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LEAST RESTRICTIVE ENVIRONMENT, NOT ONE SIZE FITS ALL FOR PRADER-WILLI SYNDROME

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ABSTRACT

Civil liberties and personal choice for individuals with intellectual disabilities have evolved over time with increasing emphasis and adoption of principles focused around "least restrictive environment" (LRE) in supportive care centers in order to maximize freedom of choice and individual autonomy for residents. However, rare disease pathology may encompass unique requirements for care inconsistent with LRE. Prader-Willi syndrome (PWS) is a rare genetic obesity -related syndrome, in which individuals are constitutionally unable, as a product of their genetic make-up, to manage feeding behaviors requiring strict oversight and limited food access to assure safety and avoid potentially life-threatening medical co-morbidities. The practical requirements for effective management of PWS has led to ethical conflicts and occasionally inappropriate prioritization of individual autonomy over personal safety. The responsible application of LRE principles to residential care settings in PWS requires adaptation to accommodate disease-specific requirements of care to optimally balance quality of life and longevity.

Recent trends that prioritize the preservation of civil liberties and personal choice for individuals with intellectual disabilities have increased emphasis and adoption of principles focused around "least restrictive environment" (LRE) in supportive care centers in order to maximize freedom of choice and individual autonomy for residents.[1] LRE is a component of federal laws that guide educational practices for students with disabilities requiring appropriate access to resources and mainstreaming which has been effective in reducing stigma, increasing access to learning and educational environments for many students. However, the nuances of rare genetic disease pathology often with unique requirements for care are frequently misunderstood and subject to misapplication of social trends including mental health care policies such as LRE.

Prader-Willi syndrome (PWS), a rare genetic obesity-related syndrome, in which individuals are constitutionally unable, as a product of their genetic make-up, to manage feeding behaviors and require intervention and strict oversight to assure their safety.[2,3] PWS is characterized by failure to thrive in infancy due to a poor suck and feeding difficulties, central hypotonia, hypogonadism and hypogenitalism in both males and females. Growth and other hormone deficiencies lead to growth failure, infertility and endocrine disturbances. A narrow upper airway, dry mouth with sticky saliva, enamel hypoplasia and central hypotonia create swallowing difficulties increasing the risk of aspiration, choking while eating and decreased gastrointestinal motility, perforation or necrosis which complicate medical care and response to treatment requiring close monitoring throughout life.[2] Perhaps most notable, PWS is characterized by hyperphagia, the unrelenting pathologic urge to consume food with unremitting hunger that typically develops in childhood leading to excessive food-seeking, tantrums when denied food, binge eating, consumption of non-food items, self-injury to obtain food and life-threatening obesity, if not controlled.[2] Hyperphagia and constant eating in the presence of food can lead to stomach necrosis or acute perforation. Morbid obesity is a significant contributor to mortality in PWS through cardiorespiratory failure, thrombotic events and associated diabetes with multi-organ failure.[4,5] Recent reports have identified a clear benefit and increase in longevity in PWS associated with modern era cohorts since the advent of better diagnostic technologies, earlier interventions to improve growth and increased awareness with vigilant monitoring to restrict access to food.[5]

These practical requirements for the effective management of PWS, unfortunately, run contrary to the

LRE principles leading to ethical conflicts and occasionally the inappropriate prioritization of individual autonomy over personal safety. While LRE principles may maximize opportunities and objectives in education, the theoretical framework is fundamentally inconsistent with the strict standards and food restrictions required for optimal health, quality of life and longevity in residential care settings for PWS as well as many other obesity-related mental disorders. Further, it is important to recognize that unrestricted access to food provides no relief from the insatiable food cravings experienced in PWS and only serves to exacerbate health risks and physical disability ultimately leading to the loss of the fundamental freedom afforded by good health, mobility and longevity that is associated with leanness. