PRADER-WILLI SYNDROME AND JIGSAW PUZZLES:
PUTTING THE PIECES TOGETHER

By

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To my family…

For everything I am

For everything I will be

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TABLE OF CONTENTS

LIST OF FIGURES

Figure Page

LIST OF TABLES

CHAPTER I

LITERATURE REVIEW

Overview

Humans rely heavily on our ability to remember and navigate through our environment. We would not be able to find our car in a parking lot or even locate the grocery store without the use of landmarks and an understanding of the space we live in. Without the use of concrete representations of space such as maps, we might waste our vacation at Disney World aimlessly wandering the park. Poor visual-spatial ability could keep us from participating in popular activities such as playing baseball, riding a bike, or building a jigsaw puzzle. These examples may seem like relatively trivial problems, but imagine if a soldier misinterpreted a map and accidentally led a unit into enemy territory. Imagine if our visual-spatial processing was not fast or accurate enough to correctly react to an erratic driver on the highway. Due to the importance of visual-spatial processing in our daily lives, and some evidence that males perform better on visual-spatial tests than females, cognitive psychologists have sought to discover when and how various visual and spatial abilities emerge.

One approach to studying the mechanisms of typical development is to focus on people, or groups of people, whose development follows an atypical developmental trajectory. This approach has been utilized in the visual-spatial field through studies of individuals with Williams syndrome, who show relative weakness in visual-spatial abilities. On the other end of the visual-spatial spectrum, a small body of research on individuals with Prader-Willi syndrome (PWS) indicates that they are particularly strong at putting together jigsaw puzzles, a task which would appear to require a number of visual-spatial abilities. Additionally, there are different genetic subtypes of the disorder and members of each subtype exhibit differences in a number of abilities (Roof, Stone, MacLean, Feurer, Thompson, & Butler, 2000; Dykens, 2002). Because of our focus on a number of these strengths and weaknesses, this examination of visual-spatial ability in individuals with PWS is an example of research aimed at creating etiology specific cognitive profiles as a step toward investigating developmental processes (see Dykens, Hodapp, & Finucane, 2000 for a review).

A major impetus for the conception of the research reported here was the incredible puzzle assembly of individuals with PWS observed by Dykens (2002). In her study, a group of individuals with PWS assembled almost three times the number of pieces as typically developing chronological-age matched controls, with 71% of the PWS group earning puzzle scores above the typical group's mean score. Observation of some of the more competent puzzle solvers revealed apparent mental rotation of pieces (e.g. they often picked the pieces up and rotated them as they carried them toward their final location) and possibly a strong ability in perceiving lines and curves (e.g. they seemed more accurate in judging the shape of pieces and therefore tried to force pieces together less often than the typically developing controls), skills that would result in the few mistakes and increased building speed observed by Dykens.

These findings related to puzzles motivated the research reported here, which used a visual-spatial battery to attempt to uncover what cognitive strengths are leading to the incredible performance of individuals with PWS on jigsaw puzzles. A battery of visual-spatial tasks was given to a group of individuals with PWS and a group of typically developing (TD) mental-age matched controls. These tasks were designed to test a broad spectrum of spatial abilities at varying levels of analytical and perceptual complexity. Parts of the battery required mental representation and manipulation of the test stimuli and other parts of the battery required the use of concrete representations of space such as diagrams and maps (i.e. more complex visual-spatial abilities). In the absence of a body of literature regarding the processes involved in assembling jigsaw puzzles, it was necessary that a wide range of tasks be selected and that more specific investigations be reserved for future studies.

Because research on jigsaw puzzles is nearly non-existent, one aim of this study was to investigate the processes by which puzzles are assembled. Other major aims of the study were to add to the very modest amount of research involving visual-spatial abilities in PWS phenotypes (Dykens, 2002) and to contribute to the existing body of visual-spatial literature; goals that could have practical uses for both groups studied. Clarifying the cognitive strengths and weaknesses of the group with PWS could have practical applications in educational settings and potentially open up new areas of employment and opportunity. Understanding normal visual-spatial development could also have implications for education, especially in the design of programs to narrow the gender gap that is evident in some spatial abilities.

Visual-Spatial Cognition

The study of visual-spatial processing in the typically developing population has a long and sometimes contentious history. In 1985, Linn and Petersen carried out a meta-analysis of

172 studies of visual-spatial cognition to summarize what was known about sex differences. Using the Hedges (1982a, 1982b) meta-analytic technique, they tested the studies they selected to find homogeneity of effect sizes (an indication that two tasks are measuring the same thing). If an acceptable level of homogeneity was not found, they divided the studies into smaller groups until they located homogeneous or nearly homogeneous groups of studies. Using this procedure they identified three categories of spatial ability that they claimed represented the skills on which all spatial abilities are built: *spatial perception*, *mental rotation*, and *spatial visualization.* Although there are concerns with any meta-analysis procedure because it is difficult to combine studies that are not exact replications, there is general agreement within the field that these categories encompass a major set of visual-spatial abilities (Liben et al., 2002; Scali, Brownlow, & Hicks, 2000; Voyer, Voyer, & Bryden, 1995).

Spatial perception involves determining a spatial relationship using a judgment based on awareness of the orientation of one's own body. In spatial perception tasks, this judgment ordinarily requires the participant to ignore some type of distracting cues such as a rotated frame of reference. The Rod and Frame Test (RFT--Witkin, 1949), possibly the most well known spatial perception task, requires participants to place a rod vertically within a frame that is rotated to varying degrees. Because the rod and frame are the only things visible to participants, they must ignore the tilted frame while using the effect of gravity on their body to determine when the rod is positioned vertically. The measure of spatial perception that I used was the Water Level Task (Piaget & Inhelder, 1956), in which participants must identify a horizontal water level while ignoring the angle at which a water bottle is tilted. The Water Level Task is described in greater detail in the measures section.

The second spatial ability that Linn and Petersen (1985) proposed, *mental rotation*, is a skill that allows individuals to mentally spin or "walk around" an object. A number of tasks have been created to measure a person's ability to mentally rotate objects. Shepard and Metzler (1971) devised individually administered tests in which reaction times could be recorded. They reasoned that mental rotation was analogous to physical rotation and that the farther an object needed to be rotated, the longer it would take to respond. In support of this hypothesis, Cooper (1982) found evidence that reaction time in adults is a linear function of angular rotation.

In an effort to test young children's awareness of their mental activity, Estes (1994; 1998) developed a computer based mental rotation task in which children needed to judge if two pictures of monkeys on the computer screen were holding up the same or different arms. Across trials, one monkey was rotated to varying degrees. Reaction times were recorded for each trial. Estes found that 15 of 20 six year-olds and 18 of 20 adults exhibited the linear relationship between degree of rotation and reaction time hypothesized by Shepard and Metzler. Of interest, most of these individuals reported mentally turning the rotated monkey in making their judgment. Those who did not allude to mental rotation took equally long to judge regardless of degree of rotation involved. This experiment provides more support for Shepard and Metzler's (1971) hypothesis that mental rotation is analogous to physical rotation. Because the pattern of results observed in previous studies with Estes' task indicates that it is a valid measure of mental rotation in adults and children, I adapted the task for use in this study.

Spatial visualization, Linn and Petersen's (1985) final category of spatial ability, is associated with complicated multi-step operations on spatial information. A task measuring spatial visualization may have components of spatial perception and mental rotation, but is differentiated by having multiple possible solution strategies. Embedded figures tasks, paper folding, paper form board, and block design are all examples of spatial visualization tasks (e.g. Witkin, 1950; Likert & Quasha, 1937). In many ways, spatial visualization is a catch-all category that encompasses tasks that do not fit into the other two categories of spatial ability. A subset of items from the Motor-Free Visual Perception Test (described in the methods section) was used as a measure of spatial visualization.

Tests measuring these three areas of spatial ability rely more heavily on lower-level processing abilities than on higher order function (e.g. planning) or abilities outside the visual-spatial realm (e.g. motor functioning and analytical ability). For example, in the Water Level Task, being able to tell if the water is horizontal relies on the participant making a judgment based on the orientation of their body. Gravity gives the participant the ability to perceive horizontal and they must then ignore the tilted lines of the bottle which serve as distracting input. Thus, solving the problem relies on a perceptual "feeling" about what is correct (Linn & Petersen, 1985). Similarly, *mental rotation* tasks must be solved by mentally representing the object and rotating it to match a referent.

In the real world, adults and children use skills from these three areas of spatial ability to solve and understand various complex problems. A number of my tasks are designed to simulate or directly copy challenging activities in which children and adults regularly engage. We all know how difficult it can be to navigate using the map of an unfamiliar city. In one of my tasks, children were asked to find, on a map, a location that was designated on a small 3-D terrain.

Another task required participants to use a 2-dimensional diagram to construct a 3-dimensional object (a simple Lego model). The skills required to use assembly diagrams do not come easily to many individuals. Children often struggle when using building sets or putting together a Barbie house. Any adult who has ever assembled a piece of furniture or a bicycle from its accompanying diagram also knows how frustrating it can be to use spatial representations.

The more complex visual-spatial task of using spatial representations may rest on a combination of Linn and Petersen's three visual-spatial abilities and may also require other forms of cognitive processing. For instance, poor mental rotation ability may affect an individual's ability to use a map of a mall to locate a store, especially if the map is not oriented with the mall. However, other skills outside of visual-spatial processing, such as motor functioning or the ability to plan a route, could have a strong effect on a person's ability to use the same spatial representation.

Prader-Willi Syndrome

Prader-Willi syndrome is a genetic disorder characterized by mild to moderate mental retardation, hyperphagia (over-eating), and a profile of physical and behavioral features. Physical characteristics include short stature, small hands and feet, thick saliva, and speech articulation difficulties (Gabel et al., 1986). A number of affected individuals exhibit obsessive/compulsive behavior, including skin picking, verbal perseveration on a narrow range of topics, impulsivity, and a preoccupation with exactness (Wigren & Hansen, 2003; State, Dykens, Rosner, Martin, & King, 1999).

About 1 in 10,000 to 1 in 15,000 people are born with PWS. Among a number of genetic causes for the syndrome, the two most common are a deletion of a small area (q11-13) on the long arm of the father's copy of chromosome 15 and maternal uniparental disomy (UPD), in which an individual inherits two chromosome 15s from the mother and none from the father (Dykens et al., 2000). In all cases, the syndrome results from an inactive or missing contribution of the father's genetic material at 15q11-q13 (an example of *genomic imprinting*, the process by which a gene is expressed differently depending on the sex of the parent from which it comes --Goos & Silverman, 2001).

In terms of intelligence, individuals with PWS generally have higher IQ's (average = 70) than do individuals with other genetic forms of mental retardation. About 5% of individuals with PWS have IQ scores that are considered average in normal populations (85 and above). A number of studies provide evidence that short-term memory is particularly weak in individuals with PWS and that long-term retrieval is relatively strong (Warren & Hunt, 1981; Conners, Rosenquist, Atwell, & Klinger, 2000). Dykens, Hodapp, Walsh, and Nash (1992) found a significant weakness on the sequential processing subscale of the Kaufman Assessment Battery for Children (K-ABC) and also found a relative weakness on the spatial memory subtest of the simultaneous processing subscale.

