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Study of the deficit in planning abilities of adults with Prader-Willi Syndrome

Séverine Estival [a,*], Johann Chevalère [a,1], Virginie Laurier [b], Fabien Mourre [b], Julie Tricot [b], Virginie Postal [a]

[a] *Laboratoire de Psychologie LabPsy EA4139, Université de Bordeaux, France*
[b] *Hôpital Marin AP-HP, Unité Prader-Willi, Hendaye, France*

A R T I C L E I N F O

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A B S T R A C T

Background: Prader-Willi syndrome (PWS) is a complex developmental genetic disorder associated with intellectual disability and deficits in executive functions which result in disorganisation and poor personal autonomy.
Aims: This study aimed to determine impairments in planning skills of adults with PWS, in relation with their intellectual disabilities, as well as the influence of food compulsions on their performance.
Methods and procedures: A modified version of the Zoo Map from the Behavioural Assessment of the Dysexecutive Syndrome was used in three groups: a group of adults with PWS in comparison with two groups both matched on chronological age, one with typical development (TD) and one with intellectual disability (ID).
Outcomes and results: Compared to TD adults, both adults with PWS and ID showed increased planning time and lower raw scores on the planning task. The execution time and the number of errors were higher in the PWS group compared to the comparison groups. All three groups performed worse in the non-food condition only for number of errors and raw score.
Conclusions and implications: Planning abilities were impaired in PWS adults. Results also showed that intellectual level plays a role in participants' performance. These findings are essential to understand the difficulties of people with PWS daily life.

What this paper adds?

This paper provides a specific study of planning abilities in adults with Prader-Willi Syndrome compared to two other groups matched on chronological age and intellectual disability respectively. It adds information on the concept of planning abilities as not a unitary concept. Results also allows to understand the importance of intellectual level. Planning abilities and intellectual level should be considered more systematically to improve care management practices in patients with PWS. Highlights of this study gave us futures directions for a cognitive rehabilitation programme designed to improve planning abilities in adults with PWS.

* Corresponding author at: 3Ter Place de la Victoire, Bordeaux Cedex, 33076, France.
E-mail address: s.estival@live.fr (S. Estival).
[1] J. Chevalère is now working at the Department of Educational Sciences, University of Potsdam, Germany.

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1. Introduction

Prader-Willi syndrome (PWS) is a complex neurodevelopmental disorder caused by a lack of paternal expression on the q11-q13 region of chromosome 15. In 60 % of cases, PWS is the result of a paternal deletion of the 15q11-q13 region of the chromosome. In 35 % of cases, the syndrome is due to maternal uniparental disomy (m-UPD). In the remaining cases, PWS is thought to be caused either by a defect in the imprinting centre or by a balanced translocation involving a breakpoint in the 15q11-q13. The percentage given relate to estimations made last century (Butler, Miller, & Forster, 2019). PWS is characterised by neonatal hypotonia, poor sucking reflexes and delayed motor development in early infancy, followed by excessive appetite (hyperphagia) and obsessive food-seeking in childhood that can lead to morbid obesity in adulthood if not kept under control (Gunay-Aygun, Schwartz, Heeger, O'Riordan, & Cassidy, 2001). The food dimension is central to the syndrome, as people with PWS have a constant sense of hunger and the presence of food can disturb their performance (Hinton, Isles, Williams, & Parkinson, 2010; Key & Dykens, 2008). PWS also involves a range of difficulties, such as physical, behavioural and intellectual symptoms (Holm et al., 1993). Key features include repetitive and compulsive behaviours (Woodcock, Oliver, & Humphreys, 2011), temper outbursts, resistance to change (Tunnicliffe, Woodcock, Bull, Oliver, & Penhallow, 2014), impaired social functioning (Dimitropoulos, Ho, & Feldman, 2013), delayed development of speech and language (Lewis, Freebairn, Heeger, & Cassidy, 2002), difficulties in understanding the behaviour and mental states of others (Whittington & Holland, 2011), a general slowdown in the processing of information, extreme fatigability and learning disabilities (Foti et al., 2015; Whittington et al., 2004). Some of these impairments can be attributed to their low intellectual level (i.e., IQ level). Previous research focusing on intellectual disabilities (ID) in people with PWS showed a deficit of cognitive processes and more specifically, impairments in higher cognitive processes, known as executive functions (Jauregi et al., 2007).

Executive functions can be defined as high-level cognitive processes that allow flexible and context-appropriate behaviour when an individual is confronted with a new or complex situation (Friedman & Miyake, 2017; Mikaye et al., 2000; Shallice & Burgess, 1996). The deficit in executive functioning leads to difficulties in managing new situations and in managing family, social or professional life (Diamond, 2013). According to Stuss and Benson (1984) "executive control" activates specific functions in order to find solutions in response to new situations: anticipation, selection of behaviour, monitoring of performance, feedback use and planning. Planning can be defined as the ability to anticipate and organise series of actions in an optimal sequence to achieve a goal (Luria, 1966). It involves using effective strategies, prioritising, anticipating and predicting the steps in a task (Shallice & Burgess, 1991). In order to do so, planning requires different abilities: maintaining the goal, planning ahead by elaborating different strategies, choosing among them the most suitable action plan to successfully achieve the goal, and initiating the selected action plan while taking account of incidents or changes necessary to achieve the goal (Levine et al., 2000), with constant feedback between those abilities. However, it remains unsure what cognitive processes underlie the planning function (Rose et al., 2015). To date, there is no consensus regarding the deficit of planning in people with ID: either the capacity is relatively preserved (Danielsson, Henry, Rönnberg, & Nilsson, 2010) or there is a deficit indeed (Alloway, 2010). Furthermore, when compared with an age-matched population, planning seems to depend primarily on mental age (Danielsson, Henry, Messer, & Rönnberg, 2012). This lack of consensus possibly relies on the difficulty to evaluate which factors are impaired: it could be the goal maintenance, the ability to follow the steps through which the goal is achieved, or the ability to adapt to unexpected changes (Levine et al., 2000).

