

MALADAPTIVE BEHAVIOR IN CHILDREN

with PWS, Down syndrome, and Non-specific 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. It thus remains unclear to what extent certain behaviors seen in people with Prader-Willi syndrome (PWS) are shared versus unique.

This study compared maladaptive behavior in 43 children with Prader-Willi syndrome, aged 4 to 19 years, to 43 age- and gender-matched youngsters with Down syndrome, and 43 matched subjects with non-specific mental retardation. As measured by the Child Behavior Checklist (Achenbach, 1991), some problems were elevated in all three groups, such as tantrums, stubbornness, and difficulties regulating attention.

Yet on average, the Prader-Willi group showed significantly more frequent and severe internalizing, externalizing, and total problem behaviors. Further, 72% of children with Prader-Willi syndrome showed scores that fell in the clinically elevated range. This rate of clinically worrisome scores was higher than rates shown by children with Down syndrome (23%) or nonspecific retardation (39%). Twelve behaviors, listed below, were significantly elevated in youngsters with Prader-Willi syndrome relative to children in both comparison groups.

	PWS	DS	NS
Skin-Picking	95%	20%	26%
Argues a lot	95%	65%	63%
Underactive	91%	35%	28%
Obsessions	88%	46%	36%
Overtired	84%	25%	23%
Overeating	80%	27%	21%
Talks a lot	74%	35%	46%
Excessive sleep	74%	11%	9%
Overweight	70%	30%	16%
Gets teased	72%	28%	32%
Compulsions	68%	32%	37%
Steals at home	58%	6%	9%

Elevated Behaviors in PWS Relative to Both

Discriminant function analyses revealed that seven behaviors predicted membership into the Prader-Willi group with 91% accuracy; these behaviors included skin-picking, overtiredness, obsessions, talking too much, impulsivity, speech concerns (all higher in PWS), and hyperactivity (lower than the other groups).

Only 3 of the 86 comparison group children were mistakenly classified in the Prader-Willi group. Findings suggest a distinctive Prader-Willi behavioral phenotype.

Though promising candidates also emerged for characteristic behaviors in Down syndrome, the Prader-Willi phenotype was more distinctive.