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Refining Behavioral Phenotypes: Personality–Motivation in Williams and Prader-Willi Syndromes

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Despite behavioral differences, individuals with Williams or Prader-Willi syndrome share a proneness to certain personality characteristics. We hypothesized that there are qualitative differences in these shared personality features. Personality–motivation (measured using the Reiss Profiles) was compared for equal numbers of age- and gender-matched individuals with Williams or Prader-Willi syndrome or mental retardation due to nonspecific causes. Each syndrome featured aberrant motivational profiles, and similarities were found across groups in various domains. Significant differences emerged in the specific stimuli that motivated behavior in several Reiss Profile domains. Implications are discussed for the "classic" sociable personality in Williams syndrome and for compulsivity in Prader-Willi syndrome. Recommendations are made for treatment and more refined phenotypic research.

In contrast to even a decade ago, more and more researchers are studying the behavior of people with specific mental retardation syndromes. Although many behavioral researchers study heterogeneous groups of subjects with mixed or unknown etiologies (Hodapp & Dykens, 1994), workers both in the United States and abroad are increasingly examining the so-called "behavioral phenotypes" of people with specific genetic syndromes (Dykens, 1995; Flint & Yule, 1994). This syndromic research, sparked by remarkable advances in molecular genetics and the Human Genome Project, facilitates understanding of gene–brain–behavior relationships and refines treatment and intervention (Dykens, in press, a).

In one approach to phenotypic work, researchers compare behavior across different syndromes and assess the extent to which syndromic behaviors are unique or shared (Dykens, 1995). Making between-syndrome comparisons is relatively easy when individuals with specific syndromes have salient or unique behaviors. Hyperphagia or overeating, for example, is a hallmark characteristic of Prader-Willi syndrome but is rarely seen in other genetic groups. Many phenotypic researchers focus on these apparently unique attributes because they hold considerable promise for making gene–behavior correlations.

Yet most syndromes do not feature unique or totally specific attributes, and even rare or unusual syndromic behaviors can occasionally be seen in others with mental retardation. More often than not, then, people with syndromes show both similarities and differences in behavior (Dykens, 1995) or partial as opposed to totally specific effects (Hodapp, 1997). Heightened anxiety, for example, is seen in people with Williams syndrome and

Prader-Willi syndrome, even as these individuals show striking differences in other traits or behaviors (Dykens, 1999). A further complication to the work is that even when behaviors seem the same across syndromes, more fine-grained studies may reveal important qualitative differences in these shared traits (Dykens, 1995).

These phenotypic complexities are aptly demonstrated in two syndromes that have recently come under considerable scrutiny: Williams and Prader-Willi syndromes. On first glance, these syndromes feature glaring behavioral differences and even possibly unique or totally specific behavioral effects. Hyperphagia, underactivity, and excessive day time sleepiness are salient in Prader-Willi syndrome (Dykens & Cassidy, 1996), whereas overactivity, restlessness, and difficulties sleeping are prominent in Williams syndrome (Einfeld, Tonge, & Florio, 1997; Tomc, Williamson, & Pauli, 1990; Udwin & Yule, 1991).

At the same time, individuals with these syndromes show *partial specificity*, or shared behaviors and personality characteristics; in separate studies investigators have even used identical terms to describe their Williams or Prader-Willi samples. People with both syndromes have been depicted as impulsive, aggressive, stubborn, anxious, obsessive, and prone to perseveration and worry (e.g., Dykens & Kasari, 1997; Einfeld et al., 1997; Pober & Dykens, 1996; Udwin & Yule, 1991). Further, people with Williams or Prader-Willi syndromes have been cast as socially withdrawn or isolated, with poor peer relationships.

Despite these social difficulties, however, people with both syndromes seem to show some social skills or strengths. Many people with Prader-Willi syndrome are described as friendly, pleasant, and polite (e.g., Cassidy, 1984), and persons with Williams syndrome are seen as having a "classic," very sociable personality (for a review see Pober & Dykens, 1996). Although researchers have yet to assess this "classic" personality using standardized measures, anecdotally people with Williams syndrome seem overly friendly, affectionate, engaging, interpersonally sensitive, empathic, and charming (Dilts, Morris, & Leonard, 1990; Gosch & Pankau, 1997; Udwin, Yule, & Martin, 1987). Indeed, many parents worry that their children's outgoing, if not indiscriminate, relating to others may lead to heightened risks of exploitation or abuse (Dykens & Hodapp, 1997).

Differences are, therefore, seen across Williams and Prader-Willi syndromes in many behaviors, especially those related to the regulation of activity, sleep, and appetite. At the same time, individuals with these syndromes share certain personality features, primarily anxiety, obsessiveness, perseveration, and impulsivity. Further, both syndromes feature contradictory descriptions of social interaction: social withdrawal and peer difficulties on the one hand and a friendly, pleasant—if not empathic—demeanor on the other.

Our purpose in the present study is to sort out these conflicting descriptors of social interaction as well as identify the distinct versus shared personality features of Williams syndrome and Prader-Willi syndrome. To this aim, we compared personality characteristics of 35 adolescents and young adults with Williams syndrome with that of 35 age- and-gender matched individuals with Prader-Willi syndrome and 35 people with heterogeneous or nonspecific mental retardation. To assess personality traits, we used the Reiss Profiles, a newly developed measure of 15 fundamental motivations found in people with or without mental retardation (Reiss & Havercamp, 1998). Within each of the 15 domains, specific stimuli are assessed that motivate people to behave in certain ways. As such, this measure is particularly well-suited to determine how syndromic groups might be the "same but different" in their personality features.

Using the Reiss Profiles, we expected to find some obvious and more subtle motivational differences across groups. Relative to their counterparts, for example,

people with Prader-Willi syndrome will be more motivated by food (due to their hyperphagia) and less motivated by physical activity or sexuality (due to their lowered metabolic rates and hypogonadism). Of more central concern, however, are the ostensibly shared personality–motivational features of Williams and Prader-Willi syndromes. Although people with Prader-Willi or Williams syndromes may have similar Reiss Profile social interaction or anxiety scores, we hypothesized that they would also show differences in the specific stimuli that motivate them to behave in certain ways.

Method

Subjects

Participants were 105 individuals (54 males, 51 females) with mental retardation: 35 with Williams syndrome, 35 with Prader-Willi syndrome, and 35 with mixed or nonspecific mental retardation. Subjects ranged in age from 14 to 50 years (M = 25.34, standard deviation [SD] = 8.69).

Subjects were matched across groups on gender (18 males and 17 females per group) and within 2 years of their chronological age. Williams syndrome participants had a mean age of 24.54 years (SD = 7.61), Prader-Willi subjects averaged 26.54 years (SD = 8.91), and the mean age of the nonspecific group was 26.05 years (SD = 9.49).

Most subjects with Williams syndrome (69%) had moderate levels of mental retardation, whereas most with Prader-Willi syndrome (71%) had mild levels of mental retardation. Level of mental retardation data were based on parental reports of previously administered intelligence tests and are quite consistent with more formal studies of IQ and cognition in each of these disorders (Crisco, Dobbs, & Mulhern, 1988; Dykens, in press, b; Greer, Brown, Pai, Choudry, & Klein, 1997). Most subjects in the nonspecific group (66%) had moderate mental retardation. Level of delay, then, differed significantly across groups, χ^2 (2, N = 105) = 14.04, $p < .001$, with more Prader-Willi subjects functioning in the mild range than did their counterparts in either the Williams or nonspecific groups. Level of mental retardation was not correlated with any of the personality variables in any of the three groups. Even so, as IQ level differed across groups, level of delay was used as a covariate in all between-group analyses. Covariance procedures were used instead of matching across groups on IQ level because matching would have resulted in a skewed, unrepresentative sample of lower functioning persons with Prader-Willi syndrome.

Participants with Williams or Prader-Willi syndromes were recruited through the local chapters of the National Williams Syndrome and Prader-Willi Syndrome Associations as well as through referrals from university-based geneticists. Interested families were recruited at local chapter meetings as well as through mailings and were invited to participate in an ongoing study on behavior and development. Participants with nonspecific mental retardation were obtained from the national normative sample (N = 515) of the Reiss Profiles. As described by Reiss and Havercamp (1998), these individuals were recruited from service agencies located throughout the United States. Age- and gender-matched subjects were drawn from this large normative sample. When more than one appropriate match was available, subjects were randomly selected from the roster.