In PWS, it has been suggested that intellectual impairments are involved in social behaviour disorders (Woodcock et al., 2011) and some behaviours observed in PWS (e.g., repetitive and perseverative speech, tantrums and intolerance to change) do suggest a deficit of executive functions (Walley & Donaldson, 2005). Previous studies have shown a planning deficit in PWS with a battery of executive tasks (Chevalère et al., 2013, 2015). These deficits may translate into disorganisation and poor personal autonomy in daily life. Most adults with PWS live in their family home or in a care system, under guardianship measures (Laurier et al., 2015). Having access to an autonomous life is complicated and executive deficits, especially on planning abilities, may determine the patient's level of autonomy (i.e., being able to take appointments, to take the bus by oneself, to attend to an activity). Considering that one of the rehabilitation priorities for families is to reduce adjustment difficulties (Pituch et al., 2010), there is a need to clarify planning abilities in adults with PWS. This step is essential to propose adapted interventions and help them enhance their independence in daily living.

The present study aimed to evaluate planning abilities in adults with PWS by conducting analyses of group differences. This first objective was operationalised using a planning task based on the Zoo Map test from the Behavioural Assessment of Dysexecutive Syndrome (BADS, Wilson, Alderman, Burgess, Emslie, & Evans, 1996) where participants planned their route on a map while complying with certain rules, thereby reproducing a situation that each participant can experience in real life. The task required elaborating all the steps needed to achieve the goal correctly while following the rules, prior to their execution. We used a task with an ecological validity meaning that it reflects a set of mechanisms that people are supposed to use in real life while offering a fine control over crucial parameters such as time, number of locations, number of actions and number of distractors (Wilson et al., 1996). We also examined whether the planning deficit was specific to PWS or explained by intellectual disability. This second objective was evaluated with two comparison groups both matched on chronological age, one with typical development (TD group) and one with non-specific intellectual disability (ID group). More precisely, the former group comparison aimed at assessing whether PWS adults showed an impairment of planning abilities and its magnitude relative to the typically developing population. The latter group comparison was useful to determine whether the expected impairment in planning abilities was accounted for by IQ level. Indeed, comparing the syndrome under study to groups with non-specific intellectual disabilities has been recommended and used in previous work to isolate syndrome-specific psychological features from general ID (Dykens & Hodapp, 2001; Hodapp & Dykens, 2001). This data will help us understand the role of ID in a high-level cognitive function such as planning. Studying the presence of these specific impairments would allow for the development of intervention programs targeting planning difficulties while considering individual IQ, which objectives would aim at reducing these difficulties in daily life. IQ indices were considered in order to discuss the possible influence of other cognitive processes on planning-related functions. Finally, this study sought to go one step further previous work showing planning impairments in PWS (Chevalère et al., 2015). Given the overwhelming influence of food in PWS (Hinton et al., 2010; Key & Dykens, 2008), our third objective was to examine how the fictional presence of food could impact planning abilities when irrelevant for the task. As the original task required visiting a number of locations while avoiding others, a modified version of the task with two conditions that included food items was proposed in our study. In one condition, participants had to draw their path through food-related locations only (e.g., a bakery). In another condition, they had to draw their path while avoiding these locations. We expected an interaction between group and condition (food vs. non-food) where planning scores across conditions should be more nuanced in PWS compared to the two control groups. Participants with PWS should show lower planning performance in the non-food condition, as obsessive thoughts relative to food should particularly distract them from the task goal, whereas these thoughts could help them maintain the task goal in the food condition.