In the clinical literature, individuals with PWS are reported to assemble a lot of jigsaw puzzles, and an affinity for jigsaw puzzles has actually become a supportive diagnostic criterion (Dykens et al., 2000). Clinical reports also suggest that individuals with PWS enjoy word search puzzles; there are anecdotal reports of this affinity even in some individuals who cannot read.

Although few studies focus on visual-spatial processing in individuals with PWS, the small amount of extant research indicates that certain domains of visual-spatial ability may be relative strengths compared to other domains of intelligence (e.g. Gabel et al., 1986; Taylor, 1988). Evidence most suggestive of relatively spared visual-spatial abilities come from a study by Dykens (2002) investigating the ability of individuals with PWS to assemble jigsaw puzzles. In the study, a group of individuals with PWS assembled an average of 28.1 puzzle pieces in 6 minutes, whereas chronological-age (CA) matched typically developing children placed an average of only 10.7 pieces. Dykens (2002) also found that her sample of individuals with PWS solved a word search puzzle almost as well as her CA matched typically developing children. A few standardized visual-spatial tasks have also been administered to individuals with PWS. On some of the tasks, individuals with PWS have outperformed their chronological-age matched TD controls (Gabel et al., 1986) and on others they have fallen behind CA matched controls but still outperformed mixed etiology IQ matched controls (Dykens, 2002). These pieces of evidence have been used to suggest that individuals with PWS may have relatively strong cognitive abilities in one or all of the visual-spatial areas. However, there are a number of issues with this conclusion. Most of the visual-spatial tests that have been administered resemble jigsaw puzzles (e.g. Object Assembly from the WISC-III--Wechsler, 1991; Triangles from the K-ABC--Kaufman & Kaufman, 1983). Additionally, many of the tasks are visual-motor tasks (in which manipulations of the stimuli provide the basis for assessment; e.g. VMI--Beery, 1997; Grooved Pegboard--Knights & Moule, 1980; Trailmaking Test--Reitan, 1958) as opposed to tasks that would be considered visual-spatial (in which the emphasis is on perceiving the stimuli and/or manipulating mental representations of them).

There is evidence that the different genetic subtypes of the disorder may be related to different cognitive and behavioral profiles. Whittington et al. (2004) found that a group with uniparental disomy had higher verbal abilities than a group with the deletion form of PWS. Additionally, it has been found that individuals with the deletion form of PWS tend to have more maladaptive behaviors (Dykens, Cassidy, & King, 1999). In addition to the deletion/uniparental disomy distinction, there is evidence that the various deletion types also have different phenotypes; individuals with larger deletions often have more compulsive behaviors and greater visual perception impairments (Butler, Bittel, Kibiryeva, Talebizadeh, & Thompson, 2004). Unfortunately, in the research reported here, the number of individuals in each subtype was insufficient to formally analyze subtype effects, but as will be discussed later, most of the differences between subtypes were in the expected direction. As more studies finding between-subtype differences emerge, it becomes more important to find genetic, neurological, and/or environmental bases for an advanced skill in individuals who otherwise are cognitively impaired.

CHAPTER II

METHOD

Participants

Two groups participated: 18 individuals with Prader-Willi syndrome (12 males and 6 females; CA = 19.7; *SD* = 12.2 years) and 16 typically developing individuals (8 males and 8 females; CA = 6.2; *SD* = 1.5 years), matched by mental-age (MA). The MA's of the group with PWS and the TD individuals were 7.47 (*SD* = 2.25) and 7.29 (*SD* = 2.38) years respectively. The group with PWS consisted of 2 individuals with maternal uniparental disomy and 6 individuals with paternal deletions. Lab results are still being processed for 7 individuals with unspecified deletions and 2 individuals who have been diagnosed with PWS, but for whom the subtype is unknown. Participants with PWS were recruited through local and national chapters of family support groups and local clinics. The sample was entirely European American with the exception of one African American male. The TD participants were recruited from a local database compiled from state birth records and by flyers distributed through after-school programs, sport programs, and various businesses frequented by children and parents. This sample was also largely European American, with the exceptions being 2 African American females and 1 Australian female.

Procedure

All participants were tested by a male researcher in a lab at Vanderbilt University. Participants were asked to complete a brief IQ test and a series of visual-spatial tasks (outlined below) during a 90 minute session. Participants received a gift certificate as a token of appreciation. The sessions were video taped, and the tapes were later coded by an assistant to look for strategies and other interesting behaviors.

Due to concerns with the length of the visual-spatial battery and participants' ability to maintain concentration throughout, tasks were presented in a designated order that interspersed short, hands-on tasks with lengthy ones that required verbal responses (such as the IQ and Motor- Free Visual Perception Test). The presentation order was: Kaufman Brief Intelligence Test (K-BIT), Placement Tasks, Water Level Task, Jigsaw Puzzles, Mental Rotation Task, Motor-Free Visual Perception Test 3rd Edition (selected items), and Lego Building. Concern over possible order effects was outweighed by the necessity to ensure that motivation stayed high enough for all subjects to complete the battery. Where possible or necessary, multiple presentation orders were used within the separate tasks.

Measures, Materials, and Scoring

The following battery of tasks was designed to asses various visual-spatial abilities and includes standardized measures, measures adapted from previous research, and newly designed tasks.

IQ Measure

KBIT

The Kaufman Brief Intelligence Test (K-BIT; Kaufman & Kaufman, 1990) allowed for MA matching between the MR group and the TD group. The K-BIT is a short-form IQ test, consisting of two distinct subscales (Verbal and Matrices). The verbal subscale is made up of two sections called Expressive Vocabulary and Definitions. The test gives standard scores for each domain and overall, and can be used for typically developing participants from ages 4 to 90. The K-BIT, in addition to being used in typically developing populations, has been successfully used in recent studies of children and adults with mental retardation (Dykens, Rosner, & Ly, 2001).

Puzzles

Jigsaw/Face Puzzle

To replicate an earlier study by Dykens (2002), I tested both groups on their ability to assemble jigsaw puzzles using a traditional, 30-piece jigsaw puzzle of a young woman's smiling face that was created for this study (see Figure 1a). Participants had 5 minutes to assemble as many pieces as possible and their score was the number of correctly attached pieces. A picture of what the jigsaw puzzle looks like when completed (similar to a picture on the box-top of a traditional puzzle) was provided to assist in assembling the puzzle. Henceforth I will refer to this picture as the box-top picture or box-top representation.

Jigsaw/Blank Puzzle

To explore strategy use and to determine which pieces of information the groups were most focused on, participants were also asked to solve two unusual puzzles. One was a blank, 30-piece jigsaw puzzle (see Figure 1b) that could only be solved using the shapes of the individual pieces. Because it was blank, no box-top picture was provided for assistance on this puzzle.

Non-Interlocking/Face Puzzle

The final puzzle was a 17-piece non-interlocking puzzle that contained the same face as the interlocking/face puzzle (see Figure 1c). Correctly assembling this puzzle relied less on the shape of the puzzle pieces (any two pieces could be placed together) and more on attending to the picture of the face.

(a)Interlocking Face (b)Blank Interlocking (c)Non-interlocking Face

Figure 1 --Puzzle Pictures

Measures of the 3 Areas of Visual-Spatial Ability

Water Level Task (Spatial Perception)

A multiple choice version of Piaget and Inhelder's (1956) Water Level Task was employed as a measure of spatial perception (for a thorough review of the Water Level Task, see Vasta and Liben, 1996). In my version, for each trial, the subjects saw drawings of five bottles tilted at the same angle (see Figure 2). Within each bottle there was a line that signified the top of the water. One line was parallel to the tabletop on which the bottles rested in the drawing (correct choice) and the rest were at random angles to the tabletop (incorrect choices). Subjects were asked to point to the one that "shows where the top of the water would be."

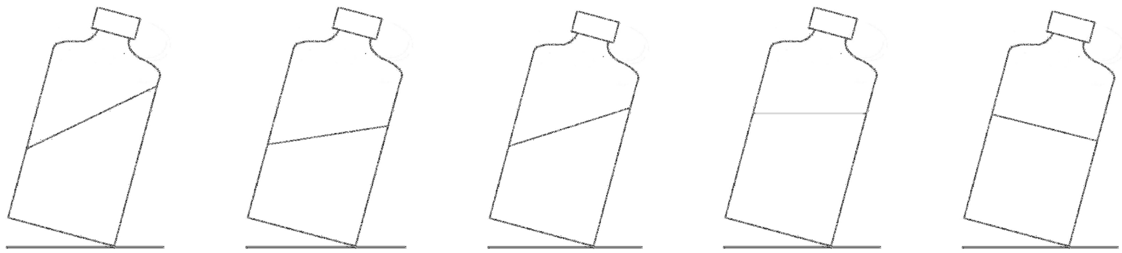


Figure 2 --Water Level Task Example

A multiple choice format was used, as opposed to a version in which the individual must draw in the water lines, to eliminate any requirements of motor coordination. Each subject was given four trials, with all the bottles tilted at the same angle (15º, 30º, 45º, or 60º) on each of the four trials. Presentation order was counterbalanced across participants. Answers were scored on a scale from correct (4 points) to the angle farthest from correct (0 points), summing to a total possible of 16 points. Because it was expected to be a common incorrect answer (e.g. Piaget & Inhelder, 1956), the researcher also counted the number of bottles chosen in which the water level was parallel to the bottom of the bottle (the last bottle in Figure 2).

Mental Rotation Task (Mental Rotation)

As a measure of mental rotation ability, a version of Estes' (1998) computer based mental rotation task was employed. The program displays two monkeys on a computer screen and requires the participants to judge, by pressing 1 of 2 buttons on the keyboard, whether the monkeys are holding up the same or different arms. The monkey on the left side of the screen always faced forward and was oriented at 0 degrees (upright). The monkey on the right side of the screen is rotated from 0 to 180 degrees from upright in 45 degree increments (Figure 3). The monkey on the right side can also be facing in the opposite direction (Figure 3--bottom panel), requiring rotation in both planes (a variation not used by Estes). The 3-D trials were introduced to increase the difficulty of the task, in order to capture the full spectrum of ability in both groups. Participants received 30 trials (3 blocks of 10 trials), with backward facing monkeys appearing on 10 of the 30 trials. The presentation of stimuli was in a pseudo-random order that prevented stimuli with the same degree of rotation from appearing on consecutive trials.

