Prader-Willi Syndrome

Factsheet

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Introduction

- Documented cases of PWS go back to the 17th Century
- In 1956, PWS was identified by A. Prader, H. Willi and A. Labhart
- Prader-Willi Syndrome (PWS) is a Endocrine Hypothalamic Disorder
- It affects 1 in 10,000 to 25,000 persons born
- There are approximately 400,000 people living with PWS worldwide
- PWS is a congenital chromosomal disorder affecting chromosome 15
- There is no cure
- Most common genetic cause of obesity

Physiology of PWS

- The hypothalamus, which controls feelings of satiety, does not function correctly in PWS patients
- Growth hormone releasing hormones (GHRH) are reduced by approximately 30% compared to the average person
- Levels of ghrelin, a potent appetite stimulating hormone, are raised and remain elevated even after meals

Characteristics of PWS

- Compulsive Hyperphagia - abnormally increased appetite
- Polyphagia - excessive eating
- Short stature with small hands and feet
- Mild retardation
- Hypogonadism - production of sex hormones and germ cells (sperm and eggs) is below normal which means they are sexually sterile
- Muscular Hypotonia - low muscle tone and decreased limb movement with poor reflexes

Exhibit non-ambulatory behaviour

- Exercise and movement are avoided
- Sedentary lifestyles are sought due to comfort

Reduced energy needs

- Metabolism is slower than average
- Dental caries
- Bizarre eating behaviours (gorging, hoarding, food stealing, eating inappropriate foods, etc.)
- Poor sucking ability and failure to thrive in infancy (may need PEG feeding during infancy)
- Varying degrees of obsessive compulsive behaviour
- Slow motor development
- Obesity related diabetes in later childhood
- Lack of emotional control
- Drowsiness

Three phases of PWS

Hypotonic Stage (prenatal-infancy)
Hyperphagic Stage (childhood)
Adolescence and Adulthood

Food security principles

No doubt
When meals will occur and what foods will be served

No hope
Of getting anything different from what is planned

No disappointment
Related to false expectations

Preventative strategies

- Food should be out-of-sight,
- Set meal times, equal portion sizes
- Clear communication & agreements
- Consistent routine
- Everything planned in advance
- Avoid change
- Set consistent staff responses (BIS plans)
- Regular counseling sessions
- Regular staff meetings / communication

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Clinical features

Stage 1 hypotonic, infant stage

- Low birth weight
- Severe muscle weakness (hypotonia)
- Suck/swallow problems, reflux, respiratory problems
- Subtle dysmorphic facial features such as triangle" shaped mouth" narrow forehead, almond-shaped eyes
- Underdeveloped sexual organs
- Delayed motor/physical milestones

Infancy

Hypotonia in infancy makes it difficult for babies to attain adequate calories, but by approximately 12 months to 36 months hyperphagia begins causing a massive influx in body weight. When hyperphagia becomes apparent the nutritional intervention methods need to be drastically changed.

Stage 2 – Hyperphagic (childhood)

- Between ages 2-6, hyperphagia symptoms begin with a preoccupation with food
- Voracious appetite begins due to lack of satiety feedback
- Slow metabolic rate
- Low energy level makes it difficult to exercise
- Emotional labiality and behavioural characteristicsWeight control and management
- Vitamin supplementation if needed
- Hypothalamic abnormalities reduce physiological sensations
- GHRH is reduced by 30% so children are shorter in stature
- Growth hormones are needed to avert early onset obesity which, in PWS patients on average occurs around age 2
- Educational and behavioural interventions are also needed for cognitive and social disabilities

Adolescence and adulthood

- The average individual with PWS has 40-60% body fat by later adolescence and general population
- Unless weight has been strictly managed throughout childhood weight-related disorders and diseases have manifested themselves
- Reduced or inhibited glucose tolerance due to diabetes
- 25% of adults with PWS have type II diabetes
- Nutritional intervention must be taken by a behavioural, medical, and an educational perspective
- Weight management plans are highly variable and depend up upon the individual and their support groups

Importance of dietitians

- Although PWS has not become more prevalent more diagnoses are seen because it is being identified earlier
- Dietitians need to be able to assist in early interventions starting at infancy and throughout adolescence and adulthood
- It is key to work with the family and or primary caregivers to educate them, implement dietary restrictions and guidelines, and to set nutritional goals and objectives
- Although dietitians are needed in the realm of nutritional intervention methods, the behavioural interventions are better suited for psychologists, psychiatrists or counselors

Exercise

- Critical component of management
- Improve muscle mass
- Weight control
- Improve strength
- Improve endurance
- Cardiac tone
- Improved lipid profile

Medical management

- Prone to more illnesses
- Regular check-ups
- PWS experienced doctor preferable
- Inform medical staff (on admission)
- Awareness of high-pain threshold
- Poor temperature regulation

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