2. Method

2.1. Participants

Three groups of participants were recruited in accordance with the Declaration of Helsinki and the World Medical Association. All ethical aspects of the recruitment processes were governed by the French Health Code. Participants were invited to participate on a voluntary basis and provided informed consent after being informed of the aims of the present study in the presence of their legal guardian when they had one, and a personnel representative of the institution where the recruitment process took place. In compliance with the Article L. 1122-2 of the French Public Health Code, the informed consent was adapted to participants' level of understanding with different versions of the consent form provided to healthy adults and adults with PWS or ID. Participants with PWS were 30 adults recruited during a stay at the Hôpital Marin of Hendaye, a dedicated centre for PWS patients in France. The recruitment process was observed under the framework of a research convention between the Hôpital Marin of Hendaye and the University of Bordeaux. The medical staff was present during the recruitment process. All adults with PWS all had genetic confirmation of the diagnosis: the group contained 23 cases of deletion (76.6 %) and seven cases of m-UPD (23.4 %). Twelve participants were men and 18 were women. The age range of participants with PWS was 20–41 years ($M = 28.53$, $SD = 5.30$ years). The group of adults with non-specific ID comprised unqualified persons who left the formal education system very early and joined medico-social structures for disabled workers who cannot carry on a professional occupation in ordinary sectors of the labour market. The recruitment process of adults with ID was observed in the framework of a Research Agreement between a Work Assistance Establishment, the Adapei Association of Gironde and the University of Bordeaux. The tutors of the medico-social structures were aware of the recruitment process. The group of adults with ID was composed of 30 people, 17 men and 13 women. The age range of the participants was 21–41 years ($M = 30.13$, $SD = 6.67$ years). None of these persons had a known medical record of any genetic disorder or neurological condition. The TD group, recruited at the University of Bordeaux, was composed of 30 participants, 12 men and 18 women with an age range of 18–41 years ($M = 26.76$, $SD = 5.82$ years).

Participant's characteristics (PWS group, ID group and TD group) are summarised in Table 1. No age difference was found between the PWS group and the TD group, $t(58) = 1.22$, $p = 0.96$. No difference between the PWS group and the ID group was found for FSIQ, $t(58) = 0.32$, $p = 0.90$.

2.2. Measures

The Wechsler Adult Intelligence Scale III edition (WAIS-III, Wechsler, 1997) was administered to all the participants in the same order as described in the user's guide. The WAIS-III evaluation allowed obtaining a Full Scale Intelligence Quotient (FSIQ) for the participants and four indexes of IQ. The verbal comprehension index (VCI) measures the capacity to apply knowledge to reasoning skills and the ability to retrieve information, vocabulary and verbal-reasoning. The perceptual reasoning index (PRI) reflects the ability to interpret, organise and think with visual information. It measures nonverbal reasoning skills and requires perceptual abilities. The working memory index (WMI) measures the capacity to maintain and manipulate visual and verbal information in working memory. The processing speed index (PSI) reflects to accuracy to identify visual information, making decisions and acting on these decisions.

Table 1
Participant's characteristics.

	PWS adults ($n = 30$)		TD group ($n = 30$)		ID group ($n = 30$)	
	Mean	(*SD*)	Mean	(*SD*)	Mean	(*SD*)
AGE	28.53	(*5.30*)	26.76	(*5.82*)	29.75	(*6.67*)
FSIQ (WAIS-III)	65.7	(*8.51*)	112.5	(*11.79*)	64.9	(*10.19*)
VCI	75.03	(*10.40*)	112.3	(*12.41*)	69.48	(*11.58*)
PRI	71.89	(*11.09*)	111.44	(*14.47*)	73.34	(*13.43*)
WMI	67.46	(*15.92*)	110.07	(*15.70*)	63.28	(*13.39*)
PSI	66.74	(*15.84*)	115.10	(*11.00*)	71.13	(*15.99*)

Note. FSIQ = Full Scale Intelligence Quotient; VCI = Verbal Comprehension Index; PRI = Perceptual Reasoning Index; WMI = Working Memory Index; PSI = Processing Speed Index.

In order to allow manipulation of the presence of food items, we adapted the original BADS Zoo Map (Wilson et al., 1996) and proposed a modified version named "The Market Map". The test is a map of a market place with different shops (shops related to food and shops unrelated to food). The organisation of the different locations is identical to the original version of the task, except that zoo sites have been replaced by market shops (e.g., bakery, fishmonger, bag seller, jewellery). The participant had to plan and then execute –draw– a path while complying with certain rules. Depending on the condition, participants had to organise their path by visiting either food related shops (food condition) or non-food related shops (non-food condition) exclusively. One measure was the average time taken by participants to anticipate and plan their future path. This planning time stopped when participants started to draw their path. The execution time started when participants began to draw their path. There was also a measure of the average number of errors including a path visited more than once, deviations from the path, failures to make a continuous line, inappropriate places visited and forgotten places (Oosterman, Wijers, & Kessels, 2013). According to the BADS' Manual (Wilson et al., 1996), the raw score for each version was created by the number of locations visited in the correct order (sequence score) with points deducted if an error was made (range: 0–8). In order to avoid a familiarisation effect, the orientation of the tests was reversed (top entry vs. bottom entry) and counterbalanced for each condition and for each participant. Similarly, the presentation of the two conditions was counterbalanced in each group. See Appendix A for more details about the Market Map.

2.3. Procedure

All the participants took the two conditions (food vs. non-food) of the test individually with an additional interference task in between. The interference task measured the theory of mind and was part of a complete protocol designed to assess cognitive and executive functioning of people with PWS. First, instructions of the modified version of the Zoo Map were read aloud with the participant, several times if necessary. Participants had to be able to rephrase the instructions in their own words before starting the test. Then, participants were invited to think about their path in their head and were encouraged to take their time to plan it best. After this planning period, participants started to draw the path on the map using a set of coloured markers providing information on the sequence of movements. The instructions remained visible at any time on the same sheet where the map was pictured, and could also be recalled by the experimenter at any time.