All subjects with Williams syndrome or Prader-Willi syndrome had previously been clinically diagnosed with their respective disorders, and the majority also had undergone molecular genetic testing that confirmed these diagnoses. In the Williams syndrome group, 69% (n = 24) had genetic testing confirming this diagnosis (deletion 7q11.23), and in the Prader-Willi syndrome group, 86% had genetic testing (26 were found to have paternal

deletion 15q11-q13 and 4, maternal uniparental disomy). No differences were found in age, IQ level, gender, or personality profiles in subjects with clinical diagnoses only versus those with both clinical diagnoses and molecular genetic testing. As per caregiver report, subjects with nonspecific mental retardation did not have Prader-Willi or Williams syndromes. Yet they may have had other genetic or organic causes for their delay, or their mental retardation may have been due to unknown causes. This heterogeneity was considered acceptable because mixed groups are widely used in behavioral research (Hodapp & Dykens, 1994), and our aim was compare the syndromic groups to a group of persons with mixed or unknown causes for their delay.

Procedures and Measures

Parents were asked to complete two questionnaires: a demographic information sheet and the Reiss Profiles. They either completed the measures during local syndrome-specific parent meetings or at home and then returned them to us in a stamped, self-addressed envelope. On the demographic information sheet, they were asked to note age, gender, IQ level, and clinical diagnoses and genetic testing information (including where and when diagnoses were made, by whom, and where and when genetic testing was conducted, and the results for the affected offspring).

Reiss Profiles. The Reiss Profiles of Fundamental Goals and Motivation Sensitivities for Persons with Mental Retardation (Reiss & Havercamp, 1998) consists of 100 statements that are rated by an informant on a 5-point scale (1 = *strongly disagree, not at all characteristic of person* to 5 = *strongly agree, definitely characteristic of the person*). A self-report version is also available for persons without mental retardation. Based on a series of psychometric reliability and validity studies on over 2,550 people, the Reiss Profiles is the first instrument that allows researchers to rigorously assess personality–motivation using normative groups of people with and without mental retardation. The Reiss Profiles differs from many other available instruments for use with people who have mental retardation in that they do not measure maladaptive behavior or psychopathology per se (see Aman, 1991, for a review of these measures). Instead, the Reiss Profiles was designed to assess motivational strengths and styles in adolescents and adults; unusually high or low motivational styles may, in turn, lead to aberrant behavior (Reiss & Havercamp, 1996).

The Reiss Profiles features a 15-factor structure, which is consistent across people with and without mental retardation. The factor structure for persons with mental retardation was based on two studies, with a total of 950 participants, in which factor analyses based on maximum likelihood extraction and confirmatory methods were used (see Reiss & Havercamp, 1998, for details). Raw scores are calculated for each of the 15 domains by summing up all items in each domain.

Of the 15 factors, 6 are particularly pertinent to this study: Anxiety (e.g., has many fears, strong feelings of inadequacy), Order (e.g., likes to do things in a precise manner, becomes upset when things are out of place), and 4 socially related domains—Social Contact (e.g., likes to do things in a group, has many friends), Rejection (e.g., highly sensitive to rejection from others, easily offended), Help Others (e.g., goes out of way to help others, tries hard to protect weak), and Attention (e.g., enjoys being a show-off, often initiates interactions with others).

We also examined the remaining 9 Reiss domains: Vengeance (e.g., mean-spirited, vindictive), Food (e.g., very hearty appetite, always thinking about food), Pain (e.g., complains about aches and pains, low tolerance for physical pain), Sex (e.g., strongly attracted to members of opposite or same sex, strong sex drive), Physical Activity (e.g., more than most people, needs to be physically active; very

energetic/vigorous person), Frustration (e.g., impatient with delays, copes with anger poorly), Independence (e.g., more than most, enjoys working independently; strongly prefers to make own decisions), Curiosity (e.g., enjoys learning, wants to understand things), and Morality (e.g., strong sense of personal honor, needs to do right/moral thing).

Results

Within-Group Analyses

Analyses were first conducted in each group to identify possible age, gender, or IQ correlates of personality data. Most of these analyses proved nonsignificant. Among subjects with Williams syndrome, a gender difference was found in the Morality domain, with females scoring higher than males (*M*s = 14.17 and 11.88, respectively), $F(1, 34) = 6.39$, $p < .01$. In the Prader-Willi group, age was correlated with increased scores on the Anxiety domain, $r = .44$, $p < .01$, and with lower scores on the Curiosity domain, $r = -.40$, $p < .01$.

