Maladaptive and Compulsive Behavior in Prader-Willi Syndrome: New Insights From Older Adults

Elisabeth M. Dykens
John F. Kennedy Center for Research on Human Development, Peabody College, Vanderbilt University

Abstract
Although maladaptive and compulsive behaviors are increasingly well-described in young persons with Prader-Willi syndrome, it is unclear how these problems manifest in older adults with this syndrome. In Part I, I compared maladaptive and compulsive behaviors in 45 older adults with Prader-Willi syndrome (ages 30 to 50 years) to 195 children, adolescents, and young adults. Young adults were at highest risk for problems. In contrast, maladaptive and compulsive symptoms diminished significantly in older adults with Prader-Willi syndrome. In Part II, I examined predictors of problems other than age: IQ, gender, BMI, and— for adults—living status. Gender and BMI were significant predictors of skin-picking and other symptoms. Possible reasons are discussed for sweeping declines in problems among older adults.

Genetic and behavioral researchers alike are increasingly examining the unique features of persons with Prader-Willi syndrome; indeed, over the last 2 decades, the number of published articles on Prader-Willi syndrome has more than doubled (Dykens & Hodapp, 2001). Although the majority of these articles are focused on the genetic mechanisms of Prader-Willi syndrome, a flurry of studies depicts a “classic” Prader-Willi syndrome behavioral phenotype. Compared to others with intellectual disabilities, for example, persons with Prader-Willi syndrome are prone to hyperphagia; temper tantrums; impulsivity; skin-picking; and repetitive, compulsive-like behaviors such as hoarding, ordering, and arranging (Clarke et al., 2002; Curfs, Verhulst, & Frys, 1991; Dykens, Leckman, & Cassidy, 1996; Dykens & Kasari, 1997; Einfeld, Smith, Durvasula, Florio, & Tonge, 1999; Stein, Keatings, Zar, & Hollander, 1994).

The majority of these behavioral studies, however, have been focused on children, primarily because early childhood is marked by the emergence of very challenging behaviors. Hyperphagia and food-seeking typically begin between 2 and 6 years of age as do tantrums and self-injurious behavior, predominantly skin-picking (Dimitropoulos, Feurer, Butler, & Thompson, 2001). Internalizing problems, such as feelings of negative self-worth, withdrawal, and sadness, may also increase across the childhood years (Dykens & Cassidy, 1995).

In contrast, much less is known about behavioral difficulties in adults with Prader-Willi syndrome. Clarke, Boer, Chung, Sturme, and Webb (1996) examined 30 adolescents and adults with Prader-Willi syndrome with the Aberrant Behavior Checklist and found high rates of tantrums, skin-picking, impulsiveness, lability, inactivity, and repetitive speech, even when compared to others with developmental delay. Similarly, Einfeld, Tonge, Turner, Parmenter, and Smith (1999) examined 35 adolescents with Prader-Willi syndrome over a 4-year period with the Developmental Behaviour Checklist. They found high rates of problems in adolescents with Prader-Willi syndrome relative to control subjects as well as non-significant increases in overall maladaptive scores in the Prader-Willi syndrome sample. Finally, Dykens, Hodapp, Walsh, and Nash (1992) examined
21 adults with Prader-Willi syndrome residing in group homes and found high rates of externalizing as opposed to internalizing problems on the Child Behavior Checklist.

Although studies to date provide snapshots of discrete developmental epochs in Prader-Willi syndrome, researchers have yet to link the child, adolescent, and adult years together for a broader view of how maladaptive behaviors might change over the entire life span. Adopting this broader view, it may be that certain maladaptive behaviors in Prader-Willi syndrome persist over time, from early childhood to old age, whereas others intensify or diminish over the course of development.

In a first attempt to examine maladaptive behavior over time, Dykens et al. (1996) found age-related increases in the overall number, but not severity, of non-food compulsive symptoms in 91 persons ages 5 to 47 years. Regarding food-related problems, some clinicians suggest that older adults with Prader-Willi syndrome “mellow” in their drive for food (Waters, 1990). Yet persistent and severe food-seeking behaviors were noted in 3 case reports of older women with Prader-Willi syndrome ages 54, 69, and 71 years who had been residing in large institutions for most of their lives (Carpenter, 1994; Goldman, 1988). Similarly, Dykens et al. (1992) found that overeating, skin-picking, stubbornness, hoarding, and tantrums were elevated across Prader-Willi syndrome participants in their teens, 20s, and 30s. Those in their 30s, however, exhibited much less destructive, impulsive behavior than did their younger cohorts, and they also had higher rates of underactivity, fatigue, and withdrawal.