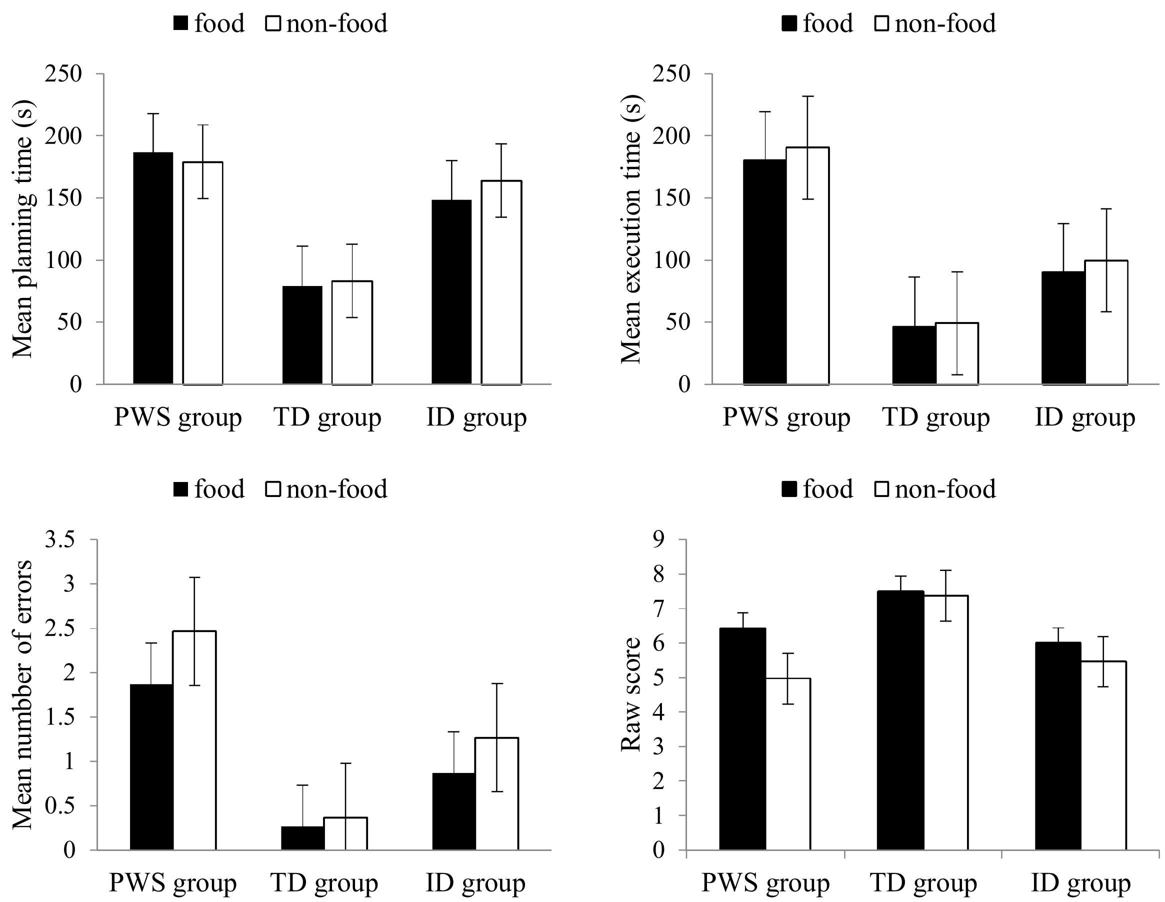

Fig. 1. Mean performance for each group.

3. Results

Data were analysed to compare group performances and examine the relationship with the FSIQ score, the four WAIS-III indexes and participant's age. Descriptive and analytical statistics were calculated in order to compare group's scores. Assuming a probability $p > 0.05$ for the normality of the distribution and regarding the sample size, we performed parametric analysis. All test results were considered significant at $p \leq 0.05$. The four dependent variables (planning time, execution time, number of errors and raw score) were analysed separately using a similar 3 (group: PWS; ID; TD) × 2 (condition: food; non-food) mixed-design ANOVA with the 3 groups as between-participants factor and the 2 conditions (food or non-food) as within-participants. Due to an error in the data collection, the execution times for 8 participants were not available (3 in the TD group and 5 in the ID group).

The 3 × 2 mixed-design ANOVA revealed group differences on the mean planning time, $F(2, 87) = 5.59$, $p < 0.001$, $\eta_p^2 = 0.11$. Both the PWS group (182.9 s, $p = 0.002$) and the ID group (156.2 s, $p = 0.02$) took longer to plan their path than the TD group (81.3 s). There was no difference between the food (138.2 s) and non-food (142 s) conditions of the task ($F < 1$) and no significant Group x Condition interaction effect on the mean planning time ($F < 1$). Regarding execution time, a similar analysis revealed a difference between the three groups, $F(2, 79) = 39.84$, $p < 0.001$, $\eta_p^2 = 0.50$. PWS adults (185.2 s) took longer to execute their path compared to ID group (96.7 s, $p < 0.001$) and TD group (48 s, $p < 0.001$). In turn, the ID group took longer than the TD group ($p = 0.002$) to execute their path. There was no difference between the food (105.9 s) and non-food (114 s) conditions of the task for any group, $F(1, 79) = 1.50$, $p = 0.22$, and no significant Group x Condition interaction effect on mean execution time ($F < 1$).

