American Journal on Mental Retardation, 1997, Vol. 102, No. 3, 228–237

Maladaptive Behavior in Children With Prader-Willi Syndrome, Down Syndrome, and Nonspecific Mental Retardation

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Although some genetic, mental retardation syndromes have well-described behavioral features, comparative studies have not yet assessed the relative uniqueness of these so-called phenotypes. Maladaptive behavior of 43 children with Prader-Willi syndrome was compared to age- and gender-matched children with Down syndrome and with nonspecific mental retardation. The Prader-Willi group showed more frequent and severe internalizing, externalizing, and total problem behaviors on the Child Behavior Checklist. Some problems were elevated in all groups, and 12 behaviors were significantly elevated in Prader-Willi subjects relative to both comparison groups. Seven behaviors predicted membership into the Prader-Willi group with 91% accuracy. Implications were discussed for research on behavioral phenotypes in general and for dual diagnosis in particular.

Advances in genetics have sparked renewed interest in the behavior of people with specific mental retardation syndromes. Researchers now find that many individuals with these syndromes show distinctive behavioral features or behavioral phenotypes (Dykens, 1995; Hodapp, 1997; O'Brien & Yule, 1995). Most researchers studying behavioral phenotypes use within-syndrome approaches, examining the extent to which certain behavioral patterns are consistently seen in people with the same genetic anomaly (e.g., Dykens, Finucane, & Gayley, 1997; Kasari, Freeman, Mundy, & Sigman, 1995). Within-syndrome studies are particularly helpful in relatively rare or newly delineated disorders, as well as in syndromes with little previous behavioral research. Prader-Willi syndrome is one condition that has benefited from such within-syndrome work.

Identified just 40 years ago, Prader-Willi syndrome occurs in approximately 1 in 15,000 births and typically results in mild to moderate mental retardation. Most cases, about 70%, are caused by a paternally derived deletion on chromosome 15; remaining cases are generally attributed to maternal uniparental disomy, or when both chromosome 15s are derived from the mother (Butler & Palmer, 1983; Nicholls, Knoll, Butler, Karam, & Lalande, 1989). In either case, Prader-Willi syndrome is associated with an absence of paternally derived information to the Prader-Willi critical region on chromosome 15.

The syndrome is perhaps best known for its striking food-related characteristics, including hyperphagia, food-preoccupations and foraging, and increased risks of obesity in affected individuals (Holm et al., 1993). In addition, many people with Prader-Willi syndrome show significant behavioral and emotional dysfunction. Common problems include temper tantrums, stubbornness, lability, impulsivity, argumentativeness, depression, anxiety, and repetitive behaviors such as skin-picking (Dykens, Hodapp, Walsh, & Nash, 1992a; Greenswag, 1987; Whitman & Accardo, 1987). These behaviors are seen throughout the syndrome's IQ spectrum and are variably expressed throughout the life cycle (Dykens et al., 1992a, 1992b; Dykens & Cassidy, 1995).

Many people with Prader-Willi syndrome also show obsessions and compulsions above and beyond their hyperphagia or food preoccupations (Dykens, Leckman, & Cassidy, 1996; Stein, Keating, Zar, & Hollander, 1994). Prominent obsessive–compulsive symptoms include hoarding (e.g., paper, pens, books), ordering and arranging objects according to certain rules or simply until they are "just right," rewriting and redoing, and repetitive telling or asking. Compulsivity is often associated with distress and adaptive impairment, suggesting increased risks of full-blown obsessive–compulsive disorder in this population (Dykens et al., 1996). These and other psychiatric vulnerabilities, combined with problem behaviors and persistent food-seeking, often lead to less-than-optimal adaptive outcomes for many people with Prader-Willi syndrome (Dykens & Cassidy, 1996).

In all this work, however, researchers have yet to compare maladaptive behavior in people with Prader-Willi syndrome to others with mental retardation. Indeed, although there has been much within-syndrome work, comparisons are rarely made across different syndromic groups (Dykens, 1995; Einfeld & Hall, 1994). It thus remains unclear to what extent maladaptive features in people with Prader-Willi syndrome are distinctive from others with mental retardation.