In all this work, however, researchers have yet to rigorously assess older adults with Prader-Willi syndrome, primarily those who are in their 30s, 40s, or older. Indeed, most adults in Clarke et al.’s (1996) study were in their teens or 20s, with just 5 adults in their 30s. Similarly, Dykens et al. (1992) included only 6 participants in their 30s.

It is unclear whether older adults have not been studied because of the long-standing research focus on children or because of a higher death rate in older versus younger persons with Prader-Willi syndrome. Using a population-based cohort in the United Kingdom, Whittington et al. (2001) estimated a 3% annual death rate overall for persons with Prader-Willi syndrome, and a 7% per year death rate for adults ages 30 years and older. Early death in Prader-Willi syndrome adults is primarily due to complications arising from obesity, especially Type 2 diabetes and respiratory problems (Butler et al., 2002; Smith, Loughnan, & Einfeld, 2001).

In Part I of this study I compare 45 older adults with Prader-Willi, ages 30 to 50, to 195 children, adolescents, and young adults with Prader-Willi syndrome, thereby painting a broader, life-span picture of maladaptive and compulsive behavior in Prader-Willi syndrome than has previously been described. I expected to find age-related increases in maladaptive behavior and compulsivity across the child and adolescent years, but did not know how problems of young or older adults might compare to those of their younger counterparts. In Part II, I identify correlates of behavioral problems and compulsivity, including IQ, gender, degree of obesity, and living status.

**Method**

**Participants**

Participants were 240 persons (49% male, 51% female) with Prader-Willi syndrome ages 3 to 50 years ($M = 20.78$ years, standard deviation $[SD] = 10.81$). To identify developmental trends across the entire 3- to 50-year cohort, I divided the 240 participants into four age groups based on developmental periods. The young group consisted of 56 young children ages 3 to 9.99 years; the adolescent group, 80 older children and adolescents ages 10 to 19.99 years; the young adult group, 59 persons ages 20 to 29; and the older group, 45 older adults ages 30 to 50 years. Mean ages and the gender composition for each group are shown in Table 1.

Participants were recruited from several sources. Twenty-eight percent (28%) participated in research conducted either at the University of California-Los Angeles (UCLA) or at annual conferences of the National Prader-Willi Syndrome Association. An additional 30% were interviewed during routine visits to the Lili Claire-UCLA Behavior Genetics Clinic, and 42% were obtained through mailings to families living in or out of state with the assistance of either the California Prader-Willi Foundation or the National Prader-Willi Syndrome Association. To rule out an ascertainment bias, we compared maladaptive and compulsive behaviors across these three recruitment sources (research appointment, clinic, mailed survey). None of these ANOVAs were significant; therefore, participants were combined
Older adults with Prader-Willi syndrome  

E. M. Dykens

Table 1. Participant Characteristics by Group

<table>
<thead>
<tr>
<th>Groupa</th>
<th>n</th>
<th>M/F</th>
<th>Age</th>
<th>IQ</th>
<th>BMIb</th>
<th>Obese (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>56</td>
<td>35/21</td>
<td>6.49</td>
<td>1.85</td>
<td>71.02</td>
<td>16.48</td>
</tr>
<tr>
<td>2</td>
<td>81</td>
<td>31/50</td>
<td>14.45</td>
<td>2.80</td>
<td>62.97</td>
<td>16.26</td>
</tr>
<tr>
<td>3</td>
<td>58</td>
<td>31/27</td>
<td>23.89</td>
<td>2.45</td>
<td>66.20</td>
<td>13.19</td>
</tr>
<tr>
<td>4</td>
<td>45</td>
<td>19/26</td>
<td>36.63</td>
<td>5.52</td>
<td>65.17</td>
<td>11.91</td>
</tr>
<tr>
<td>Total</td>
<td>240</td>
<td>116/124</td>
<td>20.78</td>
<td>10.81</td>
<td>66.34</td>
<td>15.53</td>
</tr>
</tbody>
</table>

aGroup 1, 3 to 9 years; Group 2, 10 to 19 years; Group 3, 20 to 29 years; Group 4, 30 to 50 years. bBody Mass Index.

across the three recruitment sources and subsequently divided into age groups.

Data pertaining to diagnoses of Prader-Willi syndrome were derived from parents or caregivers. For participants who were directly interviewed (58% across research and clinic programs), diagnostic information was confirmed by obtaining copies of previous lab reports; we were unable to obtain test confirmations for remaining cases. Approximately 47% of the sample reported that their offspring had paternal deletions at 15q11–13, consistent with the Prader-Willi syndrome diagnosis; 10% reported maternal uniparental disomy; 21% reported having a positive blood test but not recalling or knowing the genetic subtype; and 21% had clinical diagnoses and no recent genetic testing. Three participants (1%) had other chromosome 15 anomalies (e.g., translocations) that resulted in a clinical diagnosis of Prader-Willi syndrome.

The Kaufman Brief Intelligence Test (K-BIT; Kaufman & Kaufman, 1990) was used to obtain IQs of approximately 58% of cases, and for the remaining participants, parental reports of their offspring’s most recent intelligence testing were employed. The IQs obtained with the K-BIT (M = 63.78, SD = 14.64) were similar to those obtained via parent report (M = 65.27, SD = 12.79), and both were in the range considered typical of the Prader-Willi syndrome population. Table 1 depicts the mean IQ for each age group; IQs did not significantly differ across the four age groups nor were there gender differences in IQ.

Degree of obesity was determined by the Body Mass Index—BMI (weight in kilos/height in meters²). Mean BMIs for each age group are shown in Table 1. Young children had significantly lower BMIs than did those in remaining groups, F(3, 220) = 6.18, p < .001. Body Mass Indexes were not significantly related to gender or IQ, but among the two groups of adult participants, those residing in out-of-home placements (n = 56) had significantly lower BMIs than those living at home (n = 46), with means of 30.76 and 39.14, respectively, t(100) = 3.96, p < .001.

The majority of children and adolescents in Groups 1 and 2 were obese (73% and 71%, respectively), defined as BMIs above the 95th percentile, as based on each child’s age- and gender-specific BMI (Centers for Disease Control, 2000). Obesity among adult participants was defined as having a BMI equal to or greater than 30 (National Institutes of Health, 1998). Among adults in their 20s, 53% were obese; and among older adults ages 30 to 50 years, 58% were obese.

Measures and Procedure

Behavioral measures were administered either via the mail or in direct interviews of the parents of the participants with Prader-Willi syndrome and then checked for completeness and accuracy by trained research assistants. For participants with research or clinic appointments, research assistants obtained participants’ heights and weights and individually administered the K-BIT (Kaufman & Kaufman, 1990). The K-BIT is used to assess cognition in persons ages 4 years through adulthood and provides standard scores for Vocabulary, Matrices, and a composite IQ. The K-BIT has been successfully used in previous studies of children and adults with mental retardation (e.g., Dykens, 2002). All parents were asked for basic information about their child (age, gender, IQ, diagnostic history, height, weight, living status) and asked to complete the following instruments:

Yale–Brown Obsessive Compulsive Scale. Parents completed an informant version of the Yale-Brown Obsessive Compulsive Scale (Goodman et al., 1989), which has good reliability and validity.
(Taylor, 1995). Although this scale has yet to be independently validated in samples with mental retardation, I used it in this study to identify compulsive symptoms, not to make diagnoses of obsessive-compulsive disorder. This version of the Yale-Brown Obsessive Compulsive Scale contains 30 symptoms rated as being present ever or in the last week, and data analyses were based on current symptomatology. Informants also rate the extent to which symptoms are time-consuming, distressful, and cause social or adaptive impairment (0 = none to 5 = extreme). In addition to the overall number and severity of symptoms, skin-picking was examined as a separate item because it has substantial unique variance that seems to distinguish it from all other compulsive behaviors in Prader-Willi syndrome (Feurer et al., 1998). The presence or absence of skin-picking in the last week was identified, as were four Obsessive Compulsive Scale subdomain scores (as per Leckman et al., 1997, and Baer, 1994): Obsessions and Checking, Symmetry and Ordering, Cleaning and Washing, and Hoarding.

Child Behavior Checklist (Achenbach, 1991). In this widely used checklist, parents are asked to rate 112 problem behaviors on a 3-point scale: 0 (not true), 1 (somewhat or sometimes true), and 2 (very true or often true). Although the checklist has been used in studies of children with mental retardation, the factor structure and norms for adults with mental retardation have not been established. In this study, therefore, I used raw scores instead of the age-referenced scaled scores provided by the Child Behavior Checklist. The instrument contains an Internalizing factor (anxious/depressed, somatic complaints, withdrawn clinical scales), Externalizing factor (noncompliant and aggressive behavior clinical scales), and three additional clinical scales (Social, Thought, and Attention Problems). Raw scores from the Total score and Internalizing and Externalizing factors were used in data analyses; when these were significant, follow-up analyses were conducted with specific clinical scales.