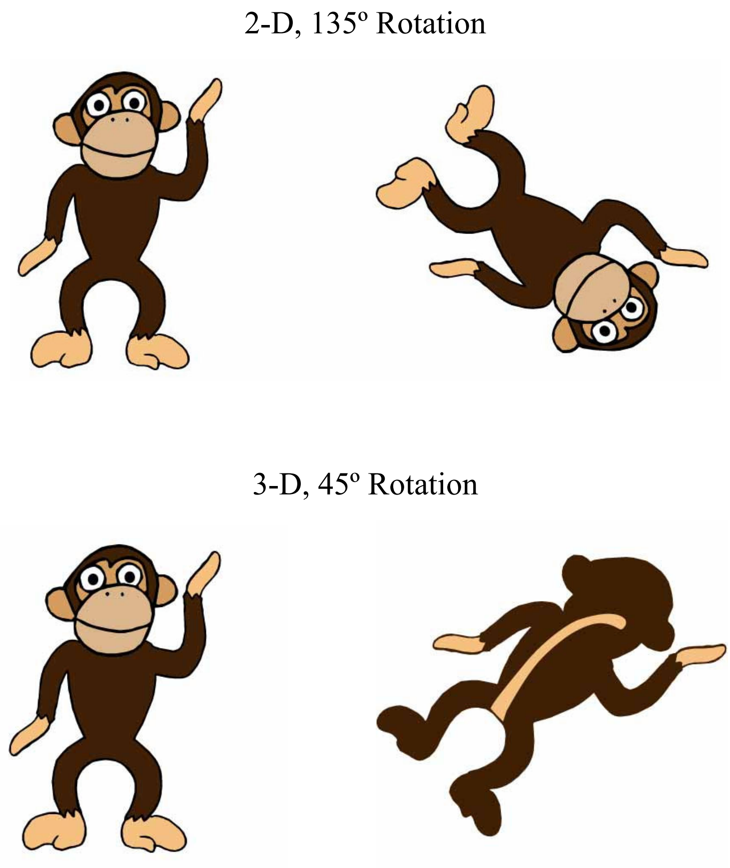


Figure 3 --Mental Rotation Examples

Participants were taught the object of the game during 10 orientation trials. These trials involved different monkey orientations and required both “same” and “different” answers. The orientation was repeated one time if the participant did not appear to understand what was required of them. In the 3 instances in which the orientation was repeated, the repetition was not effective and the subjects did not complete the task.

For each trial, the computer kept track of which monkeys were displayed, whether the answer the participant gave was correct or incorrect, and how long it took to answer. Participants were not told that speed was an important part of the task because of concerns about impulsivity in the group with PWS.

Motor-Free Visual Perception Test -Third Edition (Spatial Visualization)

Due to the length of the Motor-Free Visual Perception Test (third edition, Colarusso & Hammill, 2003), items were selected that required finding a target shape hidden within a larger array. These embedded figure items (numbers 4-8,10,12,13, and 51-55) were chosen for their similarity to other embedded figures tasks (e.g. Witkin, 1950), and were used as a test of *spatial visualization*. As a result of selecting only a portion of the test items, the norms associated with the test were uninformative, but comparison across groups was still possible.

To get a measure of how well each group could find very small differences between figures, stimuli from two other sections of the test were also presented. One section required the participants to find an incomplete figure that (if completed) would match a presented figure (even numbered items from #22 to #34). The other section required the participant to find the one figure out of four possible choices that was different from the others (odd numbered items from #35 to #45). The ability to notice small differences in the shapes of puzzle pieces could be very helpful for being a successful puzzle builder.

Measures of Complex Visual-Spatial Abilities

Placement Tasks

In addition to being interested in the participants' skill at completing tasks measuring the three areas of visual-spatial ability outlined by Linn & Petersen, I wanted to test their ability to use representational artifacts like maps and diagrams. These complex abilities are important in many real world situations. Knowing whether individuals with PWS excel at them could have many practical applications. To study the participants' ability to understand and use maps, a modified version of Laurendeau and Pinard's (1970) placement task was utilized, employing a portable 3-D Styrofoam "terrain" (50 cm x 50 cm) as well as a map (27 cm x 27 cm; see Figure 4). To begin the task, a short orientation and explanation was given to ensure that the participant was familiar with the map key and the symbols appearing on the map.

Next, participants were told a story about a man digging for buried treasure in the town. A Lego man (the treasure hunter) was moved to the different placement locations on the terrain and participants were told to, "draw an *X* on the map where the Lego man is digging." The experimenter marked the position, on each of 4 trials, using colored stickers. Then the experimenter replaced the terrain with a second map and repeated another 4 trials (map-to-map).

The two conditions were used to examine components of spatial representation ability. Success in the map-to-map case requires simply mapping between two identical arrays and does not require the participant to notice a representational relationship. In contrast, participants in the terrain task, created for this study, must notice the 1-to-1 correspondence of the map to a distinctly *different* (3-D) referent, much as real world map use requires a person to match their 3-D surroundings to locations on a 2-D map.

Four placement locations were chosen for the tasks in order to create a continuum of difficulty, depending on the presence or absence of distinctive landmarks (see Figure 4). Placements were scored using a multi-step coding scheme that reflected how close the mark was to the target location (maximum per trial = 4).

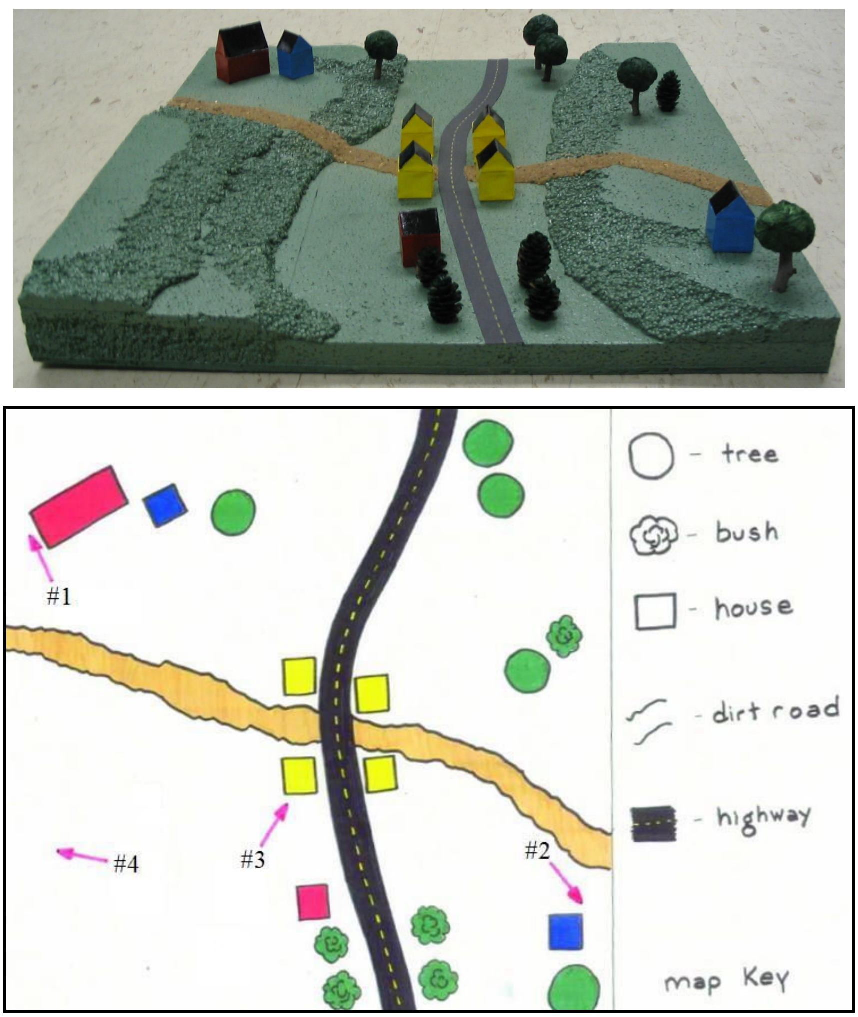


Figure 4 --Pictures of Terrain and Map with Marked Placement Locations

Lego Building

Another measure of the participant's ability to use and understand concrete representations of space involved a diagram and a set of 17 Lego blocks. Participants were given a diagram that showed the steps required to assemble a duck figure from Legos. The children were simply given the Lego pieces and the diagram, and told to build the duck using the steps shown in the diagram. Participants were given 5 minutes to get as far as they could on the task. If they said that they were finished before the end of the 5 minute period, the experimenter recorded the time at which the participant claimed to be done. Each block placed by the participant was assigned a point for being in the correct row and a point for being in the correct position within the row (determined in relation to the row below the piece in question). With 17 blocks in the model, the maximum potential score was 34 points.

CHAPTER III

RESULTS

Preliminary Analyses

Every effort was made to match mental ages (MA's) across groups on a child-by-child basis using KBIT scores, resulting in mean ages and variances that were statistically equivalent. To control for any potential remaining MA differences in the two groups, I performed ANCOVA's, covarying for mental age, to test between group differences on all of the measures. Changes in the scores were minor and the statistical significance of the findings did not change. The only significant gender differences involved the number of looks at the box-top picture in both groups (females looked more) and scores on the water level task in the TD group (males scored higher); these are presented in appendix A.

Puzzles

Jigsaw and Non-Interlocking Puzzle Scores

Participants with Prader-Willi syndrome assembled an average of 15.65 (*SD* = 10.87) pieces of the 30-piece jigsaw/face in 5 minutes compared to the 11.88 pieces (*SD* = 7.74) assembled by the MA matched controls. This difference was not significant at the 0.05 level, but there was a medium effect size (*ES*) of -0.39. Five of the individuals with PWS (but none of the TD children) finished the puzzle before the 5 minute mark, creating a possible ceiling effect.

An interesting pattern emerged for the jigsaw/blank and non-interlocking/face puzzle. The individuals with PWS outperformed the MA-matched group on the jigsaw/blank puzzle, assembling 10.44 (*SD* = 8.40) versus 4.56 (*SD* = 4.41) pieces out of 30 respectively, a difference that was significant by t-test, $t(32) = 2.51$, $p = 0.031$, *ES* = -0.84. Scores on the non-interlocking/face puzzle were similar in both groups (TD = 7.75 pieces, *SD* = 5.52; PWS = 6.56 pieces, *SD* = 3.85). This difference was not significant (*ES* = 0.25). Puzzle means are displayed in Figure 5.