This study is the first in which investigators have compared maladaptive behavior of children and adolescents with Prader-Willi syndrome with that of youngsters in two other groups: those with Down syndrome and those with nonspecific mental retardation. Down syndrome, the most common chromosomal cause of mental retardation, provides an important contrast to the more rarely occurring Prader-Willi syndrome. Further, a prevailing view of children with Down syndrome casts them as easy-going, friendly, charming, and at low-risk for psychiatric or behavioral problems (e.g., Gibbs & Thorpe, 1983; Hornby, 1995). Yet many children with Down syndrome have significant behavioral and emotional dysfunction, including oppositionality, aggression, stubbornness, and anxiety (Gath & Gumley, 1986; Meyers & Pueschel, 1991; Pueschel, Bernier, & Pezzullo, 1991). Children with Down syndrome are, therefore, far from problem-free, and many show behavioral difficulties similar to those seen in children with Prader-Willi syndrome.

We also used a comparative group of youngsters with nonspecific mental retardation. In most behavioral and dual diagnosis research, investigators select individuals with unspecified causes for their developmental delay or those with heterogeneous or mixed etiologies (Dykens, 1995, 1996; Hodapp & Dykens, 1994). Therefore, in the present study we included a mixed group so that we could more rigorously assess the distinctiveness of maladaptive behaviors in subjects with Prader-Willi syndrome and increase generalizability of findings.

We predicted that all three groups would show common problems, such as tantrums or stubbornness, but that children with Prader-Willi syndrome would exhibit more frequent and severe levels of these problems relative to age- and gender-matched subjects with Down syndrome or nonspecific mental retardation. We also expected that subjects with Prader-Willi

syndrome would show increased rates of repetitive behavior, such as skin-picking, obsessions, and compulsions. Finally, we predicted that maladaptive behavior would be more variably expressed by individuals in the two comparison groups.

Method

Subjects

Participants were 129 children with mental retardation ages 4 to 19 years who belonged to one of three etiologic groups: Prader-Willi syndrome, Down syndrome, or nonspecific mental retardation. There were 43 subjects (15 males, 28 females) in each group. They were matched across groups on both gender and age. The mean age in all three groups was 11 years.

Subjects with Prader-Willi syndrome were recruited through the California or Connecticut chapters of the Prader-Willi Syndrome Association (USA), an organization providing information to families. All subjects had a priori diagnoses of Prader-Willi syndrome, meeting clinical criteria for this disorder (Holm et al., 1993). Further, the majority (70%) also had undergone molecular genetic testing for Prader-Willi syndrome. No differences in maladaptive behavior were found between subjects with clinical diagnoses only versus those with both clinical and genetic diagnoses. As determined by Body Mass Indices—BMIs (weight in kilograms/height in meters^2), 58% of subjects with Prader-Willi syndrome were obese, with BMIs greater than 22 (Rolland-Cachera et al., 1982).

Most participants with Down syndrome (65%, or 28 subjects) were recruited through the Down Syndrome Association of Los Angeles. Remaining participants (35%, or 15 subjects) were recruited through public school systems in Connecticut. Diagnoses of trisomy 21 were confirmed in all cases by parents.

All subjects with nonspecific mental retardation were recruited through public school systems in Connecticut. As per parent and teacher report, subjects did not have Down syndrome, Prader-Willi syndrome, or any other known genetic cause for their mental retardation. As we did not follow-up caregiver reports with genetic testing, this group may have included subjects with undetected genetic anomalies. This heterogeneity was considered acceptable because our goal was to compile a group of subjects with mixed or unknown etiologies.

Participants were thus recruited from parent groups and school systems in either California or Connecticut. We checked for possible biases associated with these two resources and states; no differences were found across the two resources or states in age, maladaptive behavior, gender, or IQ. All subjects lived at home, attended their local public schools, and were identified as special education students.

As expected, mean IQs differed across the three groups, $F(2, 127) = 41.63$, $p < .001$. Based on previously administered standardized intelligence tests, the Prader-Willi syndrome group had a mean IQ of 69 (standard deviation [*SD*] = 8.44); Down syndrome subjects had a mean IQ of 48 (*SD* = 11.18); and the mean IQ of subjects in the nonspecific group was 56 (*SD* = 10.91). As such, IQ was used as a covariate in all between-group analyses.

Procedure and Measures

As previously noted, parents were notified of the study through syndrome-specific parent organizations or public school systems. Parents were invited to participate in a study on behavioral aspects of children with developmental disabilities; the term *maladaptive behavior* was avoided in an effort to reduce selection biases (e.g., volunteering because of unusually high rates of maladaptive behavior).