Results

Part I: Developmental Trends

Child Behavior Checklist Total and factor scores. Analyses of variance (ANOVAs) were used to compare the Total and Internalizing and Externalizing factor scores across the four age groups. Significant age effects were found for all three of these scores. Table 2 depicts the means and SDs for each score by age group as well as $F$ and $p$ values. Student Newman-Keuls post-hoc tests revealed that young children and older adults had significantly lower Externalizing and Total scores than did adolescents and young adults. On the Internalizing factor, young children scored significantly lower than did individuals in all remaining groups.

Child Behavior Checklist clinical scales. As significant group differences were found using the Total and factor scores, follow-up analyses were conducted with the clinical scales, using a Bonferroni corrected value of $p$ less than .006 (.05/8 clinical scales). Four of the 8 clinical scales showed significant group differences at the .006 level, and Table 2 depicts the means and $F$ and $p$ values for the clinical scales. Student Newman-Keuls post-hocs revealed that young adults in their 20s scored significantly higher than all remaining age groups on the Withdrawn and Delinquent scales. Young children scored lower than adolescents and young adults on the Withdrawn scale and lower than all remaining groups on the Delinquent and Anxious scales. On the Social Problems scale, young children and older adults scored lower than did adolescents and young adults.

Yale-Brown Obsessive Compulsive Scale total scores. Using ANOVAs, I found significant age effects for the number of compulsive symptoms and severity of symptoms, $F$s(3, 235) = 3.05 and 8.15 and $p$s < .05 and .001, respectively. Figure 1 depicts mean scores for the number and severity of compulsive symptoms across age groups. Student Newman-Keuls post-hocs revealed that the number of compulsive symptoms was similarly low in young children and in the older adult group, and these were significantly lower than the 20-year-old group. A similar pattern was seen in the severity of compulsive symptoms, with those in their 20s scoring higher than the other age groups.

Yale-Brown Obsessive Compulsive Scale subdomains. Because total scores on this instrument showed significant group differences, I conducted follow-up analyses of subdomains, using a Bonferroni corrected value of $p$ less than .01 (.05/5 subdomains or item). As shown in Table 3, significant differences were found in 2 of the 4 subdomains and in the skin-picking item. Young adults in their 20s had significantly higher Hoarding scores than did all remaining age groups. Compared to all other groups, the Ordering and
Table 2. Mean CBCL Scores by Group and $F$ Tests

<table>
<thead>
<tr>
<th>CBCL</th>
<th>Group 1</th>
<th></th>
<th>Group 2</th>
<th></th>
<th>Group 3</th>
<th></th>
<th>Group 4</th>
<th></th>
<th>$F$</th>
<th>Posthoc</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Mean</td>
<td>SD</td>
<td>Mean</td>
<td>SD</td>
<td>Mean</td>
<td>SD</td>
<td>Mean</td>
<td>SD</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>54.13</td>
<td>23.13</td>
<td>67.13</td>
<td>26.95</td>
<td>74.15</td>
<td>21.25</td>
<td>59.06</td>
<td>22.54</td>
<td>7.55***</td>
<td>1 &lt; 2, 3; 4 &lt; 3</td>
</tr>
<tr>
<td>Internalizing</td>
<td>9.53</td>
<td>7.02</td>
<td>14.32</td>
<td>8.43</td>
<td>16.57</td>
<td>7.96</td>
<td>14.37</td>
<td>7.03</td>
<td>8.25***</td>
<td>1 &lt; 2, 3, 4</td>
</tr>
<tr>
<td>Externalizing</td>
<td>15.37</td>
<td>9.13</td>
<td>19.97</td>
<td>9.85</td>
<td>22.15</td>
<td>8.95</td>
<td>16.57</td>
<td>8.27</td>
<td>6.49**</td>
<td>1 &lt; 2, 4; 4 &lt; 3</td>
</tr>
<tr>
<td>Clinical scales</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Anxious/Depressed</td>
<td>2.80</td>
<td>3.16</td>
<td>5.84</td>
<td>4.87</td>
<td>7.26</td>
<td>4.71</td>
<td>6.33</td>
<td>3.95</td>
<td>11.21***</td>
<td>1 &lt; 2, 3, 4</td>
</tr>
<tr>
<td>Withdrawn</td>
<td>4.09</td>
<td>2.76</td>
<td>5.32</td>
<td>3.03</td>
<td>6.35</td>
<td>3.16</td>
<td>4.82</td>
<td>2.72</td>
<td>5.91**</td>
<td>3 &gt; 1, 2, 4; 1 &lt; 2, 3</td>
</tr>
<tr>
<td>Somatic</td>
<td>2.68</td>
<td>2.63</td>
<td>3.26</td>
<td>2.35</td>
<td>3.08</td>
<td>2.34</td>
<td>3.22</td>
<td>2.43</td>
<td>.71</td>
<td></td>
</tr>
<tr>
<td>Aggressive</td>
<td>12.50</td>
<td>7.36</td>
<td>14.89</td>
<td>7.59</td>
<td>15.19</td>
<td>6.74</td>
<td>11.69</td>
<td>6.52</td>
<td>3.27</td>
<td></td>
</tr>
<tr>
<td>Delinquent</td>
<td>2.87</td>
<td>2.45</td>
<td>5.21</td>
<td>3.13</td>
<td>6.96</td>
<td>3.35</td>
<td>4.88</td>
<td>2.57</td>
<td>18.40***</td>
<td>3 &gt; 1, 2, 4; 1 &lt; 2, 3, 4</td>
</tr>
<tr>
<td>Social</td>
<td>6.28</td>
<td>2.65</td>
<td>8.82</td>
<td>2.85</td>
<td>9.20</td>
<td>2.92</td>
<td>6.51</td>
<td>2.93</td>
<td>16.60***</td>
<td>1 &lt; 2, 3; 4 &lt; 2, 3</td>
</tr>
<tr>
<td>Thought</td>
<td>3.30</td>
<td>2.06</td>
<td>3.85</td>
<td>2.16</td>
<td>4.45</td>
<td>2.11</td>
<td>3.20</td>
<td>7.82</td>
<td>3.88</td>
<td></td>
</tr>
<tr>
<td>Attention</td>
<td>6.59</td>
<td>3.21</td>
<td>7.47</td>
<td>4.15</td>
<td>9.10</td>
<td>2.97</td>
<td>7.55</td>
<td>4.09</td>
<td>2.05</td>
<td></td>
</tr>
</tbody>
</table>

Note. CBCL = Child Behavior Checklist.

*Group 1, 3 to 9 years; Group 2, 10 to 19 years; Group 3, 20 to 29 years; Group 4, 30 to 50 years.

**$p < .006$. ***$p < .001$. 
Older adults with Prader-Willi syndrome

E. M. Dykens

Figure 1. Mean number and severity of Yale-Brown Obsessive-Compulsive Scale (Y-BOCS) symptoms across four age groups of participants with Prader-Willi syndrome. Black bars = number of compulsions, gray bars = severity of compulsions.

Arranging domain was significantly lower in older adults. As the SDs of the Obsessive Compulsive Scale subdomain scores were often as large or larger than the means, I re-examined subdomain differences using the Kruskal-Wallis nonparametric test. The pattern of findings was the same (because means are more understandable, I reported means instead of ranks in Table 3).

The skin-picking item showed significant age effects; Table 3 shows frequencies across age groups. Follow-up analyses revealed that the frequency of skin-picking in older adults was similar to that of young children and that older adults skin picked less often than did adolescents and young adults in their 20s, $\chi^2(1, N = 184) = 10.80, p < .01$.

Part II: Correlates

To identify correlates of maladaptive and compulsive behaviors above and beyond the effect of age, I conducted hierarchical regression analyses for two groups: children and adolescents (Groups 1 and 2) and adults (Groups 3 and 4). Groups were combined to limit the number of analyses and because predictors differed slightly across age groups. For all age groups, predictors included IQ, gender, and BMI; for adults, living status was included as an additional predictor.
Outcomes were the factor scores from the Child Behavior Checklist and the Obsessive Compulsive Scale. For all regressions, the contributions of other predictors were assessed after the effect of age was taken into account.

**Children—Adolescents.** For children and adolescents, no significant regressions were found with the Child Behavior Checklist, whereas one significant regression emerged on the Obsessive Compulsive Scale. Skin-picking was predicted by gender and BMI, and these accounted for 9% of the variance, after the contributions of age were considered, $F(4, 113) = 5.63, p < .001$. Gender accounted for 6% of the variance, and follow-up analyses revealed that girls showed more frequent skin-picking than did boys ($M_5 = .77$ and $.52$, respectively), $t(133) = -3.11, p < .01$. Skin-picking was also predicted by the BMI, and this negative association, $\beta = -.19, p < .05$, accounted for 3% of variance.

