocol. Dev Neurorehabil. 2019;22(8):569–75. doi:10.1080/17518423.2019.1642414.
29. Laurier V, Lapeyrade A, Copet P,Demeer G, Silvie M, Bieth E, Coupaye M, Poitou C, Lorenzini F, Labrousse F, et al. Medical, psychological and social features in a large cohort of adults with Prader-Willi syndrome: experience from a dedicated centre in France. J Intellectual Disability Res. 2015;59(4):411–21. doi:10.1111/jir.12140.
30. De Rotrou J, Wu Y-H, Hugonot-Diener L,Thomas-Antérion C, Vidal J-S, Plichart M, Rigaud A-S, Hanon O. DAD-6: a 6-item version of the disability assessment for dementia scale which may differentiate alzheimer's disease and mild cognitive impairment from controls. Dement Geriatr Cogn Disord. 2012;33(2–3):210–18. doi:10.1159/000338232.
31. Miotto EC, Evans JJ, Souza de Lucia MC. Rehabilitation of executive dysfunction: a controlled trial of an attention and problem solving treatment group. Neuropsychol Rehabil. 2009;19(4):517–40.
32. Ylvisaker M, Feeney T. Apprenticeship in self-regulation: supports and interventions for individuals with self-regulatory impairments. Dev Neurorehabil. 2009;12(5):370–79. doi:10.3109/17518420903087533.
33. Rath JF, Hradil AL, Litke DR,Diller L. Clinical applications of problem-solving research in neuropsychological rehabilitation: addressing the subjective experience of cognitive deficits in outpatients with acquired brain injury. Rehabil Psychol. 2011;56 (4):320–28. doi:10.1037/a0025817.
34. Hart T, Design BE. Implementation of clinical trials in rehabilitation research. Arch Phys Med Rehabil. 2012;93(8 Suppl):S117–S126. doi:10.1016/j.apmr.2011.11.039.
35. Spillane V, Byrne MC, Byrne M,Leathem CS, O'Malley M, Cupples ME. Monitoring treatment fidelity in a randomized controlled trial of a complex intervention. J Adv Nurs. 2007;60(3):343–52. doi:10.1111/j.1365-2648.2007.04386.x.
36. Wilson BA, Evans JJ, Emslie H,Alderman N, Burgess P. The development of an ecologically valid test for assessing patients with a dysexecutive syndrome. Neuropsychol Rehab. 1998;8(3):213–28. doi:10.1080/713755570.
37. Burgess PW. Theory and methodology in executive function research. In: Rabbitt P, editor. Methodology of frontal and executive function. Hove (UK): Psychology Press; 1997. p. 81–116.
38. Bertens D, Frankenmolen N, Boelen DH,Kessels RPC, Fasotti L. Validity of an adapted scoring method for a modified six elements test in individuals with brain injury. Arch Clin Neuropsychol. 2015;30(2):122–29. doi:10.1093/arclin/acu093.
39. Chevalère J Executive functioning and emotional processing in Prader-Willi syndrome: studies in cognitive neuropsychology and psychophysiology [dissertation]. Bordeaux (France): University of Bordeaux; 2014.
40. Li S-C, Schmiedek F, Huxhold O,Röcke C, Smith J, Lindenberger U. Working memory plasticity in old age: practice gain, transfer, and maintenance. Psychol Aging. 2008;23(4):731–42. doi:10.1037/a0014343.
41. Arrington CM, Logan GD. The cost of a voluntary task switch. Psychol Sci. 2004;15(9):610–15. doi:10.1111/j.0956-7976.2004.00728.x.
42. Burgess PW, Shallice T. Response suppression, initiation and strategy use following frontal lobe lesions. Neuropsychol. 1996;34 (4):263–73. doi:10.1016/0028-3932(95)00104-2.
43. Tournier I, Postal V, Mathey S. Investigation of age-related differences in an adapted Hayling task. Arch Gerontol Geriatr. 2014;59 (3):599–606. doi:10.1016/j.archger.2014.07.016.
44. Kiresuk TJ, Sherman RE. Goal attainment scaling: a general method for evaluating comprehensive community mental health programs. Community Ment Health J. 1968;4(6):443–53. doi:10.1007/BF01530764.
45. Krasny-Pacini A, Evans J, Moore Sohlberg M,Chevignard M. Proposed criteria for appraising goal attainment scales used as outcome measures in rehabilitation research. Arch Phys Med Rehabil. 2016;97(1):157–70. doi:10.1016/j.apmr.2015.08.424.
46. Alderman N, Burgess PW, Knight C,Henman C. Ecological validity of a simplified version of the multiple errands shopping test.

