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Perspective

Nutritional Treatment for Prader-Willi Syndrome and its Complications in Children

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DESCRIPTION

Prader-Willi Syndrome (PWS) is a rare genetic disorder that affects various aspects of physical, mental, and behavioral health. It is caused by the loss of function of genes on the paternal chromosome 15. One of the main features of PWS is hyperphagia, which is an excessive and uncontrollable appetite that leads to obesity and its complications. Therefore, nutritional treatment is a significant part of the management of PWS in adults, along with pharmacological and psychological interventions. The goals of nutritional treatment in PWS are to prevent or reduce obesity, maintain or improve muscle mass and function, prevent or treat metabolic and cardiovascular diseases, and improve quality of life.

However, achieving these goals is challenging due to the complex interplay of genetic, hormonal, neurological, and environmental factors that affect the energy balance and food intake in PWS. Adults with PWS have lower energy requirements than the general population due to their reduced basal metabolic rate, lean body mass and physical activity. Therefore, they need to consume small amounts of calories to maintain a healthy weight. The recommended daily energy intake for adults with PWS ranges from 10 kcal/kg to 25 kcal/kg depending on their age, gender, weight status, and activity leve. However, this may vary from person to person depending on their individual characteristics and needs. A nutritionist can help determine the optimal energy intake for each patient based on their clinical assessment and dietary history.

Adults with PWS have higher protein requirements than the general population due to their increased protein turnover and reduced muscle mass. Therefore, they need to consume more protein to preserve or increase their muscle mass and function. The recommended daily protein intake for adults with PWS ranges from 1.2 g/kg to 2 g/kg depending on their weight status and severity of disease. However, this may also vary from person to person depending on their individual characteristics and needs. A nutritionist can help determine the optimal protein intake for each patient based on their clinical assessment and dietary history.

Adults with PWS have altered lipid metabolism and increased risk of dyslipidemia and cardiovascular disease due to their obesity, hormonal imbalances, and genetic factors. Therefore, they need to moderate their fat intake to avoid hyperlipidemia and atherosclerosis. The recommended daily fat intake for adults with PWS ranges from 25% to 35% of total energy intake depending on their weight status and lipid profile. Therefore, they need to ensure adequate intake of vitamins and minerals to support their immune system, antioxidant defense, bone health, and neurological function.

The recommended daily micronutrient intake for adults with PWS follows the same guidelines as the general population unless there is a specific indication for supplementation. However, some micronutrients may have adverse effects on respiratory function if consumed in excess. Therefore, adults with PWS should avoid taking high doses of vitamin A (>1500 mcg/day), vitamin E (>400 IU/day), beta-carotene (>15 mg/day), iron (>18 mg/day), or zinc (>40 mg/day) without medical supervision. Therefore, they need to consume adequate amounts of fluids to prevent dehydration and edema. The recommended daily fluid intake for adults with PWS ranges from 1.5 liters to 2 liters depending on their age, gender, weight status, and fluid balance.

CONCLUSION

A nutritionist can help determine the optimal energy and nutrient intake for each patient based on their clinical assessment and dietary history. Moreover, some dietary patterns and habits that may benefit adults with PWS are the Mediterranean diet, the ketogenic diet, eating frequency and timing, and eating environment and behavior. Nutritional treatment in PWS may be more effective when combined with other interventions, such as exercise training, behavioral therapy, medication adherence, and growth hormone therapy.

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