Regarding the number of errors, the 3 (group) × 2 (condition) mixed-design ANOVA design revealed a difference between the three groups, $F(2, 87) = 7.98$, $p < 0.001$, $\eta_p^2 = 0.15$. Adults with PWS (2.20) underperformed the TD group (0.26, $p < 0.001$) and the ID group (1.10, $p = 0.02$). There was no difference between the two latter groups ($p = 0.09$). Participants made more errors in the non-food (1.36) relative to the food condition (1.00), $F(1, 87) = 4.05$, $p = 0.03$, $\eta_p^2 = 0.04$. There was no significant Group x Condition interaction effect on the mean number of errors ($F < 1$). Regarding raw scores, results of the 3 × 2 ANOVA revealed a difference between the three groups, $F(2, 87) = 8.25$, $p < 0.001$, $\eta_p^2 = 0.16$. The raw score was higher in the TD (7.13) than in the PWS group (4.31, $p < 0.001$) and the ID group (5.05, $p < 0.01$). There was no difference between the two latter groups ($p = 0.29$). The raw score was higher in the food (5.81) than in the non-food (5.18) version of the task, $F(1, 87) = 4.12$, $p = 0.04$, $\eta_p^2 = 0.05$. There was no significant Group x Condition interaction effect on the raw score, $F(1,87) = 1.55$, $p = 0.21$. All the results are displayed in Fig. 1.

Next, we described each group's pattern of errors in more details by dividing errors types into 5 categories: using a path more than once, deviations from the path, failures to make continuous line, forgotten locations and inappropriate places visited (Oosterman et al., 2013). Results are displayed in Fig. 2. The most common mistake was using a path more than once (25.5 % of all participants in the food condition and 32.2 % in the non-food condition). This was the principal error in the TD group (13.3 % in the food condition and 16.6 % in the non-food condition). There was only one participant from the PWS group who deviated from the path in both conditions reunited. In the food condition, 10 % of PWS adults and 13.3 % of the ID group failed to make continuous lines, those results increased in the non-food condition with 20 % of PWS adults and 23.3 % of ID group. However, none of the PWS adults forgot to visit locations, whereas this specific error was committed by the TD group (3.3 % in the food condition and 10 % in the non-food condition) and the ID

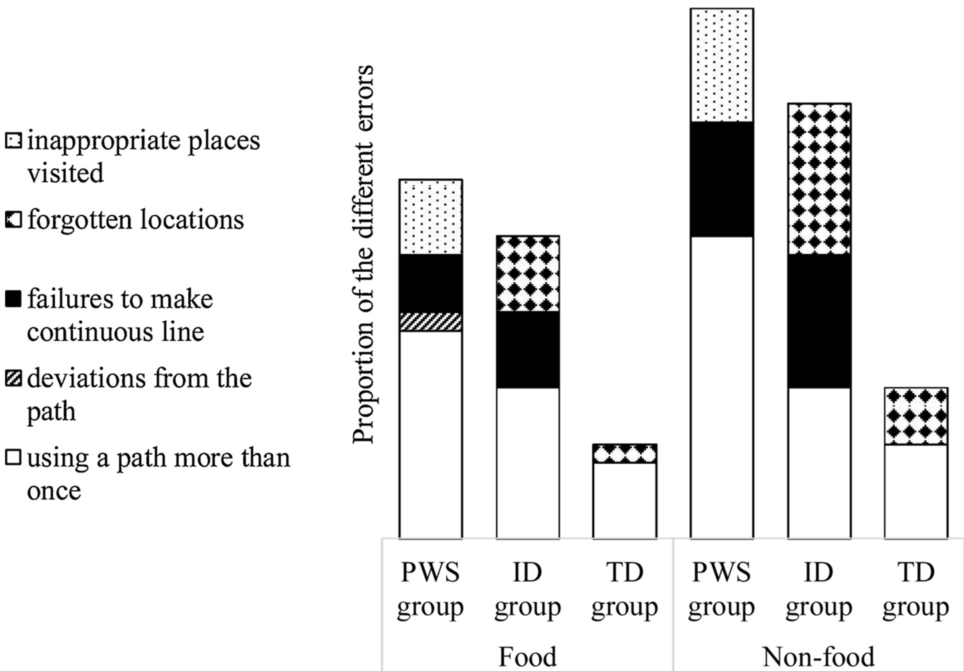

Fig. 2. Pattern of errors according to the group.

group (13.3 % in the food condition and 26.6 % in the non-food condition). Finally, in comparison to the two other groups, adults with PWS were the only ones to visit inappropriate places (13.3 % in the food condition and 20 % in the non-food condition).