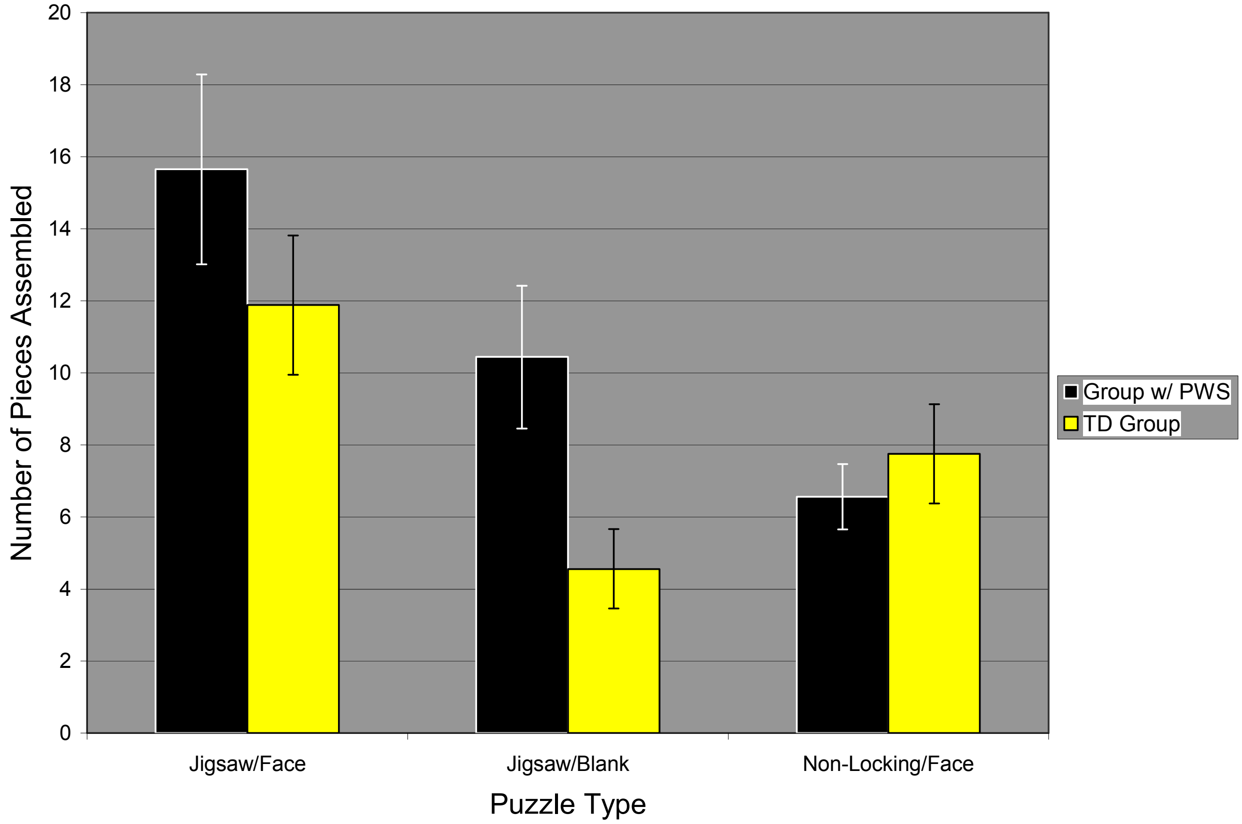


Figure 5 --Puzzle Performance

Correlations between scores on the 3 puzzle types revealed different patterns for the two groups (see Table 1). In the group with PWS (but not the TD group), performance on the two *jigsaw* puzzles (face and blank) was highly correlated (Spearman's *rho* = 0.90, $p < 0.001$). In the TD group, the highest correlation (Spearman's *rho* = 0.68, $p = 0.004$) was between scores on the two face puzzles (non-locking and jigsaw), which both contained a *picture*. These scores were similarly correlated in the individuals with PWS. Finally, scores on the jigsaw/blank and the non-locking/face puzzles were significantly correlated in the TD group but not the group with PWS. This pattern of results suggests that the group with PWS relied primarily on shape information in constructing the puzzles, whereas the TD group seemed to favor information derived from the picture appearing on the puzzle.

Table 1 --Puzzle Correlations

		Jigsaw/ Face	Jigsaw/ Blank	Non-Lock/ Face
Jigsaw/Face	R Sig. N		**.900**** **.000** **17**	**.537*** **.026** **17**
Jigsaw/Blank	R Sig. N	.245 .360 16		.313 .205 18
Non-Lock/ Face	R Sig. N	**.677**** **.004** **16**	**.579*** **.019** **16**	

All correlations are non-parametric Spearman Correlations
* p < 0.05 (2-tailed)
**p < 0.01 (2-tailed)

Key

Key
Group w/ PWS
TD Group

Video Coding and Strategies

Dykens (2002) reported that participants with PWS were more likely to start with the edge pieces of jigsaw puzzles. To determine if my participants with PWS used that strategy, the number of edge pieces out of the first five pieces that each participant attempted to put together were recorded. On both puzzles, participants with PWS were more likely than TD participants to start with the edge pieces. The use of edge pieces increased for both groups on the blank jigsaw puzzle, where shape was the only information provided by the pieces. Table 2, shown below, gives the results of the edge strategy analyses.

Table 2 --The Use of Edge Pieces at the Onset of Puzzle Building

		N	Mean (of 5)	Standard Deviation	Significance
Jigsaw/Face Puzzle	Group w/ PWS	15	3.13	2.15	$p < 0.05$
	TD Group	14	1.50	2.75	
Jigsaw/Blank Puzzle	Group w/ PWS	15	4.57	2.16	$p < 0.05$
	TD Group	14	3.73	1.87	

From the videotapes of participants assembling the puzzles, we coded the number of looks they directed toward the box-top picture. On the jigsaw/face puzzle, there was a non-significant trend for the group with PWS to look less at the box-top picture ($M = 1.33$ looks, $SD = 2.15$) than the TD group ($M = 3.00$ looks, $SD = 2.75$), $t(27) = -1.83$, $p = 0.079$, $ES = 0.68$. When working on the non-interlocking puzzle, both groups looked at the box-top picture more often (PWS--$M = 5.73$ looks, $SD = 6.98$; TD--$M = 5.57$, $SD = 3.97$) and the groups did not differ. To examine how children chose to use the box-top picture as a strategy, I classified participants in each group who looked at the box-top picture 3 or more times as using a looking strategy. The TD group had significantly more participants who relied on the looking strategy for both the jigsaw/face puzzle (TD = 50%, PWS = 13%), $\chi^2(1) = 4.55$, $p = 0.033$, and the non-interlocking/face puzzle (TD = 65%, PWS = 40%), $\chi^2(1) = 4.41$, $p = 0.035$. Another interesting finding is that those individuals in the group with PWS who used the looking strategy for the non-locking/face puzzle (40% of the sample), looked at the box-top picture significantly more frequently than the TD participants employing this strategy (13.17 looks, $SD = 5.07$ vs. 6.68 looks, $SD = 3.71$), $F(1,25) = 4.36$, $p = 0.047$.

There are a number of possible explanations for this pattern of results. One possibility is that a deficit in short-term and/or spatial memory for the group with PWS makes the box-top picture difficult to use. This interpretation would explain why some individuals did not attempt to use a looking strategy (they already learned that it was a difficult strategy to use), while those that used the looking strategy tended to look at the picture more often than typically developing controls on the non-interlocking puzzle (i.e. when piece shape was unavailable as a clue to piece placement).

“Expert” Effects

It became apparent that the high scores of a small group of puzzle “experts” in the group with PWS were raising the means for the jigsaw/face puzzle. All of these experts completed the puzzle in less than 5 minutes, creating somewhat of a ceiling effect for that puzzle. I examined whether there were any ways in which these puzzle “experts” differed from other participants, including those from the TD group who performed well on the jigsaw/face puzzle. For these analyses, I chose the five individuals with PWS that reached ceiling on the jigsaw/face puzzle (30 pieces assembled) and the top four puzzle assemblers from the TD group (19 to 24 pieces assembled).

I performed an ANOVA to determine how the experts differed from the rest of their respective groups and from each other. The 2 (group: PWS vs. TD) x 2 (classification: expert vs. non-expert) ANOVA and post-hoc LSD tests showed that the experts with PWS significantly outscored all of the other groups on both the jigsaw/face and jigsaw/blank puzzles (see Figure 6). The jigsaw/blank puzzle was their biggest triumph: they achieved a mean of 18.60 pieces (*SD* = 0.89) out of 30. The rest of the group with PWS had the next highest mean score at 5.75 (*SD* = 5.71), which was not significantly different from the score of the other groups. Note that the within-group standard deviation for the blank puzzle is very small for the PWS experts--less than ¼ as large as the standard deviations for all of the other groups, perhaps the result of an artificial ceiling created for this puzzle when the individuals ran out of the 18 edge pieces and had to try to assemble the more difficult inside pieces.

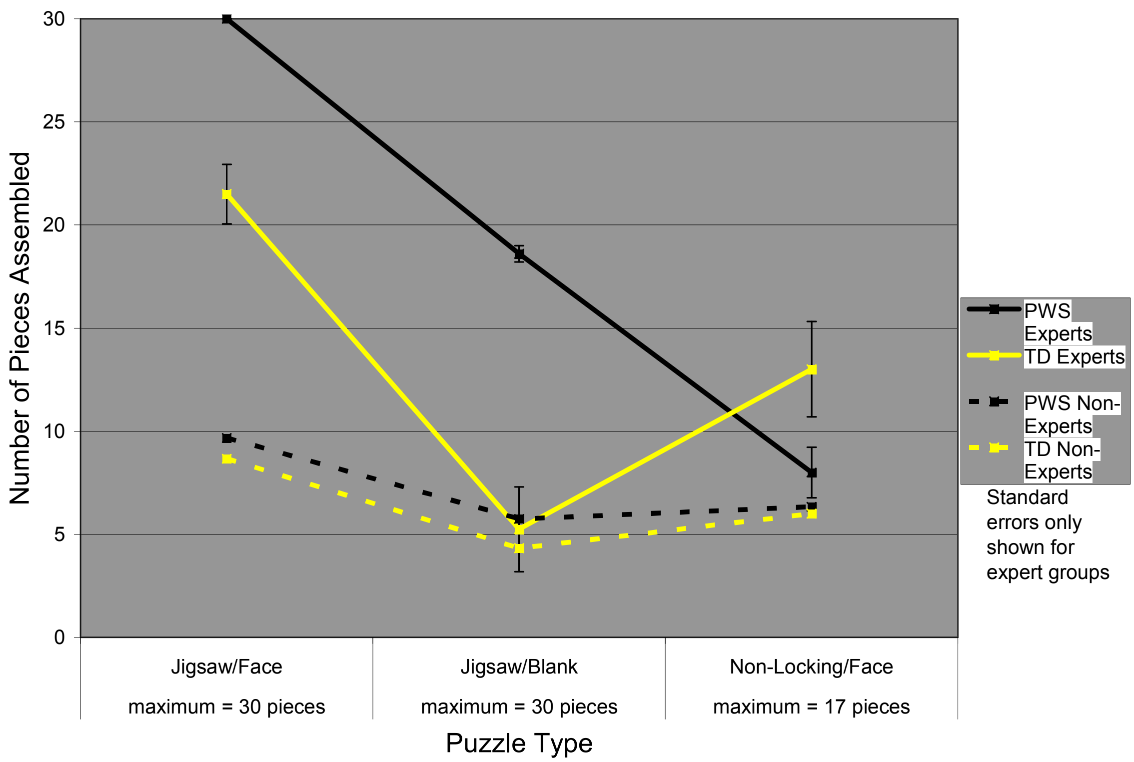


Figure 6 --Expert and Non-Expert Puzzle Scores

Although the experts with PWS significantly outscored everyone on both of the jigsaw puzzles, their performance on the non-interlocking puzzle was not significantly different from that of any of the other groups. The TD experts outscored both of the non-expert groups on this puzzle; the two expert groups' scores (TD and PWS) were statistically equivalent.