**Adults.** After age was considered, significant effects of other predictors were found for Child Behavior Checklist Externalizing behaviors and on the Obsessive-Compulsive Scale, for skin-picking, the severity of compulsive symptoms, and hoarding, $F_5(5, 99) = 6.77, 3.73, 5.37, and 4.21$, respectively, $p < .01$. Gender explained 6% of Externalizing variance. Because the overall domain was significant, the two Externalizing subdomains were examined. Gender emerged as a significant predictor for the Aggression subdomain, with males scoring higher than did females ($M_5 = 15.56$ vs. 11.87), $t(101) = 2.83, p < .01$.

For skin-picking, the BMI was the sole significant predictor, and this inverse relationship, $\beta = -.26, p < .01$, accounted for 7% of skin-picking variance. The severity of compulsive symptoms was predicted by gender (6% of variance) and living status (5% of variance). Follow-up analyses revealed that males had significantly higher severity scores than did females ($M_5 = 6.04$ vs. 4.41), $t(99) = 2.68, p < .01$, and that those living in programs had significantly higher severity scores than did those residing at home ($M_5 = 6.92$ vs. 5.51), $t(99) = -2.11, p < .05$. Because the overall severity score of the Obsessive Compulsive Scale was significant, I examined the four subdomains of this scale; only the Hoarding subdomain was significant. Hoarding was predicted by BMI, and this inverse relationship, $\beta = -.27, p < .01$, accounted for 7% of Hoarding variance.

**Discussion**

Maladaptive and compulsive behaviors showed unexpected peaks and dips across a wide age range of persons with Prader-Willi syndrome. As predicted, maladaptive and compulsive behaviors generally increased from the child to adolescent years. Unlike previous depictions of the Prader-Willi syndrome behavioral phenotype, however, it appears that persons with this syndrome in their young adult years are at high risk for maladaptive behaviors and compulsive symptoms, even as compared to the adolescent period. In contrast, both maladaptive and compulsive behaviors diminished to an astonishing degree in older adults with Prader-Willi syndrome.

Consistent with previous research, children with Prader-Willi syndrome seem to exhibit more maladaptive behaviors as they develop. Age-related increases were found in skin-picking, compulsive symptoms (such as hoarding and ordering), and externalizing symptoms (such as food theft and lying—predominantly about food). Further, participants showed age-related increases in internalizing problems, especially anxiety and sadness. Age-related increases in sadness and low self-worth were previously found in children and adolescents (Dyken & Cassidy, 1995), and age-related increases in compulsive symptoms, tantrums, and skin-picking have also been identified in both preschool and older children (Dimitropoulos et al., 2001; Dyken & Kasari, 1997).

An unexpected finding, however, was the consistent pattern of elevated problems among young adults in their 20s. With few exceptions, these young adults scored highest on both the Child Behavior Checklist and the Obsessive Compulsive Scale. Indeed, they had significantly higher scores than did all other age groups in delinquent or noncompliant behaviors (including food theft), withdrawal, hoarding, and the severity of compulsive symptoms. Further, compared to either young children or older adults, those in their 20s were highest in their externalizing and total Child Behavior Checklist maladaptive behaviors, social problems, and the number of different compulsive symptoms.

The reasons for elevated problems in young adults with Prader-Willi syndrome are unknown.
and may be associated with a host of new stressors often encountered by these individuals and their families. These include graduation from school and the need to find other activities, such as work, vocational training, or social/recreational programming. Due to their child’s maladaptive and food-seeking behaviors, many families encounter difficulties both obtaining and retaining appropriate programs in this “school to work” transitional period (Seguin & Hodapp, 1998). Young adults may also be more acutely aware of their differences relative to others because they are generally unable to emulate siblings or peers who are driving, getting married, having children, and living independently. Young adulthood may, thus, be a period of new psychosocial adjustments and heightened behavioral problems for many with Prader-Willi syndrome, although not necessarily for persons with heterogeneous causes of mental retardation (see Tonge & Einfeld, 2003).

In sharp contrast to young adults, older adults with Prader-Willi syndrome demonstrated markedly reduced rates of both maladaptive and compulsive behaviors. Generally, this older age cohort showed levels of problems similar to those of young children with the syndrome, much below those of the adolescent and/or young adult groups. Relative to adolescents and young adults, these older adults showed significantly less skin-picking as well as significantly fewer externalizing symptoms, social problems, total maladaptive behaviors, and number of compulsive symptoms. Further, compared to all the other groups, even young children, the oldest group showed significantly less ordering and arranging. Many older adults with Prader-Willi syndrome ages 30 to 50, thus, seem to “mellow” behaviorally, showing considerably less of the syndrome’s characteristic features, including skin-picking, compulsivity, and noncompliance.