Those volunteering for the study completed an informational sheet asking for the child's age, gender, results of intelligence testing, and diagnostic history as well as the Child Behavior Checklist

(Achenbach, 1991), a widely used measure of maladaptive behavior in children and adolescents. For this instrument, a 3-point scale is used to assess 112 items (0 = not true; 1 = somewhat or sometimes true; 2 = very true or often true); the 2 items that tap "other" problems were not used in the study. Consisting of eight narrow-band and two broad-band domains, the Child Behavior Checklist assesses internalizing problems (withdrawn, somatic complaints, anxious/depressed); and externalizing problems (delinquent behavior, aggressive behavior). Remaining clinical domains include social problems, thought problems, attention problems, and other problems. A total Child Behavior Checklist score is based on findings from the entire measure. In this study we used Child Behavior Checklist raw scores; total Child Behavior Checklist *t* scores were also examined to assess clinical severity. The reliability and validity of this measure has been well-established, and the instrument has also been successfully used in previous studies of persons with mental retardation.

Results

Within-Group Analyses

Separate analyses were conducted for each of the three etiologic groups to assess relations between maladaptive behavior and gender, IQ, and age. No gender differences were found in Child Behavior Checklist scores for subjects in the Prader-Willi or Down syndrome groups. Males in the nonspecific group showed more externalizing problems than did females (means = 22.75 vs. 9.75), $t(41) = 3.15$, $p < .001$, and IQ was not significantly correlated with Child Behavior Checklist domain scores within any of the three etiologic groups.

Age emerged as a significant correlate of maladaptive behavior only in subjects with Down syndrome. In this group, age was associated with internalizing problems, $r = .45$, $p < .01$, primarily anxiety/depression and withdrawal, $rs = .31$ and .42, $ps < .01$, respectively. For the Prader-Willi group, the BMI was not correlated with Child Behavior Checklist scores.

Between-Group Analyses

Domains. To assess group differences in the internalizing and externalizing domains and total Child Behavior Checklist, we conducted three analyses of covariance, with IQ as the covariate. Subjects with Prader-Willi syndrome had significantly higher scores in all three domains relative to the Down syndrome or nonspecific groups. Table 1 summarizes *F* values, and domain mean raw scores and *SD*s across the three groups. Newman-Keuls post hoc tests revealed significantly higher internalizing and total scores in the Prader-Willi group relative to both comparison groups. For the externalizing domain, subjects with Prader-Willi syndrome differed significantly from those in the Down syndrome group only.

To further assess symptom severity, we converted total Child Behavior Checklist scores to *t* scores and identified the proportion of subjects in each group who showed clinically elevated *t* scores. As described by Achenbach (1991), total *t*

Table 1
Mean Domain Raw Scores, *SD*s, and *F* Values on the Child Behavior Checklist by Group

Child Behavior Checklist (CBCL)	Prader-Willi syndrome		Down syndrome		Nonspecific MR*		
	Mean	*SD*	Mean	*SD*	Mean	*SD*	*F*
Internalizing	12.93	6.85	6.37	5.65	8.26	6.37	7.91**
Externalizing	18.49	8.91	11.42	9.81	14.67	13.25	3.61*
Total CBCL	63.24	20.69	38.65	22.57	44.37	28.97	11.27**

*Mental retardation.
*$p < .05$. **$p < .001$.

scores greater than or equal to 64 successfully differentiate typical from clinically referred samples. Among subjects with Prader-Willi syndrome, 72% (31 of 43 subjects) showed clinically elevated total *t* scores. Clinical significance was reached for only 23% of subjects with Down syndrome (10 of 43) and 39% of subjects with nonspecific mental retardation (17 of 43). A 3 (group) × 2 (above/below clinical cutpoint) chi-square analysis was significant, $\chi^2(2) = 21.49$, $p < .0001$. Follow-up 2 × 2 chi-square tests revealed that the Prader-Willi group had a greater proportion of clinically elevated scores than did both comparison groups.

Specific Behaviors. Three approaches were used to examine specific behaviors. We first identified frequently occurring Child Behavior Checklist items, or those behaviors that were shown by 50% or more of subjects in any of the three groups. Twenty-four behaviors occurred frequently, and the percentages of subjects in each group showing these behaviors are listed in Table 2.