A look at the strategies observed from the video coding revealed one interesting pattern among the experts. The results related to the number of impossible connections attempted while assembling the blank jigsaw puzzle. An impossible connection was defined as any attempt to put pieces together that could not possibly go together based on the shape of the pieces (e.g. attempting to connect an inside piece adjacent to the straight side of an edge piece or trying to put two sides with male connectors together). A 2 (group) x 2 (expert classification) ANOVA followed by post-hoc LSD tests revealed that the experts with PWS attempted the fewest number of impossible connections on the blank jigsaw puzzle ($M = 0.40$, $SD = 0.89$) and the TD experts committed the greatest number ($M = 24.75$, $SD = 22.50$). This is an enormous difference, amounting to the TD experts having committed on average more than 60 times the number of mistakes as the experts with PWS. The two non-expert groups were intermediate (PWS Non-expert--$M = 12.40$, $SD = 12.51$; TD Non-expert--$M = 10.45$, $SD = 6.74$). The difference between the two expert groups was significant ($p = 0.004$). The TD experts also committed significantly more impossible attempts than the TD non-experts ($p = 0.041$). Thus, the TD children who did best on the face jigsaw puzzle apparently resorted to a random guessing strategy when confronted with a blank jigsaw puzzle.

Recall that previous evidence from Dykens' (2002) work revealed that deletion type was related to puzzle performance. In this study, all five of the PWS experts had one of the deletion subtypes: 2 had type I deletions, 1 had a type II deletion, and the final 2 individuals had unspecified deletions. The experts in both groups of participants also had somewhat higher chronological ages compared with the non-experts, though neither difference was significant. The average difference in mental age (MA) between experts and non-experts was only 3 months in the group with PWS (n.s.) but 39 months in the TD group ($p = 0.015$). Comparing the two groups of experts, the average MA of the TD experts was approximately 2 years older than that of the experts with PWS (also n.s.). Thus, the PWS experts, who were so much *more* skilled at assembling jigsaw puzzles, were somewhat *less* mentally advanced than the experts in the TD group.

The 3 Areas of Visual-Spatial Ability

Scores for the TD group were either higher or equivalent to the group with PWS on every measure of spatial ability, including the more complex tasks requiring the use of spatial representations. A brief task-by-task analysis for the visual-spatial measures follows.

Water Level Task (Spatial Perception)

On the Water Level Task, both groups scored slightly below a chance score of 8.00 (PWS $M = 7.38$, $SD = 3.54$; TD $M = 6.44$, $SD = 4.44$). The below chance scores apparently resulted from some children systematically choosing bottles with the water level parallel to the bottom of the bottle (parallel bottles), a choice that results in the lowest score for most of the angle presentations. TD children tended to either systematically choose incorrectly (8 children chose 3 or 4 parallel bottles across 4 trials), or to avoid parallel bottles and perform very well on the task. The pattern of choices in the group with PWS is more consistent with random guessing; the greatest number of participants (9 out of 16) chose one incorrect bottle (the number expected by chance).

At first glance, the data for the TD group appear to fit the expected developmental pattern that Piaget described for the water level task (Vasta & Liben, 1996), in which younger children begin by placing the water level parallel to the bottom of the bottle and slowly revise their thinking until they are able to answer correctly. However, although the highest scores in the TD group were all recorded by older children, and mental age and Water Level Task scores were significantly correlated (Spearman's $rho = 0.521$, $p = 0.039$), chronological age and Water Level Task scores were not.

Mental Rotation Task (Mental Rotation)

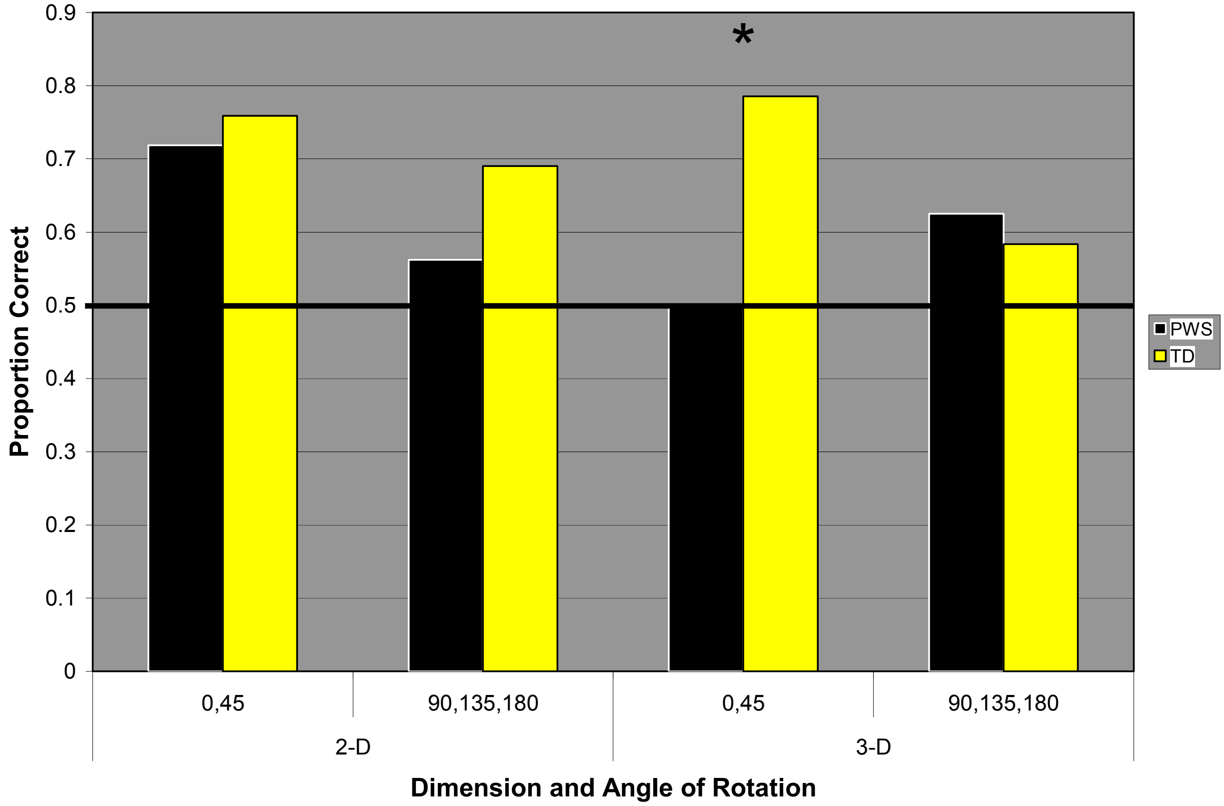
On the computerized Mental Rotation Task, average scores were computed across trials with the same rotation angle (0, 45, 90, 135, 180) and dimensional presentation (2-D--forward facing monkey; 3-D--backward facing monkey) yielding 10 scores for each participant. Scores were then compiled into small rotation angles (0, 45) and large rotation angles (90, 135, 180) within each dimension (2-D and 3-D) and each was compared to a chance level of 1.0 by t-test.

As can be seen in Table 3, the means for the TD group were above chance for all but the most difficult angles (large, 3-D rotations). In contrast, the group with PWS was above chance for only the small, 2-D rotations. Figure 7 below shows that the TD children significantly outscored the individuals with PWS for the 3-D, small rotation presentations, $t(24) = -2.32$, $p = 0.029$, *ES* = 0.91. There is also a trend in that direction for the 2-D, large rotation trials (*ES* = 0.62). An average across all angles and dimensional presentations revealed that the TD group scored slightly higher than the group with PWS (70.0% vs. 60.6% respectively), although this difference was not statistically reliable (*ES* = 0.56). Overall, the data indicate that the individuals with PWS did not exhibit an advantage in mental rotation and that mental rotation may actually be a weakness. See appendix B for more Mental Rotation Task results.

Table 3 --Mental Rotation Data

	Dimension of Rotation	Angle of Rotation	Mean Percent Correct	Standard Deviation
TD Group	2-D	0-45	**75.9%****	32.3%
	2-D	90-180	**69.1%****	21.0%
	3-D	0-45	**78.6%****	25.7%
	3-D	90-180	58.3%	26.8%
Group w/ PWS	2-D	0-45	**71.9%****	17.0%
	2-D	90-180	56.2%	20.5%
	3-D	0-45	50.0%	36.9%
	3-D	90-180	62.5%	25.7%

** Sig. different from chance (2-tailed, $p<0.01$)



* p < 0.05 (2-tailed)

Figure 7 --Mental Rotation Data

Motor-Free Visual Perception Test (Spatial Visualization)

A subset of items from the Motor-Free Visual Perception Test was used as a measure of spatial visualization. The TD participants significantly outscored the group with PWS, correctly answering an average of 16.23 (*SD* = 2.86) versus 11.91 (*SD* = 3.70) of the tests 25 questions, $t(22) = -3.23$, $p = 0.004$, yielding a very large effect size of 1.32. On one group of 6 similar items that required participants to find an incomplete figure that, if completed, would match the stimulus, scores from the two groups were not significantly different (TD--*M* = 3.46, *SD* = 1.33; PWS--*M* = 3.00, *SD* = 1.00). However, there was no evidence that individuals with PWS excelled at spatial visualization as measured by the Motor-Free Visual Perception Test items.

The 3 Areas of Visual-Spatial Ability and Puzzle Assembly Scores

Several scores on the spatial measures were significantly correlated with scores on the 3 puzzle types for the TD group (see Table 4). Most of the significant correlations for both groups involved the non-interlocking puzzle, indicating that assembly of this puzzle calls on spatial ability. Also, given that the group with PWS did not outperform the TD group on any of the spatial measures, it seems apparent that exceptional spatial ability (at least as measured by my tasks) is not the source of their ability to assemble jigsaw puzzles.