To examine the relations between participants' performances on the Market Map and their characteristics in age and IQ (FSIQ, VCI, PRI, WMI and PSI), we conducted Pearson's bivariate correlational analyses. Results are displayed in Table 2. The correlation between planning time and execution time and between planning time and number of errors were significant and positive in the TD group (respectively $r = 0.62$, $p < 0.001$ and $r = 0.44$, $p = 0.015$), but non-significant in the PWS and ID group. No other meaningful relation was found in the TD group. A similar pattern of group specificities (i.e., PWS vs. ID groups) was found regarding the IQ indexes-planning indicators links. However, a specific negative relationship between execution time and FSIQ ($r = -0.46$, $p = 0.009$) was found only in the PWS group. Furthermore, in the PWS group, VCI was moderately related both to planning time ($r = 0.39$, $p = 0.039$), number of errors ($r = -0.41$, $p = 0.03$) and raw score ($r = 0.46$, $p = 0.014$), WMI was moderately related to execution time ($r = -0.39$, $p = 0.045$), number of errors ($r = -0.40$, $p = 0.043$), and raw score ($r = 0.64$, $p < 0.001$), and PSI was moderately related to raw score ($r = 0.43$, $p = 0.025$). These relations were absent in the ID group. Conversely, in the ID group, PRI was moderately related to planning time ($r = -0.37$, $p = 0.043$) and execution time ($r = -0.40$, $p = 0.038$), and PSI was moderately related to execution time ($r = -0.50$, $p = 0.009$). These relations were absent in the PWS group. Finally, there were no correlations between age and planning indicators, except in the ID group, where it negatively predicted the raw score.

4. Discussion

The first aim of this study was to assess planning abilities in people with PWS. Given the presence of ID in PWS, the second objective was to determine whether the presence of any deficit could be attributed to syndrome-specific features or explained by global intellectual level. Our third objective was to examine possible associations between deficits and food obsessions/compulsions that people with PWS might experience when performing an ecological planning task involving food items. With our modified version of the BADS' Zoo Map, "The Market Map", we were able to assess several planning functions: the ability to understand and maintain the goal of the task, the ability to anticipate the steps of the task during the planning time, the ability to sequence the plan into sub-goals following the rules of the task and lastly the ability to take incidents into consideration during execution if the participant realizes that there is an error. Moreover, two conditions of the task (to visit or to avoid food-related places) allowed studying the influence of food during planning in order to highlight its potential facilitative or deleterious effects. The Market Map, based on the Zoo Map by Wilson et al. (1996), is particularly relevant for people with PWS because instructions were presented visually throughout the testing period, which reduces the use of working memory during the task and genuinely measures planning ability (Oosterman et al., 2013). Considering PWS adults' cognitive impairments, it was important to propose a task where only planning abilities were evaluated, as far as possible.

First, we examined planning time, which corresponds to the mean time taken by participants to elaborate and anticipate the path

Table 2
Pearson's bivariate correlations.

		Mean time of planning *r*	Mean time of execution *r*	Mean number of errors *r*	Raw score *r*
PWS group	FSIQ	*0.33*	*−0.46***	*−0.53***	*0.68***
	VCI	*0.39**	*−0.28*	*−0.41**	*0.46**
	PRI	*−0.01*	*−0.29*	*−0.44**	*0.49***
	WMI	*0.26*	*−0.39**	*−0.40**	*0.64***
	PSI	*0.15*	*−0.18*	*−0.33*	*0.43**
	Age	*−0.08*	*0.05*	*0.30*	*−0.31*
	Mean time of planning	–	*0.16*	*0.26*	*0.30*
	Mean number of errors	*0.26*	*0.32*	–	*−0.80***
TD group	FSIQ	*−0.10*	*−0.00*	*−0.03*	*0.07*
	VCI	*−0.28*	*0.01*	*0.07*	*−0.06*
	PRI	*−0.08*	*−0.03*	*−0.28*	*0.33*
	WMI	*−0.18*	*0.03*	*−0.14*	*0.19*
	PSI	*−0.10*	*0.00*	*0.23*	*−0.09*
	Age	*0.00*	*0.09*	*0.28*	*−0.19*
	Mean time of planning	–	*0.62***	*0.44**	*−0.34*
	Mean number of errors	*0.44**	*0.43**	–	*−0.91***
ID group	FSIQ	*−0.33*	*−0.28*	*−0.52***	*0.48***
	VCI	*−0.27*	*−0.10*	*−0.29*	*0.31*
	PRI	*−0.37**	*−0.40**	*−0.56***	*0.59***
	WMI	*−0.18*	*0.03*	*−0.23*	*0.03*
	PSI	*−0.15*	*−0.50***	*−0.35*	*0.22*
	Age	*0.28*	*0.15*	*0.31*	*−0.50***
	Mean time of planning	–	*0.11*	*0.02*	*−0.24*
	Mean number of errors	*0.02*	*0.28*	–	*−0.84***

Note. FSIQ = Full Scale Intelligence Quotient; VCI = Verbal Comprehension Index; PRI = Perceptual Reasoning Index; WMI = Working Memory Index; PSI = Processing Speed Index.