Table 2
Percentages of Subjects in Each Group Showing 24 Frequently Occurring Behaviors

Behavior	Prader-Willi	Down syndrome	Nonspecific MR[a]
Skin-picking	95	20	26
Argues a lot	95	65	63
Stubborn	93	72	60
Underactive	91	35	28
Obsessions	88	46	37
Tantrums	88	47	48
Clumsy	86	44	55
Overtired	84	25	23
Disobeys	81	72	45
Overeating	80	27	21
Mood changes	77	39	56
Speech problems	76	77	44
Talks too much	74	35	46
Excessive sleep	74	11	9
Impulsive	74	44	72
Gets teased by peers	72	28	32
Overweight	70	30	16
Compulsions	68	32	37
Prefers being alone	67	63	28
Can't concentrate	67	72	79
Lies, cheats	67	23	35
Kids don't like	65	32	39
Steals at home	58	6	9
Hyperactive	21	35	60

[a]Mental retardation.

Second, using mean scores, we then conducted analyses of covariance across these behaviors, with IQ as the covariate. Given the number of comparisons, the Bonferroni correction procedure was employed, resulting in a significance level of *p* less than .002. Of the 24 behaviors, 15 showed differences across the three groups. Means, *SD*s, and *F* values for these 15 behaviors are presented in Table 3.

Twelve behaviors were significantly elevated in the Prader-Willi group relative to both the Down syndrome and nonspecific comparison groups. Most of these 12 behaviors are characteristic of Prader-Willi syndrome: overeating, overweight, underactivity, sleeps too much, skin-picking, obsessions, argues a lot, and stealing at home (food or money to buy food). Other elevated behaviors in the Prader-Willi group have not been as widely noted, including compulsions, talking too much, and getting teased a lot by peers.

Of the three remaining differences across groups, two behaviors were significantly higher in the Prader-Willi and Down syndrome groups relative to nonspecific subjects: speech problems and preferences to be alone (see Table 3). Hyperactivity emerged as significantly higher for subjects in the nonspecific group compared to those in the two syndromic groups.

Third, we performed a step-wise discriminant function to identify those behaviors that best separated and predicted membership into the three groups. Seven behaviors best differentiated the groups: skin-picking, overtired, obsessions, impulsivity, speech problems, talks too much, and hyperactive. Table 4 lists the global pattern of these seven behaviors in discriminating the three groups.

The discriminant analysis correctly classified 91% of Prader-Willi cases, 80% of Down syndrome cases, and 70% of nonspecific cases. The overall classification rate was 80%. As indicated in Table 5, only 4 Prader-Willi cases were misclassified, and just 3 comparison group

Table 3
Means, *SD*s, and *F* Values of 15 Behaviors Showing Significant Differences Across Groups

Group difference/ Behavior	Prader-Willi syndrome (PWS) Mean	*SD*	Down syndrome (DS) Mean	*SD*	Nonspecific MR[a] (NS) Mean	*SD*	*F*
PWS > DS, NS							
Skin-picking	1.63	.58	.26	.54	.21	.51	67.10
Argues a lot	1.51	.59	.79	.67	.95	.84	7.05
Obsessions	1.44	.70	.60	.73	.60	.85	10.83
Underactive	1.33	.64	.42	.63	.35	.61	23.42
Overeating	1.26	.79	.40	.69	.30	.64	17.85
Talks too much	1.26	.85	.44	.67	.86	.97	6.31
Excessive sleep	1.19	.82	.14	.41	.14	.47	30.68
Overweight	1.14	.86	.40	.66	.26	.62	13.05
Overtired	1.12	.66	.26	.44	.26	.49	20.93
Gets teased	1.05	.79	.30	.51	.42	.66	8.72
Compulsions	.98	.80	.40	.62	.60	.85	6.61
Steals at home	.70	.67	.09	.37	.09	.37	17.17
PWS, DS > NS							
Speech problem	1.37	.85	1.30	.83	.65	.81	10.70
Rather be alone	.84	.69	.77	.68	.33	.57	8.12
PWS, DS < NS							
Hyperactive	.23	.48	..47	.70	.86	.80	6.53

Note. $p < .002$ for all comparisons.
[a]Mental retardation.