Table 4 --3 Areas of Visual-Spatial Ability/Puzzle Correlations

		TD Group			PWS Group		
		MVPT	WLT	MRT	MVPT	WLT	MRT
Jigsaw/ Face	r	**.608***	.312	.474	.208	.333	.281
	Sig.	**.028**	.240	.087	.540	.207	.403
	N	**13**	16	14	11	16	11
Jigsaw/ Blank	r	-.051	.410	.194	.309	.379	.417
	Sig.	.868	.115	.506	.355	.148	.202
	N	13	16	14	11	16	11
Non-Lock/ Face	r	**.561***	**.614***	**.760****	**.752****	.327	.198
	Sig.	**.046**	**.011**	**.002**	**.008**	.216	.560
	N	**13**	**16**	**14**	**11**	16	11

All correlations are non-parametric Spearman correlations
* $p < 0.05$ (2-tailed)
** $p < 0.01$ (2-tailed)

MVPT = Motor-Free Visual Perception Test (Spatial Visualization)
WLT = Water Level Task (Spatial Perception)
MRT = Mental Rotation Task (Mental Rotation)

Complex Visual-Spatial Abilities

There was a trend for the TD group to outperform the group with PWS on the placement task. The scores for the map-map portion of the test were 12.60 (*SD* = 3.09) versus 10.06 (*SD* = 3.94), $t(29) = -2.54$, $p = 0.057$, yielding an effect size of 0.45. The scores for the map-terrain portion of the task were 10.07 (*SD* = 3.94) and 8.19 (*SD* = 4.40), $t(29) = -1.25$, $p = 0.221$, resulting in an effect size of 0.45.

In assembling a small Lego model (Lego task), the TD group outscored the group with PWS, attaining a mean score of 17.44 (*SD* = 13.45) versus 8.33 (*SD* = 8.71) out of 34, $t(29) = -2.25$, $p = 0.033$, *ES* = 0.80. To ensure that the score differences did not result from differences in fine motor control (that is, that the TD individuals were not simply able to put more piece together in the given time), I analyzed the average number of points awarded per block (a measure of accuracy). The resulting means were 1.23 (*SD* = 0.67) points per block (out of 2) for the TD group and 0.76 (*SD* = 0.61) for the group with PWS. This relationship, approaching significance, $t(28) = -2.00$, $p = 0.055$, *ES* = 0.73, suggests that the TD group was faster and more accurate.

CHAPTER IV

DISCUSSION

The original question motivating this research was whether individuals with PWS have exceptional visual-spatial abilities that lead to their heightened skill with jigsaw puzzles. The results of this study lead to the conclusion that jigsaw puzzle skill in individuals with PWS does not stem from spatial ability, as measured by our battery. The group with PWS scored lower than, or at best equal to, the TD group on every spatial measure. Also, their scores on the puzzles were not correlated with the spatial measures (with one exception--shown in Table 4). This result was unexpected given previous research that suggested that individuals with PWS may have strong visual-spatial ability. However, this is the first study that was specifically aimed at testing visual-spatial ability in this group with traditional visual-spatial measures (i.e. tests that are not similar to puzzles and that do not require motor-based responses). Eliminating this plausible-seeming but apparently erroneous candidate explanation is an important step in figuring out factors that do underlie this phenomenon. Results and observations from the puzzle portion of the battery may provide clues to possible sources of the ability.

In Dykens' (2002) study of puzzle assembly in Prader Willi Syndrome, individuals with PWS outscored typically developing individuals of the same chronological age, assembling nearly three times the number of pieces of traditional jigsaw puzzles containing pictures (on average, 28.10 vs. 10.71 pieces). Task differences in the current study may explain why the effect sizes were smaller (mean scores = 15.65 vs. 11.88 pieces). First, participants in Dykens' study were given two different 40-piece jigsaw puzzles (puppy and pizza) in turn and had three minutes to work on each puzzle (total time = 6 minutes; total = 80 pieces). In contrast, in the current study, the participants had 5 minutes to work on one traditional (face) jigsaw puzzle (total pieces = 30). Given that the individuals with PWS appeared to use an edge-piece-first strategy in both studies, it is possible that their rate of puzzle assembly was faster at the beginning of a puzzle, when many edge pieces were available. As participants ran out of edge pieces, assembly rate would slow. If that was the case, being given two "starts" (and short sessions) with different puzzles would result in higher scores than a single "start" followed by an extended session. Also, there were fewer pieces (and edge pieces) in the puzzle used for this study; participants with PWS may have slowed down or struggled sooner as they ran out of edge pieces to assemble. Finally, having fewer overall pieces may have impacted the scores in that some participants with PWS (but no TD children) finished before the 5 minutes were up. Given more pieces to assemble during the remaining time, their elevated scores would likely have raised the mean scores of the group.

The typically developing children in the current study assembled about the same number of pieces as did the participants in the Dykens study, despite having a much lower mean chronological age (6.2 vs. 9.6 years) and less time to assemble puzzles (5 vs. 6 minutes). The use of a very familiar stimulus picture on the puzzle (a face), compared to the less familiar/more amorphous pictures on Dykens' puzzles (a puppy and a piece of pizza) may have contributed. As the participants with PWS paid less attention to the box-top picture than did TD children (the same finding as in Dykens' research), they may have missed out on clues presented by the face picture that helped the young TD children to achieve relatively high scores on the face puzzle. A final possibility is that the small sample sizes in both studies of individuals with PWS (an omnipresent difficulty in research on rare populations) simply contained different numbers of puzzle experts.

Information from previous research indicates that there are phenotypic differences between the different PWS subtypes. Individuals with UPD generally have higher IQ's than individuals with deletions and those with smaller deletions have higher IQ's than those with larger deletions. Additionally, Dykens (2002) found that individuals with deletions were significantly better at regular jigsaw puzzles than individuals with uniparental disomy. There were not enough instances of each type of PWS in this study to expect significant differences between the subtypes. However, the current data follow the patterns expected on the basis of current knowledge. First, mental ages seem to fit the expected trend on IQ tests, with individuals with UPD having MA's similar to the small deletion types, and those with small deletions having higher MA's then those with large deletions (microdeletion > type II deletion > type I deletion). The pattern for MA holds despite the group with UPD having the lowest mean chronological age. Individuals with all deletion forms scored better on the traditional jigsaw puzzle than the individuals with UPD, which is what would be expected from previous research with puzzles. What is surprising is that the 2 individuals with UPD scored better than any of the other subtypes on the blank jigsaw puzzle. This difference was relatively large even for the closest subtype (UPD $M = 21.50$ pieces vs. Type II $M = 14.50$ pieces).

Interpretation of Puzzle Results

Besides replicating the general pattern (if not magnitude of difference) of Dykens' (2002) findings and finding patterns of phenotypic differences similar to those that have been established by previous research, there were a number of other interesting results. Understanding the implications of these results requires an analysis of the possible sources of information available on a puzzle. Pieces in a traditional jigsaw puzzle contain several types of information (summarized in Table 5). Various strategies for approaching jigsaw puzzles may make primary use of different pieces of information. On the basis of my results, I will argue that (1) strategies form general approaches to a given set of problems, (2) if circumstances are similar

Table 5 --Summary of Strategies and Information Used in Constructing Jigsaw Puzzles

Strategy	**Examples of information used**	**Information source**	**Process**	**Requires representations (mental or concrete)**
Local Surface (Surface Features of Jigsaw Pieces)	Piece shape Color Contour Texture	Puzzle piece	Piece matching	No
Local Content (Elements of Picture Content)	Individual elements (e.g. *tree*) Areas of color (e.g. *sky*) Areas of texture (e.g. *lake*)	Puzzle piece Representation (physical picture or mental image)	Constructing and matching individual elements	Yes; secondary role
Global Content (Overall Content of Picture)	Spatial arrangement of content elements (e.g. *lake in front of mountains and beneath a cloudy sky)*	Representation (physical picture or mental image)	Mapping between elements of content and overall spatial arrangement to construct a coherent picture	Yes

enough, a person is likely to use a similar initial strategy, and (3) different strategies will result in measurable differences in outcomes, based on the adequacy of the strategy given the demands of the task. At the same time, I acknowledge that it is possible to use multiple pieces of information simultaneously or to change strategies (either between tasks or within a given task). I will use the mountain scene portrayed in Figure 8 and the picture of water lilies in Figure 9 to illustrate different types of information available in a puzzle and how these kinds of information may map onto different strategies. In my framework, the different strategies form a hierarchy of increasing reliance on knowledge about the picture appearing on the puzzle.

Local Surface

Each piece in a jigsaw puzzle has its own distinct *shape* that is a physical property of the piece itself. Shape is the only kind of information that ultimately determines where a piece can be placed. In addition, pieces contain information related to the local elements of the picture appearing on the puzzle. *Color* is probably the most obvious type of information because pieces that appear next to each other generally have similar colors on them. Piece 1 in Figure 8 illustrates this; the pieces surrounding it would be expected to include white snow and dark rocks. When a piece contains more than one distinct color, the resulting picture *contours* can be a helpful clue as to where the piece needs to be placed. In Piece 2, the top of the glacier in the middle-ground forms a straight line across the piece. Pieces to the left or right could be expected to have a similar contour line. The *textures* depicted on pieces could also be important in deciding which pieces they attach to. A good example of texture helping to determine placement is seen on Piece 3, where part of the lake at the base of the mountains is reflecting the sky. This reflection looks similar to the sky except for small ripples from the lake, which helps to disambiguate the types of pieces that could be expected to appear near it (i.e. pieces containing ripples instead of pieces containing wisps of clouds). Piece shape, color, picture contours, and texture are all local surface features of the pieces. Individuals focusing primarily on local surface features would be expected to show evidence of strategies that indicate they are connecting puzzle pieces based on the properties of individual pieces. Because it requires only information available from individual pieces, a local surface strategy does not require the use of a mental representation of the overall configuration of the scene or use of a concrete representation (the box-top picture).