Between-Group Analyses

Reiss Profiles—Domains. To assess group differences across the 15 Reiss Profiles domains, we conducted a multivariate analysis of covariance (MANCOVA), with IQ level as the covariate. This proved significant, $F(30, 176) = 6.91$, $p < .001$, with 11 of the 15 domains showing significant differences. Table 1 summarizes the means, *SD*s, *F* and *p* values, and post-hoc analyses for these 11 domains. The Anxiety domain approached significance, $F(2, 101) = 2.29$, $p < .10$, and nonsignificant differences were found for the Independence, Morality, and Pain domains.

As indicated in Table 1, Newman-Kuels post-hoc analyses revealed several patterns of group differences. First, relative to the nonspecific group, both Williams syndrome and Prader-Willi syndrome subjects showed elevated scores in Help Others, Rejection, and Frustration. Second, compared to the Prader-Willi group, the Williams syndrome and nonspecific groups had elevated scores in Social Contact, Physical Activity, and Sexuality (sexuality scores were relatively low, however, even in the Williams and nonspecific groups). Third, the Prader-Willi group was

Table 1
Means, *SD*s, and *F*s for Reiss Profile Domains Across Groups

	Group[a]						
	WS		PWS		NS		
Domain	Mean	*SD*	Mean	*SD*	Mean	*SD*	*F*
PWS > WS, NS							
Order	16.71	4.84	20.82	4.45	17.22	4.46	8.46***
Food	20.71	5.25	27.17	4.88	18.65	6.59	45.85***
WS, PWS > NS							
Help others	26.91	5.16	26.77	6.28	23.60	5.62	3.28*
Rejection	26.20	5.00	26.40	5.04	22.77	5.78	3.54*
Frustration	22.60	5.41	24.63	5.09	19.71	5.91	5.40**
WS, NS > PWS							
Social contact	27.25	4.40	24.11	4.37	27.63	5.73	4.11**
Sex	7.65	3.43	10.88	3.46	10.46	3.27	9.26***
Physical activity	16.40	4.34	14.08	4.99	18.31	5.25	5.87**
PWS, NS > WS							
Vengeance	15.26	4.67	17.20	4.58	17.97	4.60	3.24*
Curiosity	23.74	5.71	27.17	4.88	27.40	5.83	4.72**
WS > PW							
Attention	25.20	5.13	21.85	4.32	23.25	5.45	3.63*

[a]WS = Williams syndrome, PWS = Prader-Willi syndrome, NS = nonspecific.
*$p < .05$. **$p < .01$. ***$p < .001$.

singularly high in their motivations for Food and Order, and they shared with the nonspecific group elevations in the Curiosity and Vengeance domains. Finally, Attention-Seeking was high in the Williams syndrome relative to the Prader-Willi syndrome group.

Reiss Profiles—Items. To flush out these significant domain findings, we conducted follow-up MANCOVAs, with IQ level as the covariate, using the specific items that comprised each of the 11 significant domains (the Anxiety domain was also analyzed because we hypothesized differences across specific items in this domain). All MANCOVAs were significant, $ps < .001$ to .002, including the Anxiety domain; but consistent with our hypotheses, only certain items in each domain showed significant group differences. Using a Bonferroni-corrected p value for each domain (.05/number items per domain), we identified those items in each domain that differed across groups, based on $ps < .001$ to .006 (see Table 2).

To identify which of the items in Table 2 best differentiated the three groups, we conducted a step-wise discriminant function analysis. Because the Food domain showed the same pattern of findings across 6 different items, only 2 representative food items were entered into the analysis. Of the 21 items entered into the analysis, 9 emerged as the best predictors of group membership: upset with changes in routine, does things in a precise manner, has many friends, feels terrible when others are hurt, low tolerance for teasing, very energetic, enjoys puzzles, eating is more important than for most others, and has many fears. Overall, these 9 items predicted group membership with 91% accuracy, with correct classification of 89% of Williams syndrome subjects, 94% of Prader-Willi subjects, and 91% of the nonspecific group. Based on these 9 items, only 9 subjects were misclassified: 4 with Williams syndrome, 2 with Prader-Willi syndrome, and 3 with nonspecific mental retardation.

Discussion

Although certain aspects of the "classic" Williams syndrome personality were supported, both similarities and differences were found across the Williams, Prader-Willi, and nonspecific groups in social interaction, anxiety, and orderliness. Nine personality items predicted group membership with 91% accuracy, and this high prediction rate highlights the need to probe for specific differences in shared personality features across syndromes. Findings refine the Williams syndrome and Prader-Willi syndrome behavioral phenotypes as well as demonstrate the need for more detailed studies of behavior in persons with other genetic syndromes.