Table 4
Relations of Seven Behaviors in Discriminant Factors by Group

Behavior	Prader-Willi syndrome	Down syndrome	Nonspecific MR[a]
Skin-picking	High	Low	Low
Overtired	High	Low	Low
Obsessions	High	Low	Low
Impulsive	High	Low	High
Speech problem	High	High	Low
Talks too much	High	Low	Low/High[b]
Hyperactive	Low	Low	High

[a]Mental retardation. [b]Lower than subjects with Prader-Willi syndrome/higher than subjects with Down syndrome.

Table 5
Discriminant Function Classification Rates Across Groups

Group	Prader-Willi syndrome *n*	%	Down syndrome *n*	%	Non-specific MR[a] *n*	%
Prader-Willi syndrome	39	91	4	9	0	0
Down syndrome	2	4	34	80	7	16
Nonspecific MR	1	2	12	28	30	70

[a]Mental retardation.

cases were misclassified as belonging to the Prader-Willi group.

Discussion

Children with Prader-Willi syndrome showed more frequent and severe maladaptive behaviors relative to their age- and gender-matched peers with Down syndrome or with nonspecific mental retardation. Further, a blend of certain maladaptive behaviors appears to be quite distinctive to Prader-Willi syndrome and highly predictive of this disorder. Overeating, food obsessions, and sleep disturbances are salient for people with Prader-Willi syndrome, yet other obsessions and repetitive, compulsive-like behaviors also emerged as central distinguishing features of this disorder. Such data suggest a unique Prader-Willi behavioral phenotype and have important implications for further research on this and other genetic mental retardation syndromes.

Comparing Child Behavior Checklist domains across groups, we found that subjects with Prader-Willi syndrome had significantly higher total, internalizing, and externalizing scores, and a remarkably high proportion (72%) showed clinically elevated total *t* scores. Relative to two different peer groups, then, children with Prader-Willi syndrome appear to be at increased risk for significant behavioral

dysfunction, underscoring their needs for intensive, long-term management and care (Dykens & Cassidy, 1996).

Further light is shed on this increased risk by examining specific behaviors across groups. Scores from 12 behaviors were significantly higher in subjects in the Prader-Willi group relative to subjects in *both* the Down syndrome and nonspecific comparison groups. Some of these are quite predictable, as they relate to the syndrome's characteristic hyperphagia and excessive daytime sleepiness (e.g., Hertz, Cataletto, Feinsilver, & Angulo, 1993). Other elevated behaviors involve repetitive themes, specifically skin-picking, obsessions, compulsions, and talking too much. Indeed, although food obsessions have long been appreciated in Prader-Willi syndrome, recently investigators have identified increased rates of obsessions and compulsions not related to food, such as hoarding, rewriting and redoing, needing to tell or ask, and ordering and arranging (Dykens et al., 1996). All these features—compulsions, overeating, obesity, and sleepiness—are likely associated with the Prader-Willi group's increased argumentativeness and being an easy target for teasing from peers.

The extent to which these and other behaviors best differentiate the three groups was further assessed in the stepwise discriminant analysis. Findings suggest a relatively distinct Prader-Willi behavioral phenotype, with 91% of these cases correctly classified, and just 3 of 86 comparison group subjects mistakenly assigned to the Prader-Willi group. Seven behaviors best discriminated the groups, with the Prader-Willi syndrome group being singularly high in skin-picking, overtiredness, obsessions, and talking too much, and the nonspecific group showing increased hyperactivity and a low incidence of speech problems.

The discriminant analysis was also quite accurate in predicting membership into the Down syndrome group, with 80% of Down syndrome cases correctly classified. Participants with Down syndrome showed low impulsivity and were not particularly talkative, which may reflect the relative weaknesses in expressive language and grammar shown by many children with Down syndrome (Fowler, 1990; Miller, Leddy, Miolo, & Sedey, 1995). Other behavioral concerns in the Down syndrome group include high rates of speech problems, stubbornness, preferences to be alone, difficulties concentrating, and disobedience. Relative to their peers, however, subjects in the Down syndrome group showed the lowest domain scores, and only 23% had clinically elevated total scores. Other researchers have also found lower rates of psychopathology among children with Down syndrome (e.g., Meyers & Pueschel, 1991), yet increased risks of depression among adults with Down syndrome (Collacott, Cooper, & McGrother, 1992; Warren, Holroyd, & Folstein, 1989). Interestingly, children in the Down syndrome group were unique in the significant relation found between advancing age and internalizing problems, particularly withdrawal and anxiety/depression. Although preliminary, these age-related shifts may be harbingers of adult-onset depression or of the Alzheimer-like dementia seen in many adults with Down syndrome (Zigman, Schupf, Zigman, & Silverman, 1993).