* $p < 0.05$.
** $p < 0.01$.

mentally before drawing it. Planning time was significantly longer in adults with PWS and ID relative to healthy adults. This indicates that a longer planning time was not specific to PWS adults and suggests the involvement of IQ level in this performance. While PWS and ID groups showed similarities in planning time, their determinants differed. In the PWS group, we found that a better verbal comprehension was associated with longer planning time whereas in the ID groups longer planning time were associated with better perceptual reasoning skills. Those results are in line with Stuss and Benson's (1984) view of executive control involving various processes like anticipation, selection of behaviour, monitoring of performance, feedback use and planning components. The variety of processes involved may explain why the origin of deficits may be different in PWS and in more general ID (Jauregi et al., 2007; for a review, see Whittington & Holland, 2017). Second, regarding execution time, it appears that PWS adults took longer to draw their path compared to the two other groups. However, processing speed did not account for this longer execution time, as it did in the ID group. Rather, execution time was predicted by the working memory index in the PWS group. Weakness in short-term memory is a well-known characteristic of PWS adults (Conners, Rosenquist, Atwell, & Klinger, 2000) and may reflect difficulties in maintaining the planned actions during execution in this specific population. Third, a large number of errors suggests specific impairment of planning abilities in the PWS group as these participants showed the highest error rates, which were not compensated by longer planning times. This suggests that the anticipation of the steps as well their execution may have been difficult for them and that low verbal comprehension and working memory may have played a role in it. After the task started, participants did not tend to reread the instruction. This was especially true for the PWS group despite the total time taken to complete the task. This is consistent with the fact that these patients tended to lose sight of their goal during the task, but were not much concerned about it, unlike the other two groups. Furthermore, our exploratory analysis of the pattern of errors in each group revealed a specificity in the PWS group: adults with PWS never forgot to visit locations but tended to visit inappropriate places, unlike the two control groups. This important finding points out to difficulties in inhibiting irrelevant information, which is a major function of working memory (Baddeley, 1986; Engle, 2018; Hasher & Zacks, 1988). This is consistent with PWS-specific influences of working memory on execution time, number of errors and raw score, and with previous research showing deficits in PWS on inhibition tasks (Chevalère et al., 2015). Regarding the number of errors in particular, a deficit of inhibition processes is favoured over a short-term deficit as there were no forgotten locations but only inappropriately-visited locations. Furthermore, although task switching deficits are well documented in the PWS (Woodcock et al., 2011), our results rather favour an interpretation in terms of an inhibition deficit, as we did not observe a higher number of errors when the second version of the task was administered. Further studies should emphasize more directly on the how inhibition deficits may influence error rates and error types in ecological planning tasks.

Considering the central dimension of food in PWS, we investigated whether the fictional presence of food could modulate planning performance by having participants plan their imaginary shopping activity in two conditions manipulating the presence vs. avoidance of food-related shops as the goal of the task. Results showed that planning time and executive time were not affected by condition, but all participants performed worse in the non-food condition in terms of number of errors and raw score. This is consistent with studies showing automatic attentional capture to task-irrelevant food items in healthy participants affecting task performance (Cunningham & Egeth, 2018; Motoki, Saito, Nouchi, Kawashima, & Sugiura, 2018). However, these results do not allow us to confirm that avoiding food-related shops (i.e., the non-food condition) would make planning more difficult especially in PWS. The examination of error patterns across conditions revealed that the number of inappropriately visited places in PWS did not vary across conditions, thus not supporting the hypothesis that inhibition deficits would have increased when presented with distracting, to-be-avoided food items (i.e., consistent with well-documented food-related obsessions and compulsions in PWS, Hinton et al., 2010; Key & Dykens, 2008). This is surprising given that the food condition elicited more errors in all participants, demonstrating that the task was sensitive enough to elicit task-irrelevant food-related thoughts. However, this study does not conclude whether it was the nature of the food itself or the actual categories' names presented during the instruction (an identifiable "food" category versus a coarse "non-food" category) that caused more errors in the food condition.

It might be that adults with PWS, who are usually aware of their food-related problems, made a particular effort to do well in the task and reach seemingly normal performance ranges, as evidenced by the surprising absence of forgotten places (in regard to known short-term memory impairments), an error type that even the TD group committed. This hypothesis could be tested in further studies using the same task among participants expressing high vs. low concerns about their daily food-related problems. An alternative explanation of the results is that the use of food names distracted people with PWS so that they could not maintain the goal in memory. Because they were present in both conditions, food names may have been equally salient for adults with PWS and as a result, equally distractive, which may alternatively account for the similarity of performance in both conditions.

The main finding confirms the deficit of planning in people with PWS, as found in previous studies (Chevalère et al., 2013, 2015). Participants with PWS failed to anticipate the steps of the task and to initiate a plan while taking account of incidents, for example, being capable to correct themselves when using the same path more than once. Using effective strategies was also difficult for them, for example, in ensuring that they visited appropriate locations only. To our knowledge, the present study is the first to offer a precise examination of planning abilities in order to describe specific impairments. Further studies should ascertain that the higher number of incorrectly visited locations observed in the PWS group is caused by a deficit in inhibition or because they lost sight of the goal due to distractive food-related thoughts. This could be tested by additionally comparing the Market Map to the original Zoo Map, where food would not be a distraction.