Although many people with Williams syndrome may have a sociable personality style, these features may not be unique. Both the Williams and nonspecific groups had virtually identical scores in the Social Contact domain, which included items such as "likes to do things in groups," and "more than most people, desires emotional support from others." At the same time, however, Williams syndrome participants had higher scores than did their counterparts in either group on two items in the Attention domain: "often initiates interactions with others" and "never goes unnoticed when in a group."

Interests in others were seen as well in the Help Others domain, with both the Williams and Prader-Willi groups showing "strong desires to help others" and being "very happy when others do well." The Williams syndrome group, however, was singularly high in the item "feels terrible when others are hurt." Although empathy is a complex concept, researchers generally agree that these abilities—to respond to both the well-being and pain of others—are defining characteristics of empathy (see Feshbach, 1997, for a review).

Although empathy has yet to be directly examined in persons with Williams syndrome, this characteristic may relate to an unusual cognitive–linguistic

Table 2
Means, *SD*s, and *F*s for Items in Reiss Profiles Domains Across Groups

Domain/Item	Group[a] WS Mean	WS *SD*	PWS Mean	PWS *SD*	NS Mean	NS *SD*	*F*	Post-hoc analyses
Order							4.89***	
Upset with changes in routine	3.71	.127	4.00	1.13	2.80	1.10	8.61***	WS,PW>NS
Does things in precise manner	2.51	1.17	3.63	1.19	2.95	1.04	10.48***	PW>WS,NS
Strong need to put things in order	2.08	.95	3.09	1.09	2.05	.99	11.18***	PW>WS,NS
Social Contact							2.42**	
Has many friends	3.06	1.37	2.57	1.09	3.68	.90	5.44**	NS>WS,PW
Help Others								
Feels terrible when others hurt	4.06	.68	3.28	.86	3.17	.89	12.00***	WS>PW,NS
Strong maternal/paternal instincts	2.68	1.07	3.51	1.12	2.74	1.04	7.33***	PW>WS,NS
Rejection							4.49***	
Low tolerance for teasing	3.86	.97	4.28	.78	2.71	1.12	19.20***	PW>WS>NS
Attention-Seeking							3.41***	
Often initiates interactions	4.26	.78	3.65	1.23	3.45	1.12	5.32**	WS>PW,NS
Never goes unnoticed in groups	4.00	.89	2.85	1.11	3.40	1.11	7.88***	WS>NS>PW
Physical Activity							3.25***	
Very energetic	2.94	1.16	1.96	.89	3.25	1.21	10.94***	WS,NS>PW
Likes sports/athletics	3.05	1.09	2.31	1.27	3.26	1.19	6.74***	WS,NS>PW
Sexuality							2.90**	
Strong sex drive	2.51	1.03	1.60	.88	2.37	.94	8.37***	WS,NS>PW
Above average interest in sex	2.60	1.06	1.71	.98	2.68	.93	9.23***	WS,NS>PW
Curiosity							5.47***	
Enjoy puzzles or mysteries	2.31	1.25	4.37	.91	3.00	1.00	32.22***	PW>NS>WS
Vengeance							3.03***	
Mean-spirited	1.26	.44	1.68	.67	1.82	.62	10.09***	PW,NW>WS
Uncaring	1.51	.61	1.77	.60	2.17	.89	6.75**	NS>WS,PWS
Food							13.95***	
Enjoys eating more than most	3.17	1.17	4.77	.42	3.20	1.25	30.77***	PW>WS,NS
Always thinking about food	3.03	.97	4.40	.88	2.42	1.09	36.77***	PW>WS,NS
Very hearty appetite	3.20	1.18	4.57	.61	3.34	1.27	15.69***	PW>WS,NS
Eating more important than most	2.60	1.00	4.54	.78	2.37	1.06	49.34***	PW>WS,NS
Often asks about next meal	4.00	.90	4.65	.48	2.47	1.14	50.48***	WS,PW>NS
Anxiety								
Has many fears	3.44	1.13	2.33	1.83	2.60	.81	9.82***	WS>PW,NS
Frustration							2.70***	
Low frustration tolerance	3.82	.89	3.88	1.02	3.02	1.07	5.90**	WS,PW>NS
Impatient with delays	3.48	1.07	3.77	.91	2.60	1.14	11.16***	WS,PW>NS