J Int Neuropsychol Soc. 2003;9(1):31–44. doi:10.1017/S1355617703910046.
47. Norris G, Tate RL. The Behavioural Assessment of the Dysexecutive Syndrome (BADS): ecological, concurrent and construct validity. Neuropsychol Rehabil. 2000;10(1):33–45. doi:10.1080/096020100389282.
48. Renison B, Ponsford J, Testa R,Richardson B, Brownfield K. The ecological and construct validity of a newly developed measure of executive function: the virtual library task. J Inter Neuropsychol Soc. 2012;18(3):440–50. doi:10.1017/S1355617711001883.
49. Nasreddine ZS, Phillips NA, Bédirian V,Charbonneau S, Whitehead V, Collin I, Cummings JL, Chertkow H. The Montreal Cognitive Assessment, MoCA: a brief screening tool for mild cognitive impairment. J Am Geriatr Soc. 2005;53(4):695–99. doi:10.1111/j.1532-5415.2005.53221.x.
50. Gouveia PAR, Brucki SMD, Malheiros SMF,Bueno OFA. Disorders in planning and strategy application in frontal lobe lesion patients. Brain Cogn. 2007;63(3):240–46. doi:10.1016/j.bandc.2006.09.001.
51. Bennett PC, Ong B, Ponsford J. Measuring executive dysfunction in an acute rehabilitation setting: using the dysexecutive questionnaire (DEX). J Int Neuropsychol Soc. 2005;11(4):376–85. doi:10.1017/S1355617705050423.
52. Diamond A, Ling DS. Conclusions about interventions, programs, and approaches for improving executive functions that appear justified and those that, despite much hype, do not. Dev Cogn Neurosci. 2016;18:34–48. doi:10.1016/j.dcn.2015.11.005.
53. Lewis MW, Babbage DR, Leathem JM. Assessing executive performance during cognitive rehabilitation. Neuropsychol Rehabil. 2011;21(2):145–63. doi:10.1080/09602011.2010.543867.
54. Rath JF, Simon D, Langenbahn DM,Sherr RL, Diller L. Group treatment of problem-solving deficits in outpatients with traumatic brain injury: a randomised outcome study. Neuropsychol Rehabil. 2003;13(4):461–88. doi:10.1080/09602010343000039.
55. Spikman JM, Boelen DHE, Lamberts KF,Brouwer WH, Fasotti L. Effects of a multifaceted treatment program for executive dysfunction after acquired brain injury on indications of executive functioning in daily life. J Int Neuropsychol Soc. 2010;16(1):118–29. doi:10.1017/S1355617709991020.

APPENDIX A. Procedural fidelity checklist

Procedural fidelity items	%
Feasibility of the ETAPP programme	
Participants understand the session content	100
OT encourages monitoring of errors	33.66
Participants help each other	34.43
OT encourages everyone to participate	100
OT keeps participants on track (if attentional drift)	100
Participants understand the story (step "PAUSE")	100
Participants are able to "brainstorm" about the task's steps with the help of the OT (step "LIST")	66.7
Participants write down the steps or paste pre-printed steps in their work book (step "LIST")	66.7
Participants use the step list while performing the task (step "DO IT")	100
Participants show ability to return to the list of steps (step "DO IT")	33.3
Participants verify that they have followed the plan (step "VERIFY")	100
Content of sessions	
PAUSE: presentation of the story	100
PAUSE: choosing a target activity on which the strategy will be applied	100
PAUSE: illustration of the step	100
DEFINE: recall of the target activity	100
DEFINE: explanation of the step	100
DEFINE: OT questions about the difficulty of the task	100
DEFINE: OT makes participants think about their strengths and weaknesses	100
DEFINE: OT makes participants think about potential obstacles	100
LIST: recall of the target activity	100
LIST: feedback on the step	100
LIST: illustration of the step	100
LIST: explanations of the advantages of listing steps in a task	100
LIST: listing the steps needed for the target activity	100
DO IT: Recall of the target activity	87.5
DO IT: Presentation of the step	87.5
DO IT: Re-read the list of steps	87.5
DO IT: Beginning of target task execution	87.5
DO IT: Execution following the steps' list	87.5
DO IT: OT recall the step 'PAUSE' during execution	25
DO IT: Target task execution	100
DO IT: OT makes participants verify if they follow the list	100
EVALUATE: Recall of the target activity	100
EVALUATE: Presentation of the step 'EVALUATE'	100
EVALUATE: Checking target task execution	100
EVALUATE: OT asks about goal achievement	100
EVALUATE: OT asks about the benefit of evaluation	100
Feedback on assignments and punctuality	
Checking punctuality sheets	94.4
Checking assignments: succeeded/attempted but failed/not attempted	100
OT asks participants how they feel	100
Advice for next time	94.4
Explanations of new assignments	100

APPENDIX B. Examples of goal types

General objective	Examples of personalized objectives
Calendar/weekly organization	Have a global vision of your daily life (ESAT, holidays, activities, medical appointments) by using an adapted tool Record in the agenda provided by the occupational therapist the activities she has every day and add the occasional appointments Learn to use the phone calendar for appointments with an audible reminder when preparing to leave (e.g., 15 minutes before) Be able to quantify the time required to prepare herself in the morning and to get to the activities (estimated travel time)
Weight management	Take ownership of the weight curve and invest it during his stay in order to be able to continue his follow-up at home
Suitcase	Be able to prepare your suitcase for your departure from Hendaye in an organized way in the presence of a caregiver Become aware of the notions of limited quantities and deviations from the inventory of the suitcase to come to Hendaye
Laundry	Learn to become less dependent in the management of your laundry by developing your ability to make a washing machine during your stay Get more involved in the management of your business by tracking and calculating the laundry budget over 3 weeks