With two control groups, we examined whether the deficit was linked to PWS specifically or could be attributed to IQ level and were able to highlight the global influence of intellectual level on planning performance, which can translate in daily behavioural problems. The four IQ indexes were related to all planning measures in the PWS group revealing the involvement of distinct cognitive mechanisms underlying planning activities. In adults with PWS, working memory, processing speed, verbal comprehension and perceptual reasoning appeared to predict performance of adults with PWS in the Market Map. Conversely, none of the four indexes were related to any planning measures in the TD group. In the ID group, only the processing speed and the perceptual reasoning were linked to the measures. As such, the IQ indexes can help to better understand the processes underlying performance in line with the idea that planning is not a unitary construct (Burgess, Simons, Coates, & Channon, 2005; Burgess, Veitch, de Lacy Costello, & Shallice, 2000; Rose et al., 2015). This study also provides important information for adapting medical care and cognitive intervention. The particularly strong involvement of working memory capacity in in PWS adults' performance invites to consider the implementation of rehearsal strategies while focusing on task execution during rehabilitation activities. The impact of verbal comprehension on planning performance also teaches us that any task that adults with PWS may plan must be extensively described beforehand, but also shows that even though the task is seemingly understood (participants were asked to repeat the instructions aloud before the beginning of the task to ensure comprehension), comprehension problems may still occur and impact task execution, possibly due to difficulties in suppressing distracting information. Therefore, in cognitive rehabilitation, special attention should be placed on the description and the comprehension of the rehabilitation programme's aims, goals and procedures in PWS adults. Because they often failed to correct themselves during the present task, cognitive rehabilitation should insist on strategies such as the monitoring of errors to ensure a better realisation of everyday tasks. Finally, longer planning and execution times found in the present study in adults with PWS teaches us to tailor the procedures of rehabilitation programmes at a slow pace, taking fatigability into account.

Author contributions

All authors have contributed to, seen, and approved of the manuscript and agree to the order of authors as listed on the title page.

Data availability statement

The data that support the findings of this study are available on request from the corresponding author. The data are not publicly available due to privacy or ethical restrictions.

Declaration of Competing Interest

The authors report no declarations of interest.

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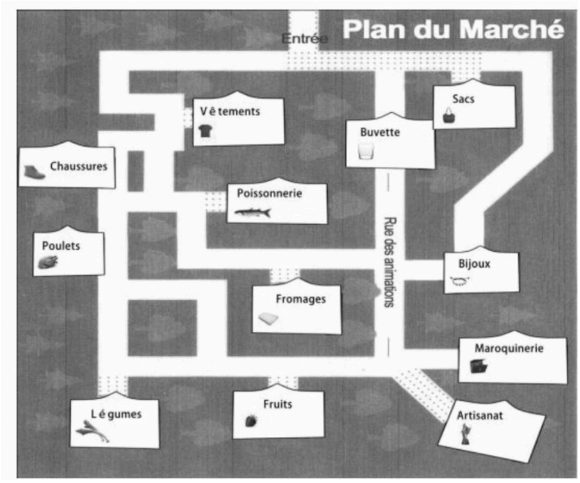
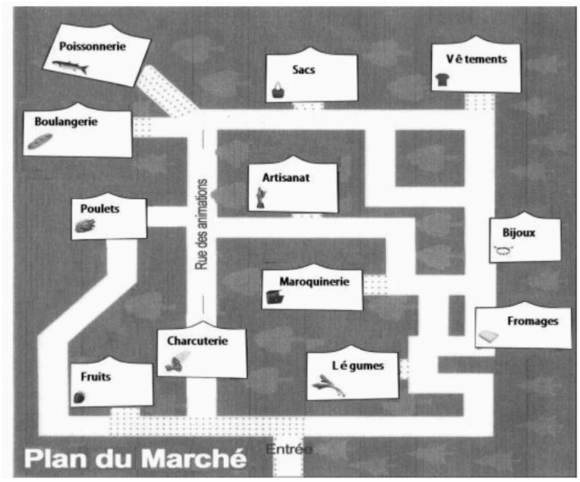
Appendix A. The Market Map

The Market Map is a slightly modified version derived from a valid standardized subtest (the Zoo Map) of the Behavioural Assessment of Dysexecutive Syndrome battery (Wilson et al., 1996). In the Market Map, some of the original location names were changed by food-related shops names. The original Zoo Map task itself does not allow a proper internal validity assessment on its own due to a single raw score and items of different metrics such as time and errors (contrasting with the whole BADS where various subtest raw scores based on the same metric are used to compute internal validity), however, a particular attention was paid to its construct validity. In support of its high construct validity, a) the task indeed yield different results across groups and conditions confirming its sensitivity with regard to planning impairments (mainly between ID and TD groups) and b) regarding the high correlations reported in the literature between planning tasks and IQ, we have compare the correlation obtained between the FSIQ and the present market map's raw score ($r = 0.68$) to the one obtained in Chevalère et al. (2013) ($r = 0.62$) between FSIQ and the original Zoo map's raw score as well as to the one obtained between the Commissions test (Martin, 1972) and FSIQ in Chevalère et al. (2015) ($r = 0.64$). We can see that the links between IQ and raw planning scores of these tasks are highly similar.

"Non-food condition – top entry"



"Food condition – bottom entry"



Note. The translations of the terms used in the Market Test are as follow: "artisanat" = crafts, "bijoux" = jewelry, "buvette" = refreshment bar, "chaussures" = shoe, "fromages" = cheese, "légumes" = vegetable, "maroquinerie" = leather goods, "poissonnerie" = fishmonger's, "poulets" = chicken, "sacs" = bag, "vêtements" = clothing.

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