Although promising, this study is limited in several ways. First, rates of Child Behavior Checklist internalizing symptoms may have been underestimated because informants such as parents often underreport internal states relative to overt behavior. Second, we relied on a single informant measure and did not directly observe the maladaptive behaviors under study. Child Behavior Checklist findings were, however, consistent with previous reports of characteristic features in Prader-Willi syndrome and Down syndrome as well as with our clinical observations of these populations.

A third limitation is that we did not match across the three groups on degree of obesity. Associations between degree of obesity and maladaptive behavior have

yet to be determined in individuals with Prader-Willi syndrome. Although no relations between BMIs and Child Behavior Checklist domains were found among children in this study, preliminary findings among adults suggest an inverse relation. Specifically, adults with lower BMIs may show increased internalizing difficulties, such as depression, distress, anxiety, and distorted thinking relative to adults with higher BMIs (Dykens & Cassidy, 1995). Such findings underscore the need for future studies in which investigators control for degree of obesity across subjects with Prader-Willi syndrome and other etiologies.

Despite these limitations, findings have implications for future research on behavioral phenotypes in general and dual diagnosis in particular. As little phenotypic work has yet been done to compare various syndromes, questions remain unanswered regarding the relative uniqueness of many behavioral phenotypes. The uniqueness question is important, as some workers posit that a behavioral phenotype only exists when all or most people with a genetic syndrome show a specific and unique behavioral outcome (Flint & Yule, 1994). Yet even unique or hallmark features, such as hyperphagia in Prader-Willi syndrome, are not expressed the same way across all individuals with the syndrome and, instead, vary in their severity, age of onset, and developmental course. Further, as demonstrated in this study, although some behavioral features of a syndrome may prove relatively unique, this does not necessarily hold true for other behaviors in the same syndrome.

In this way, then, behavioral phenotypes may best be viewed as "the heightened probability or likelihood that people with a given syndrome will exhibit certain behavioral or developmental sequelae relative to those without the syndrome" (Dykens, 1995, p. 523). All genetic syndromes have phenotypes—vulnerabilities to some behaviors more than others. Some of these behaviors, although characteristic of a syndrome, are likely to be shared. In the present study, for example, we found high rates of stubbornness, tantrums, mood changes, and poor concentration across subjects in all three groups. Yet other behaviors are relatively unique, or less likely to be seen in other genetic groups or in those with nonspecific mental retardation, such as skin-picking in Prader-Willi syndrome. In the probabilistic view of phenotypes, then, behaviors within a specific syndrome vary in their likeliness to be shared. Syndromic behaviors also vary in their degree of expression; not every child with Prader-Willi syndrome showed skin-picking or compulsions. Using a probabilistic view, then, researchers can examine genetic and psychosocial sources of within-syndrome individual differences.

The relative uniqueness of behavioral phenotypes also has important implications for research in dual diagnosis. It is now well-appreciated that people with mental retardation are at increased risk for behavioral and psychiatric dysfunction. Yet the exact causes of this increased risk are unknown and likely are associated with a confluence of psychological, cognitive, social, neurological, and genetic factors (e.g., Matson & Sevin, 1994). The search for causal mechanisms may be hindered by heterogeneity in subject groupings; indeed, in the vast majority of research in dual diagnosis, investigators have used groups of subjects with mixed or unknown etiologies (Dykens, 1996). By using homogeneous groups of people with well-delineated syndromes, researchers may identify possible genetic contributions to psychiatric vulnerabilities as well as differentiate genetic from psychosocial risk factors.

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Received 8/25/96, first decision 11/29/96, second decision 1/23/97, accepted 3/21/97.

This research was supported, in part, by National Institute of Child Health and Human Development Grant No. 03008. The authors thank Robert M. Hodapp for his careful reading of an earlier draft of this manuscript. We are also grateful to the Down Syndrome Association of Los Angeles and the Connecticut and California chapters of the Prader-Willi Syndrome Association (USA) for their continued involvement in our work. Requests for reprints should be sent to Elisabeth Dykens, Prader-Willi Syndrome Clinic, Neuropsychiatric Institute, University of California at Los Angeles, 760 Westwood Plaza, Los Angeles, CA 90024-1759.