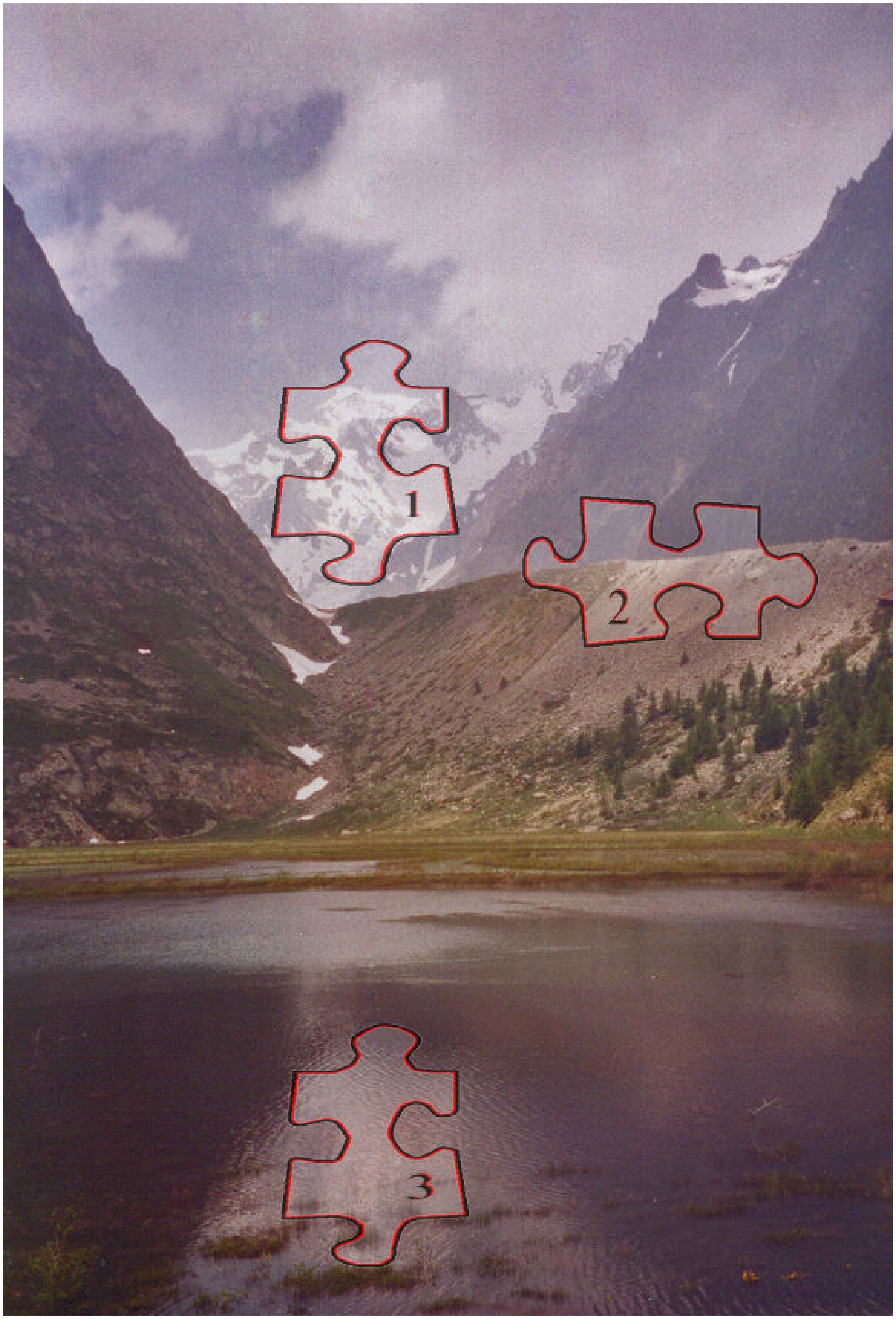


Figure 8 --Puzzle Information Example

Local Content

As an extension of the local surface features mentioned above, it is possible to notice and use *local, content based information*. This approach would involve employing local surface features to construct specific elements of the picture. What differentiates this approach from the local surface approach is that it requires the use of a representation of elements of the picture (either a mental image or the box-top picture). An example of this would be seeing pieces with green on them and attempting to assemble a tree, which depends on the person referring to an idea (mental representation) or image (box-top picture) of a tree. Consciously building elements of a picture requires the understanding of and use of a representation of particular content (i.e. knowledge that the puzzle pieces are part of some larger whole).

Global Content

A final strategy involves focusing on the *global content* of the puzzle. This strategy goes beyond the local content strategy because it not only requires the use of a representation (a mental image or the box-top picture) to identify individual elements (e.g. a tree), but also entails perceiving and making sense of the overall picture and analyzing the spatial location of individuals elements. In other words, using this strategy requires not only knowledge of *what* the different elements of the picture are (e.g. the sky, a mountain, a lake), but *where* the elements appear in the scene (e.g. the sky goes above the mountains and the mountains go above the lake).

Sometimes the spatial relations in a box-top or puzzle picture are relatively transparent (e.g. obviously, the sky does not go below the lake). However, spatial relations can be ambiguous, making it difficult to construct a useful mental representation that will enable the spatial mapping of individual elements. Repeating patterns incorporated into the picture may make it difficult to use the box-top picture (or a mental representation of that picture) as an accurate spatial map of the locations of individual elements.

For example, in Figure 9, lily pads and water lilies form an irregular pattern with repeating elements. Unlike the mountain scene in Figure 8, the spatial arrangement of the elements is complex and difficult to characterize. In this instance, knowing that a piece is part of a flower will not inform the puzzle assembler where to place the piece, as there are 8 possible spatial locations for a flower. If the piece depicts part of a lily pad, the problem is an even larger one (there are at least 21 possible placements). It appears clear that the more repetitive a pattern, the harder it is to take advantage of a global content strategy. Thus, a global strategy is not necessarily the most efficient one for all puzzles.

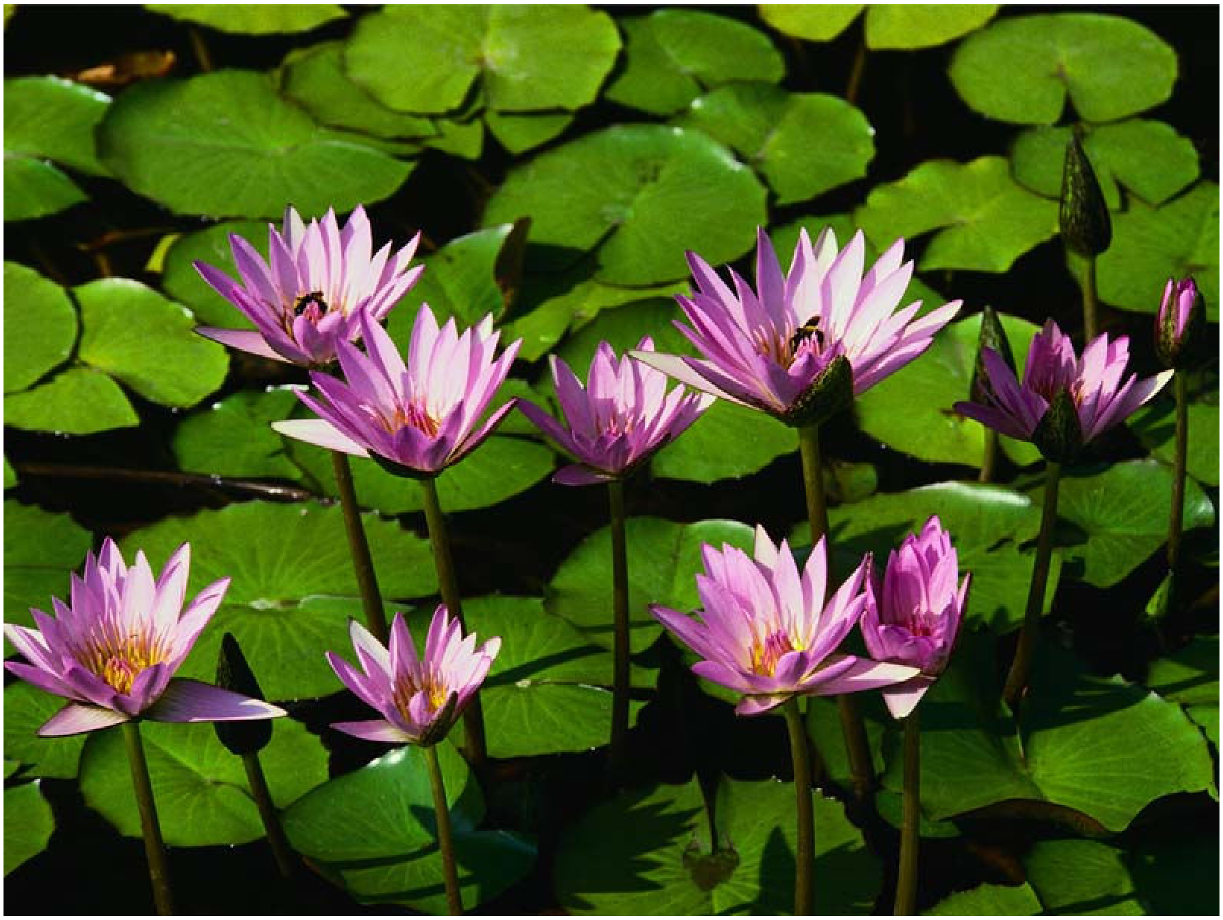


Figure 9 --Repeating Pattern Example

Using this framework of strategies with which a puzzle may be approached, what can be learned from the current pattern of results? For the purposes of this discussion, I will refer to the different types of puzzles used in this study as “containing”, or “not containing”, distinct pieces of information. The blank jigsaw puzzle contains no color, contour, or texture information. Because it lacks these, it also contains no content-based information. Therefore, a local surface strategy, with a strong focus on piece shape, will produce the best outcomes for the blank jigsaw puzzle. The non-interlocking face puzzle, consisting of pieces having all straight edges, was designed to remove shape as a useful information source (mimicking puzzles that are used in a number of IQ tests). Although the pieces still do contain some shape information, the shapes in this puzzle, unlike those in a jigsaw puzzle, cannot confirm a placement because any two pieces with straight edges can be placed next to one another. Therefore, for simplicity sake, I will refer to this puzzle as having no shape information. Matching color, contour, and texture might still be somewhat effective, but these local surface cues could lead to confused placements when used without the benefit of confirmatory shape information. For instance, in the face puzzle they might lead to a scrambled arrangement of facial features. Therefore, utilizing a content-based strategy, in which the confirming information comes from whether or not the pieces go together to make a coherent picture, would probably be most effective.

Because the modified puzzles were intended to remove specific pieces of information, it is not surprising that scores were higher for both groups on the traditional jigsaw puzzle compared to the modified puzzles. What is interesting is that individuals with PWS outperformed TD children on the blank jigsaw puzzle, while falling slightly behind the MA-matches on the non-interlocking puzzle. This pattern of results suggests that the two groups may have used the different types of available information in remarkably different ways.

For the group with PWS, performance was higher on the two interlocking puzzles (the ones providing shape information) and the correlation between scores on these two puzzles was large. This result suggests that the individuals with PWS used shape as the primary source of information for piece placement. Their scores on the two face puzzles were moderately correlated, suggesting that the individuals with PWS also made use of information common to both puzzles (color or spatial arrangement) to solve them. Of interest, for the group with PWS, only one measure of spatial ability (the Motor-Free Visual Perception Test--a measure of spatial visualization) correlated with scores on a single type of puzzle--the non-interlocking one, which requires the use of information about spatial arrangement for success. Their scores on the jigsaw puzzles were not related to visual-spatial ability. Also, the individuals with PWS were less apt to use the box-top picture when shape information was available (on the jigsaw/face puzzle) than when shape information was absent (on the non-interlocking/face puzzle). This converging evidence leads to the conclusion that the individuals with PWS concentrated the most on piece shape, then used other local cues (color, contour, and texture), falling back on content-based information only when surface information failed them.

The TD group had a markedly different pattern of results, suggesting a different pattern of information use. Their puzzle scores correlated with their scores on several of the spatial measures, suggesting that underlying spatial abilities were used for both kinds of tasks. The TD participants' scores on the two face puzzles (those containing spatial arrangement information and color/contour/texture) were much higher than their scores on the blank puzzle (on which they performed the worst) and their scores on the two face puzzles were highly correlated. This pattern of findings leads to the conclusion that the TD group favored a global content strategy rather than focusing on local content or local surface information alone.