The dampening of problems in older adults with Prader-Willi syndrome may differ from others with mental retardation, including those with genetic syndromes. In Down syndrome, for example, advancing age is associated with increased risks of depression and Alzheimer-type dementia (e.g., Collacott, Cooper, & McGrother, 1992; Zigan, Silverman, & Wisniewski, 1996). Among those with mental retardation in general, age-related increases in psychiatric problems such as depression and anxiety have been found for clinical as well as epidemiological samples (Cooper, 1997; Day, 1987). Although comparative studies are needed to confirm this finding, being an older adult with Prader-Willi syndrome may confer some protection against significant mental health problems seen in other adults with mental retardation.

It is not yet clear why older adults with Prader-Willi syndrome show such sweeping, across-the-board drops in their behavioral difficulties. One possibility relates to selective survival issues, and the idea that those who live longer may differ from those who do not (Widaman, Birthwick-Duffy, & Powers, 1994). Most deaths in Prader-Willi syndrome relate to complications of obesity, and there is a higher death rate in older versus younger adults with this disorder (7% vs. 5%, respectively; Whittington et al., 2001). Older adults with Prader-Willi syndrome may, thus, be less obese; show fewer obesity-related medical problems; and, although speculative, they may be doing better behaviorally as well. Although this is a reasonable hypothesis, the majority of adults (58%) in the older sample ages 30 to 50 years were obese and, therefore, at risk for food-seeking behaviors and medical complications related to obesity. Both obese and thinner participants were included in the older group, making it less likely that findings are completely explained by the selective survival hypothesis.

Older adults may also show less compulsive and maladaptive behaviors because of unknown hormonal or physiological factors or because they are finally reaping the benefits of being exposed to years and years of sustained behavioral and dietary interventions. Age may, thus, be a proxy for the cumulative exposure to dietary, behavioral, and other interventions routinely given to persons with Prader-Willi syndrome (Hanchett & Greenswag, 1998).

One such intervention, more common in the adult years, is out-of-home placement in a group home or residential program. Based on anecdotal and clinical impressions, Hanchett and Greenswag (1998) observed that behavioral problems typically decline when adults with Prader-Willi syndrome enter structured group living situations. If so, then reduced problems in the older adults in this study may be attributed to the fact that many of them (71%) resided in out-of-home placements.

Although a reasonable explanation, residential status did not emerge as a significant predictor of psychopathology among adults in this study. On the one hand, relative to those at home, adults with Prader-Willi syndrome in out-of-home
placement had significantly higher compulsive behavior severity scores. Higher symptom severity may have been a contributing factor in the decision to seek out-of-home placement. On the other hand, adults in out-of-home placement had significantly lower BMIs than did their counterparts residing at home (see also Greenswag, 1987). Out-of-home placement was, therefore, associated with being thinner, yet it was not a significant predictor of externalizing or internalizing problems or compulsivity.

In Part II of the study other correlates were assessed, and gender and BMI were found to be significant predictors of compulsive and maladaptive behavior, even after accounting for the effects of age. Among children and adolescents, girls were more apt to skin-pick than were boys, and among adults, males had higher Externalizing, Aggression, and Compulsive Severity scores than did females. In each instance, gender accounted for 6% of variance. Although males have previously been shown to have higher Compulsive Symptom Severity scores (Dykens et al., 1996), other researchers have not found gender differences in skin-picking, aggression, or externalizing problems (Dykens & Kasari, 1997; Einfeld et al., 1999; Symons, Butler, Sanders, Feurer, & Thompson, 1999). Future studies are needed to clarify these discrepancies in gender differences.

Interestingly, BMI was inversely associated with skin-picking in children and adults, accounting for 4% and 7% of variance, respectively. Adults also showed an inverse relationship between BMI and hoarding. Similarly, thinner as opposed to obese persons with Prader-Willi syndrome may have increased anxiety, sadness, distress, confused thinking, and other psychiatric concerns (Dykens & Cassidy, 1995; Whitman & Accardo, 1987). Collectively, findings suggest a counterintuitive relationship, with elevated compulsivity and internalizing types of symptoms in thinner persons with Prader-Willi syndrome.

Both psychological and physiological mechanisms may be implicated in these BMI findings. Achieving adequate weight control is inherently stressful for most persons with Prader-Willi syndrome; they are “always hungry, never full” (Prader-Willi Syndrome Association, 1999) and, thus, are in a constant state of denial. The denial and stress involved in successful weight loss may exacerbate anxiety and sadness as well as contribute to distorted thinking and compulsive behavior, such as skin-picking or hoarding.

