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Romanian Prader-Willi Association

ASOCIACIÓN MADRILEÑA  
PARA EL SÍNDROME DE  
PRADER-WILLI



## PHENOMENOLOGY AND MANAGEMENT OF MOOD ACTIVATION IN PRADER-WILLI SYNDROME

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**INTRODUCTION:** Mood disorders with or without psychotic features have been reported in Prader-Willi syndrome (PWS). Leibenluft et al (2003) characterize symptoms of mania in juvenile onset bipolar disorder as narrow phenotype (elation, grandiosity, episodicity) or broad phenotype (irritability, emotional reactivity, chronicity). The authors review the phenomenology and management of mood activation in PWS, characterize symptoms of mania, and comment on predisposing, precipitating and perpetuating factors.

**METHODS:** The authors collect from their clinical experience with PWS a cohort of individuals with mood activation with or without psychosis.

**RESULTS:** Mood activation was observed in one third of individuals with PWS referred for psychiatric evaluation and treatment. The cohort includes 38 children and adults, ages 8-39 years; there is no gender bias, and the genotypic frequency reflects the accepted occurrence rates of deletion and UPD conditions. Within the cohort, there was no genotypic bias among those presenting with the narrow phenotype. All of these patients were psychotic, and almost all of them required mood stabilizers and antipsychotic agents to resolve their symptoms. Those presenting with the broad phenotype were more likely to have the deletion condition. Only half of these patients were psychotic, and they required less pharmacotherapy. Among the developmental cohort, more children with the deletion condition than UPD present with psychotic mood disturbance; after the developmental period, the ratio reverses. The major precipitating factor for mood activation among all groups of patients was pharmacotherapy with selective serotonin reuptake inhibitors (SSRI's). Symptoms did not remit until the SSRI was discontinued, even in the presence of environmental management.

**DISCUSSION:** Mood activation is a serious, poorly recognized problem among individuals with PWS resulting in referral for psychiatric evaluation. For the most part, mood activation is an iatrogenic problem arising from the use of SSRI medications. If the symptoms of mood disorder persist after the discontinuation of the SSRI, then anticonvulsant mood stabilizers and lithium can be used effectively in concert with environmental interventions. In these cases, mood activation may uncover an underlying diathesis for Bipolar I Disorder which is usually associated with the narrow phenotype of mania. Surprisingly, individuals with UPD were not the most likely persons in this cohort to experience the narrow phenotype of mood activation. However, individuals with deletion condition were more likely to present with the broad phenotype, especially among the youngest cohort. Symptoms of mood disorder in PWS are effectively managed with mood stabilizers in concert with environmental interventions when mood activating agents are discontinued.