What is Prader-Willi Syndrome

Prader-Willi Syndrome is a rare and very complex non-inherited genetic disorder. Symptoms associated with Prader-Willi Syndrome (PWS) are believed to be caused in part by a defect in the hypothalamus, an important supervisory center in the brain that controls metabolism of fats and carbohydrates, sugar levels in the blood, body temperature, blood pressure, heartbeat, the expression of emotions, the regulation of the sleep-wake cycle, the development of muscle tone, and many more functions of the body. The hypothalamus is connected to the pituitary gland, often called the "master gland of the body" because all the other endocrine glands depend upon its stimulation. Prader-Willi Syndrome impacts the function of the pituitary gland which controls the release of important hormones, including growth and sex hormones.

Infants with Prader-Willi Syndrome fail to thrive and require varying degrees of assistance in order to survive. Essential medical and therapeutic interventions for children with PWS include growth hormone therapy, occupational therapy, physical therapy, oral-motor and speech and language therapy, optometric or ophthalmologic care, rigorous dental care, behavioral and often psychiatric intervention, social skills therapy, and special education services.

Beginning some time in childhood, the brain fails to regulate appetite normally. For a person with Prader-Willi Syndrome there is a constant preoccupation with food accompanied by an unrelenting, overwhelming, overriding physiological drive to eat called hyperphagia. Normal satiety (the feeling of fullness after eating) does not exist.

The physiological drive to eat is so powerful that most individuals with Prader-Willi Syndrome will go to great lengths to eat large quantities of food; many try to sneak food and some may even try to steal. Some people may eat food discarded in the trash or even non-edible items. Along with hyperphagia, the metabolic rate is about half what it should be, so individuals with Prader-Willi Syndrome can gain an enormous amount of weight in a very short period of time.

There is no cure for Prader-Willi Syndrome and there is no known medication that will control or even reduce the hyperphagia symptoms. No one with Prader-Willi Syndrome is able to live independently because without 24/7/365 supervision for the entirety of their lives, individuals with PWS will die prematurely as a result choking, stomach rupture, stomach tissue necrosis, or from complications caused by morbid obesity.

Prader-Willi Syndrome is *not* an eating disorder; it is a physiological defect in the brain that causes myriad symptoms. Prader-Willi Syndrome is a life-long, life-threatening genetic disorder that requires a multi-disciplinary treatment team. The majority of individuals with Prader-Willi Syndrome test in the mildly mentally retarded range of IQ and are therefore eligible to receive services from the State of California. The remainder of individuals with Prader-Willi Syndrome should qualify to receive services through the "Fifth Category."

Parents and care providers of an individual with Prader-Willi Syndrome experience the *highest* levels of stress. Throughout their child's lifetime they will require the assistance of a multitude of professionals. Working together, we can help individuals born with Prader-Willi Syndrome live a meaningful and productive life and pursue their hopes and dreams to the full extent of their talents and capabilities.

For more information about Prader-Willi Syndrome contact

Prader-Willi California Foundation
310.372.5053 · 800.400.9994 Toll-free in CA · www.pwcf.org

Prader-Willi Syndrome Association (USA)
800.926.4797 · www.pwsausa.org

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Supporting People with Prader-Willi Syndrome