Note. Bonferroni-corrected *p* values.
[a]WS = Williams syndrome, PWS = Prader Willi syndrome, NS = nonspecific.
**$p < .01$. ***$p < .001$.

profile seen in some persons with this disorder. Many people with Williams syndrome show significant relative weaknesses in visual–spatial functioning, including integrating parts into a whole (Bihrle, Bellugi, Delis, & Marks, 1989). Despite these deficits, however, facial recognition seems well-preserved, and many people with this disorder show marked interests in the faces and emotional expressions of others (Bellugi, Wang, & Jernigan, 1994). In addition, some individuals with Williams syndrome show relative strengths in expressive language, including loquacious "cocktail-party" speech, with well-developed affect, prosody, and vocabulary (Reilly, Klima, & Bellugi, 1990; Udwin & Yule, 1990). Although further studies are needed, it may be that strengths in facial and emotional

recognition, as well as the tendency to infuse affect into speech, come together in what is perceived as an empathic stance toward others.

Yet despite their propensity to readily approach others, and their possible empathic streak, people with Williams syndrome do not appear to fare well in establishing friendships. Indeed, relative to Williams or Prader-Willi syndrome participants, subjects in the nonspecific group had significantly higher scores on the item "has many friends," and this item was discrepant enough across groups to contribute to the discriminant function analysis. Conceptualizing and measuring friendships in people with disabilities is complex (Freeman & Kasari, in press), and friendships were not the focus of this study. Even so, it appears that people with Williams syndrome find it hard to translate their sociability into friendships (see also Dilts et al., 1990; Gosch & Pankau, 1997).

Reiss Profiles findings suggest some reasons for these difficulties making friends. Many people with Williams syndrome appear to have a low tolerance for frustration and teasing, and they may, therefore, seem impatient and short-fused. Yet many individuals are also loquacious and quick to approach others, to the point where they are rarely unnoticed in groups. These socially disinhibited individuals may, therefore, come on too strong with others and find it hard to shift gears and modulate their behavior in order to engage in the types of reciprocal interactions commonly seen in friendships.

Social disinhibition and difficulties modulating interactions may be associated with the high levels of inattention and motoric overactivity often seen in people with Williams syndrome, especially in the childhood years. Yet disinhibition may also be related to anxiety, and elevated worry and anxiety is a finding in virtually all studies to date in which maladaptive behavior associated with William syndrome is examined (Dilts et al., 1990; Einfeld et al., 1997; Udwin & Yule, 1991; Udwin et al., 1987). Interestingly, however, participants with Williams syndrome in this study primarily showed heightened Anxiety domain scores in just one item, "has many fears," and this item was one of the 9 that successfully differentiated groups. Indeed, recent work shows a wide variety of more frequent and intense fears in people with Williams syndrome relative to others with mental retardation as well as more fear-related social-adaptive impairment (Dykens, 1999). Friendships may be an aspect of social-adaptive functioning that is particularly thwarted by excessive anxiety and fears.

The findings in this study also clarify and expand key features of the Prader-Willi syndrome behavioral phenotype. Not surprisingly, results of the Reiss Profiles showed that people with Prader-Willi syndrome are highly motivated by food and not particularly motivated by sexuality or physical activity. Similarly, their impatience and low frustration tolerance have also been previously well-described. Beyond these expected results, three sets of findings are new or noteworthy.

First, relative to their counterparts in either of the other two groups, subjects with Prader-Willi syndrome were more likely to "enjoy puzzles or mysteries"; indeed, this item was one of the 9 that best differentiated the three groups. This finding verifies a long-held clinical impression that people with Prader-Willi syndrome often excel at jigsaw puzzles, and we have observed as well that some of these individuals are fascinated by "word search" puzzles. Interests in both types of puzzles may be associated with the relative strengths in spatial-perceptual organization, visual processing, and reading decoding shown by many persons with this syndrome (see Dykens, in press, b, for a review). Although an unusual skill with jigsaw puzzles is actually noted as a minor supportive finding in the consensus diagnostic criteria for Prader-Willi syndrome (Holm et al., 1993), researchers have yet to examine this relatively unexplored aspect of the Prader-Willi behavioral phenotype.

