

The PWS Personality:

What is *within normal limits*?



Pittsburgh Partnership

Janice L. Forster, MD
Linda M. Gourash, MD

Background: Among individuals with PWS, some temperamental and behavioral characteristics are so commonly present that they are considered to be part of the **behavioral phenotype**. The behavioral phenotype of PWS defines the **PWS PERSONALITY**; it corresponds to the DSM IV Axis I diagnosis of *Personality Change Due to a Medical Condition (310.1)*. In this psychiatric disorder symptoms occur as a direct physiological consequence of a medical condition. Characteristics of this diagnostic category include affective instability, poor impulse control, behavioral outbursts of aggression or rage (out of proportion to the situation), apathy (indifference) or motivational problems, and cognitive abnormalities (such as thought distortions or impaired judgment). The symptoms of this disturbance cause developmental deviation, distress and dysfunction across family, social, and educational and/or occupational settings.

Methods: A review of the literature and the clinical experience of the authors inform this work.

Results: The psychiatric and behavioral symptoms of the PWS PERSONALITY can be organized conceptually across five domains that correlate with the diagnostic description for *Personality Change Due to a Medical Condition*. The five domains of the PWS Personality are: food related behaviors, oppositional defiant behaviors, cognitive rigidity/inflexibility, anxiety/insecurity, and skin picking.

Discussion: The PWS PERSONALITY provides a qualitative description of clinical signs and symptoms observed among all individuals with PWS. However, each individual with PWS may display their own unique pattern of these characteristics. Universally, stress can increase the intensity of symptoms in any of these domains. Further, the diagnosis of a PWS PERSONALITY does not exclude the possibility that additional psychiatric diagnoses may be warranted. For example, a child with PWS PERSONALITY may also have concurrent Attention Deficit Hyperactive Disorder, or Mood Disorder, or Anxiety Disorder or Stereotypic Movement Disorder. Proper recognition of the PWS PERSONALITY will lead to more accurate psychiatric diagnosis, inform realistic treatment goals, and guide the selection of appropriate therapeutic interventions.

References:

1. The Diagnostic and Statistical Manual of Mental Disorders, fourth edition; American Psychiatric Association, 1994.

2. Managing Prader-Willi Syndrome: A Primer for Psychiatrists; The Pittsburgh Partnership, 2005.

www.PittsburghPartnership.com

ConsultForPWS@aol.com

I. Food related behaviors

- overeating of typical food
- eating atypical food (frozen, raw, spoiled food or pet food)
- sneaking food in the home
- night time foraging in the home
- arguing or manipulating to get food
- tantruming to get food
- opportunistic food theft (shoplifting from a store or stealing food from school or work)
- planned food foraging expeditions in the neighborhood or community
- nonconfrontational, invasive food access (breaking locks on cabinets, refrigerator or freezer)
- confrontational food access (using verbal or physical threats and/or actual aggression to access food)

FREQUENCY

INTENSITY

II. Cognitive rigidity/inflexibility

- perseveration, "sticky thinking"
- difficulty with transitions or changes
- rituals
- selective interests (jig saw puzzles, word searches)
- impaired judgment
- single mindedness: difficulty taking multiple view points
- egocentrism

III. Oppositional/defiance

- noncompliance
- argumentativeness
- tantrums
- shut downs
- manipulation
- lying/confabulation

IV. Anxiety/insecurity

- stress sensitivity
- inability to tolerate uncertainty
- somatic complaints
- dependency
- constant need for reassurance
- collecting and hoarding
- affective reactivity

V. Skin picking: (occurs commonly, but not universally)

- habit behavior (opportunistic typography):
 - ✦ cuticles, finger nails
 - ✦ arms, face and scalp
 - ✦ hair pulling (scalp, eyelashes)
 - ✦ nasal septum
 - ✦ peeling skin from the soles of the feet
 - ✦ pulling out toe nails, teeth
- intense, severe, reactive
 - ✦ gouging, self mutilation
- related to chronic stress
 - ✦ rectal and/or vaginal picking

FREQUENCY

INTENSITY