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Abstract 21-1-01

Title: Medical and Psychiatric Management of Prader-Willi Syndrome

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Introduction: Prader-Willi Syndrome (PWS) is a rare neurodevelopmental disorder which occurs in roughly 1 in 22000 births (1). PWS is caused by abnormal parent-specific imprinting within the Prader-Willi critical region 15q13 on chromosome 15 (2). Most cases of PWS are due to loss of a segment of the paternal chromosome 15 (2). Children with PWS have more severe affective and somatic problems compared to controls (2). Management of PWS requires multidisciplinary care.

Case Presentation: The patient is a 19 yo Caucasian male with PWS, moderate intellectual disability, Generalized Anxiety Disorder, Impulse control disorder, Major Depressive Disorder and with comorbid diabetes. He presented with increasing apathy, tantrums, and nightmares over a several month period. During this time, his mother noted his lack of regular scheduled extracurricular activities due to disruption from the COVID-19 pandemic. He was currently treated with Fluoxetine 60 mg daily, Oxcarbazepine 300 mg twice a day, Lithium 600 mg twice a day, Bupropion XL 300 mg daily, Metformin 2000 mg daily, NovoLog 36 mg daily, and Lantus 33 mg daily. His height was 183 cm and his weight was 152 kg with a BMI of 45, indicating morbid obesity.

Discussion: PWS is a complex medical disorder with an array of somatic and behavioral problems. PWS presents in infancy as hypotonia and failure to thrive. The most common affective disorders in patients with PWS are Major depressive disorder and Bipolar disorder. Children with PWS also classically have cognitive delay. Most strikingly, defects in satiety leads to obsession with food, hyperphagia, and obesity, with the latter contributing to the development of obstructive sleep apnea (4, 5). Endocrine conditions include hypogonadism leading to sex hormone deficiency and impaired sexual development and growth hormone deficiency leading to short stature, osteoporosis or scoliosis from deficient calcium intake (5). Life expectancy of a patient with PWS depends on obesity and comorbidities. Medical supervision becomes increasingly important after age 40. (3) This case highlights the need for long-term multidisciplinary care including a comprehensive psychiatric assessment in patients with PWS.

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