A second key finding was that relative to the other groups, the Prader-Willi group was more motivated by orderliness. They were, for example, more apt to need to do things in a precise manner and to put things in order. People with Prader-Willi syndrome invariably obsess about food, yet non-food obsessive–compulsive behaviors have now been consistently well-described in these individuals, including symptoms such as ordering and arranging, hoarding, skin-picking, and being concerned with cleanliness, symmetry, and exactness (Dykens et al., 1996). Importantly, these compulsive symptoms go beyond the needs for sameness in routine often shown by people with mental retardation, including the subjects with Williams syndrome in this study. Skin-picking and other compulsions accurately differentiate people with Prader-Willi from other groups of individuals with mental retardation (Dykens & Kasari, 1997; Dykens & Smith, 1998; State, Dykens, Rosner, Martin, & King, in press), and increased risks of obsessive–compulsive disorder are suggested in this population (Dykens et al., 1996).

A final set of findings concerns social interactions. Curiously, although people with Prader-Willi syndrome are prone to withdrawal and poor peer relations, subjects with this disorder in the present study were just as motivated to help others as were participants with Williams syndrome. On closer look, the Prader-Willi group showed particularly strong maternal/paternal instincts. Clinically, we have observed that many people with Prader-Willi syndrome are overly nurturant and protective of their household pets and often want to work with animals or with babies and children in day care centers. These yearnings may relate to recent reports of elevated levels of oxytocin in the cerebrospinal fluid of adults with Prader-Willi syndrome (Martin, State, Kaye, North, & Leckman, in press). Oxytocin, a neuropeptide, mediates a host of human behaviors, including eating, grooming, aggression, pair-bonding, reproduction, and infant attachment (see Leckman et al., 1994, for a review). Further work is needed to assess to what extent increased maternal/paternal instincts or nurturant behaviors are linked to altered levels of oxytocin in this population.

Findings from the present study have implications for intervention and for phenotypic research in general. With regard to intervention, the Reiss Profiles is based on the idea that individual differences across 15 basic motivational needs are key to predicting human behavior (Reiss & Havercamp, 1996). Simply put, aberrant motivations, or those that are too high or too low, lead to aberrant behaviors. Instead of specific behaviors, then, Reiss and Havercamp (1998) have postulated that aberrant motivations might be the more appropriate targets for intervention.

It remains unclear, however, to what extent treatment can really alter the unusual motivational profiles seen in people with Prader-Willi or Williams syndromes, especially if these profiles are associated with genetic predispositions to certain styles. On a practical level, then, interventionists typically deal with more "downstream" effects of aberrant motivation (i.e., maladaptive behaviors and emotions). Although treatment recommendations for maladaptive behaviors in persons with Williams or Prader-Willi syndromes are detailed elsewhere (Dykens & Hodapp, 1997), findings from this study highlight certain of these recommendations. Findings about individuals with Williams syndrome underscore the need for social skills training programs, especially ones that teach persons how to make friends and be appropriately wary of others. For people with Prader-Willi syndrome, findings emphasize the need for behavioral and pharmacologic treatments that reduce compulsivity as well as the need for intensive, highly restrictive interventions that target the drive for food (for details see Dykens & Hodapp, 1997, and Hanchett & Greenswag, 1998).

Findings also highlight certain caveats in conducting behavioral phenotype

research in general. Considerable research has now identified global behavioral patterns in people with Williams, Prader-Willi, and other genetic syndromes, in part because of the recent proliferation of psychometrically sound behavior rating scales (see Aman, 1991, for a review). Yet phenotypic work should not stop with global rating scales; researchers should also conduct detailed, follow-up studies to further specify unique syndromic behaviors or demonstrate qualitative differences in ostensibly shared behaviors (Dykens, 1995). This level of behavioral specificity is critically important in the search for gene–brain–behavior relationships, and discovering these correlations is one of the prime reasons for engaging in phenotypic research. It is, thus, less helpful for advancing gene–behavior correlations to observe that people with Williams or Prader-Willi syndromes are anxious or obsessive and more helpful to relate anxiety in Williams syndrome to specific fears or social disinhibition and anxiety in Prader-Willi syndrome to compulsivity. Similarly, although people with Williams or Prader-Willi syndromes are equally motivated to help others, they seem to do so in very different ways and through potentially different causal pathways. In the future, then, researchers need to move away from global to more refined, well-differentiated behavioral descriptions of Williams, Prader-Willi, and other genetic mental retardation syndromes.

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