Because traditional jigsaw puzzles contain both surface and content features, puzzle assembly could be considered a type of dual representation task (i.e. a task requiring a switch between two possible ways of representing the same object--c.f., DeLoache, 2000). However, unlike many other dual representation tasks (e.g. the dimensional change card sort task, which requires children to keep in mind two levels of decision rules--Frye, Zelazo, and Palfai, 1995), it is possible to assemble jigsaw puzzles using only local surface features of the puzzle pieces. In many puzzles, especially difficult ones with repeating patterns, local surface features are the most informative type of cue. In fact, piece shape, a local surface feature, is the single most constraining kind of information because it is the only information that always indicates the one position that a given piece can occupy. Therefore, while using a global content strategy might be considered more advanced (requiring the integration of local information and interpretation of the box-top picture), it is not likely to be the most effective strategy for difficult puzzles. Indeed, puzzles with repeating content would be more difficult to complete relying solely on a global content strategy. Thus, the most effective strategy is different depending on the information available on a puzzle.

Future Directions

In future studies I will construct puzzles to disambiguate the use of a global content strategy from the use of local surface features other than shape (e.g. color, contour, and texture information). The conclusion that the TD children do not focus on local surface features was drawn largely from their performance on the blank puzzle, which only tested their ability to use piece shape and not other local surface features such as color and contour. Also, separating color from content will help clarify the extent to which individuals with PWS use global content cues or other local features when they are prevented from using shape (as on the non-interlocking puzzle).

The current data support the use of two very different strategies for puzzle assembly in the groups that were tested. The group with PWS showed a tendency to focus more on local cues (surface features) than on global cues (content information). This tendency, whether occurring due to a phenotypic strength or weakness or learned through practice, may be advantageous in the assembly of puzzles and may lead to distinctive strategies in other life situations. A first step in determining if this tendency is learned through practice on jigsaw puzzles, or if it is part of the Prader-Willi syndrome phenotype, is to determine if the preference for local cues and local processing is a pervasive characteristic of this group. Individuals with other syndromes and disorders (e.g. Williams syndrome and Autism) exhibit abnormal local/global processing tendencies. Although Prader-Willi syndrome involves a different, etiology-specific cognitive profile, the fact that abnormal local/global processing characterizes other etiologies makes this an area that warrants further examination.

Another step involves observing puzzle expertise in typical and atypical development. Assuming that individuals with PWS develop the strategy through practice, then TD experts should develop the same strategy and scores for both groups should follow a similar pattern. On the other hand, TD experts' scores on all 3 types of puzzles might improve, while their relative success across puzzle types remained stable. If TD experts use a different strategy, then it would be safe to rule out knowledge gained through experience as the driving factor behind the distinctive pattern of performance in individuals with PWS.

A possible future direction involves potential reasons that individuals with PWS would be drawn to jigsaw and word search puzzles (two seemingly different types of puzzles). Two initial ideas for why individuals with PWS seem to have an expertise in both types of puzzles is that it results from their obsessive/compulsive-like symptoms (which might explain the development of *expertise* in a chosen area, but does little to explain their initial *attraction* to the puzzles) and that puzzles serve as a natural outlet for exceptional visual-spatial abilities (an explanation that the results of this project evidently have ruled out).

One possibility, which I intend to investigate further, is that individuals with PWS have a high need for cognition (curiosity or interest in and enjoyment of the act of thinking--Caccioppo, Petty, & Kao, 1984). A strong interest in various puzzle types may be an indication that these individuals enjoy particular cognitive challenges. That jigsaw and word search puzzles seem to be the most commonly sought out types of puzzles may be due to their availability (these are extremely common puzzles). Additionally, although jigsaw and word search puzzles may not appear to have a lot in common, one characteristic of both is that they are always solvable if you follow a strategy and stick to it. This common property would probably be attractive to an individual with a low IQ but a high need-for-cognition, because the puzzles will not easily frustrate them, but will still provide a challenging activity that can be successfully completed.

Lower average IQ's may explain why individuals with PWS are not drawn to more advanced puzzles that require either extensive background knowledge (e.g. crossword puzzles) or insight to solve them (e.g. the nine dot problem).

These proposed explorations may lead to better knowledge of strengths and deficits within the PWS phenotype and offer more definitive explanations for strong puzzle assembly skills in individuals with PWS. The current project provides initial information about how visual-spatial abilities and puzzle solution strategies overlap and differ in individuals with PWS and typically developing children. The study also helps to round down the number of possible explanations for the puzzle assembly phenomenon in individuals with PWS and to focus future research efforts.

APPENDIX A

SEX DIFFERENCES

In the group with PWS, the females looked at the picture significantly more often (M = 11.40, SD = 6.61) on the non-interlocking/face puzzle than their male counterparts (M = 2.90, SD = 5.45), $t(14) = 7.09$, $p = 0.020$. Individual females were also more likely to use a looking strategy (defined as 3 or more looks) on this puzzle; 80% of the females versus only 20% of the males, $\chi^2(1) = 5.00$, $p = 0.025$. A similar gender difference in the number of looks for the jigsaw/face puzzle was evident (40% of women compared to 0% of men), $\chi^2(1) = 4.615$, p = 0.032.

In the TD group, a trend similar to the pattern found in the group with PWS was found for the number of looks at the picture and the number of individuals using the looking strategy on the non-interlocking puzzles. Females had an average of 7.57 (SD = 4.29) looks compared to 3.57 (SD = 2.52) for the males, t(13) = 4.53, $p = 0.055$, and 100% of the females compared to 57% of the males used a looking strategy, $\chi^2(1) = 3.818$, $p = 0.051$.

Males significantly outperformed females on the Water Level Task [Male M = 8.75 out of 16 (SD = 4.62); Female M = 4.13 (SD = 2.95), $t(15) = 5.69$, $p = 0.032$]. This was probably due to the females' tendency to choose parallel bottles, which they did significantly more than their male counterparts [Female M = 3.00 (SD = 1.41) out of 4 bottles; Male M = 1.13 (SD = 1.55), $t(15) = 6.38$, $p = 0.024$]. None of the other spatial tasks revealed any significant sex differences, with only very small mean differences.

APPENDIX B

MENTAL ROTATION

For the Mental Rotation Task, there was an intriguing, unexplained result. Scores in the TD group followed the expected pattern, decreasing as the angle and the dimension of rotation increased. However, the group with PWS exhibited a somewhat different pattern. Looking at the scores for each presentation angle separately revealed that the scores for the group with PWS increase across the 3-D rotations (except for the 180º trials). For the 3-D rotation at 135º, the participants with PWS actually scored significantly above chance [$t(11) = 2.57$, $p = 0.026$] correctly answering 75.0% of the trials (1 of only 3 angles in which the group with PWS exceeded chance). The difference between this high mean score and that of the TD group (M = 50%) approaches significance, $t(24) = 1.88$, $p = 0.073$, $ES = 0.72$. Also, the reaction times of the group with PWS tended to decrease as their scores increased across the 3-D rotations (except for at the 180º rotation). Thus, not only did the individuals with PWS respond more accurately to the 3-D, 135º rotation than any of the other 3-D trials, but they also tended to respond faster to it.

A closer look at the data revealed a subset of 5 individuals with PWS whose 0-45º composite scores for the 3-D trials were below chance, but whose 90,135, and 180º composite scores were above chance. These scores are compared in Figure 10, a scatter plot (with trend lines), with those whose 0-45º composite scores were at or above chance. These same 5 individuals had extremely fast reaction times to the 135º rotation compared to the rest of the group (4.11 seconds compared to 10.88 seconds, n.s.). An approach to the Mental Rotation Task that would result in this pattern of results is currently unknown.

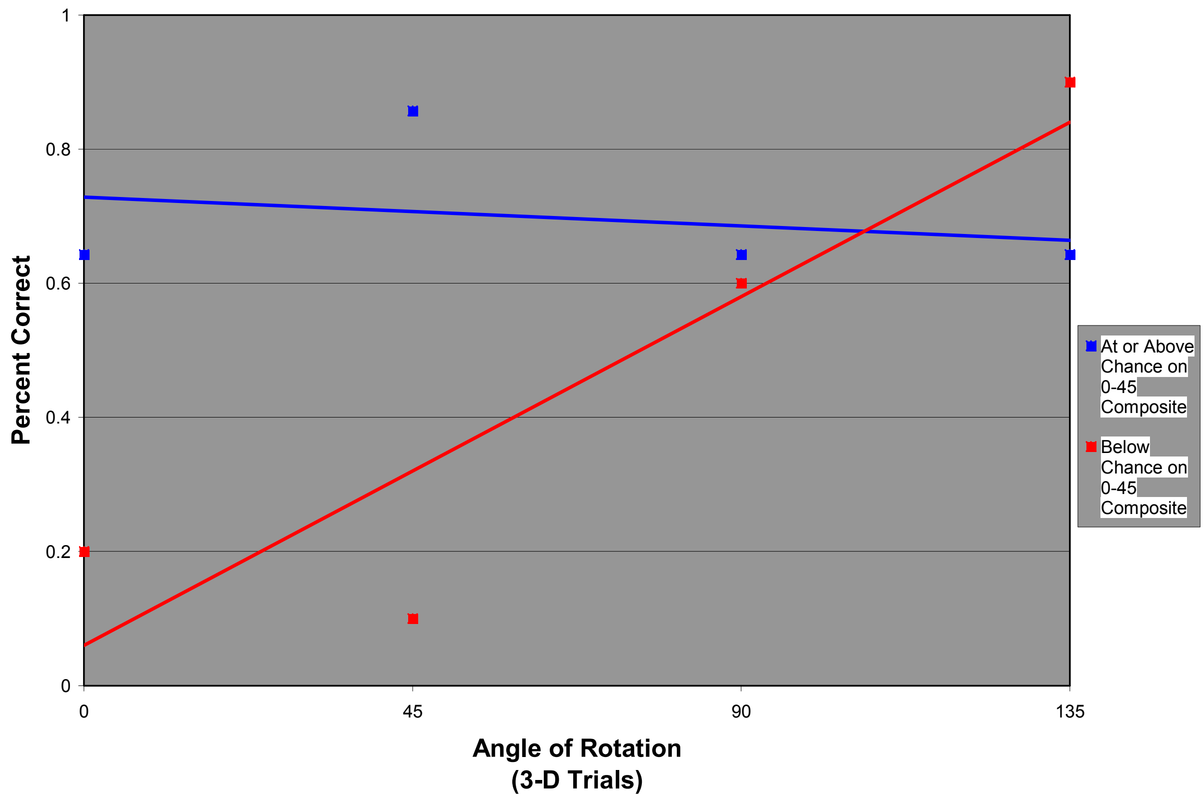


Figure 10 --PWS Mental Rotation Scores:

Individuals Scoring Below Versus At or Above Chance

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