Although speculative, this explanation garners some support from recent breakthroughs involving Prader-Willi syndrome and a novel hormone, ghrelin, which is secreted by the stomach and is a powerful stimulant of appetite and food consumption. Compared to obese and lean control subjects, children and adults with Prader-Willi syndrome have markedly high plasma levels of ghrelin (Cummings et al., 2002; Delparigi et al., 2002; Haqq et al., 2003). Ghrelin levels may remain high in persons with Prader-Willi syndrome even after food consumption (Delparigi et al., 2002). If eating provides persons with Prader-Willi syndrome with some psychological relief or satisfaction, however, then thinner persons may experience more tension or distress because they have high levels of ghrelin (and are just as hungry as their obese counterparts), but are presumably consuming less food or fewer calories. Such denial may, in turn, be associated with increased anxiety, sadness, distress, and compulsivity.

The elevated levels of ghrelin in Prader-Willi syndrome are similar to those found during starvation states, including those of individuals with anorexia nervosa (Delparigi et al., 2001). Notably, anorexia is also characterized by distorted perceptions, depression, compulsive behaviors, and increased risks of obsessive-compulsive disorder (e.g., Halmi et al., 2003; O’Brien & Vincent, 2003). Further studies are needed that assess BMI and ghrelin as possible moderators of distress and compulsive behaviors in persons with Prader-Willi syndrome and perhaps other populations with elevated ghrelin levels as well.

This study has several strengths and weaknesses. The most important strengths include the large number of participants in general and older adults in particular. Other strengths include the wide age range of participants and the use of standardized measures of both compulsivity and maladaptive behaviors.

Despite these strengths, the present study also has several shortcomings. First, in approximately half of the sample, I relied on parental reports of genetic subtyping, and many parents did not know or could not recall this information. Unlike prior studies, then, data were not analyzed by genetic subtype. When genetic subtypes are accurately compared, persons with maternal uniparental disomy may show less skin-picking and other maladaptive behaviors (Dykens, Cassidy, & King, 1999; Symons et al., 1999), but heightened vulnerabilities in young adulthood for psychiatric
Older adults with Prader-Willi syndrome

E. M. Dykens

disorders, such as atypical psychosis and affective disorders (Beadsmore, Dorman, Cooper, & Webb, 1998; Boer et al., 2002; Vogels, Matthijs, Legius, Devriendt, & Fryns, 2003).

A second weakness is that the residential data were preliminary in scope. More robust effects of living status may be found in future studies in which researchers assess residential programs in a fine-tuned way, including the type and duration of previous and current placements. In a similar vein, the present study lacked detailed medical histories of participants, and these data would have been helpful for evaluating the selective survival hypothesis.

Finally, data were cross-sectional, and despite the large sample, these developmental findings must be considered preliminary. Follow-up longitudinal studies are needed to clarify the peaking of problems in young adulthood and the sweeping decline of problems in mid-adulthood. It is not known how this pattern might extend into older adulthood, specifically for persons in their 50s, 60s, and beyond.

In the future researchers conducting longitudinal studies also need to assess promising correlates of longevity and positive adult behavioral outcomes in Prader-Willi syndrome, including degree of obesity, physiological and hormonal factors, medical health, treatment histories, and residential status. These studies may shed new light on risk and protective factors associated with successful aging in persons with mental retardation in general. In the meantime, findings underscore the fluid and changeable nature of syndromic behavior, and emphasize the need to adopt a broad, life-span, developmental approach in research on Prader-Willi syndrome.

References


Older adults with Prader-Willi syndrome

E. M. Dykens

Clinical Endocrinology and Metabolism, 87, 5461–5464.


Older adults with Prader-Willi syndrome

E. M. Dykens


The author thanks the many families who participated in this study, recruited with help from the National Prader-Willi Syndrome Association (USA), the California Prader-Willi Foundation, and the Lili-Claire Foundation. This work was supported by Grant R01135681 from the National Institute of Child Health and Human Development. The author thanks Robert M. Hodapp for his helpful comments on an earlier draft of this manuscript; and Beth Rosner, Tran Ly, and Jaclyn Sagun for their assistance with data collection. Requests for reprints should be sent to Elisabeth Dykens, Vanderbilt Kennedy Center, 230 Appleton Place, Peabody Box 40, Nashville, TN 37203. E-mail: elisabeth.dykens@Vanderbilt.Edu.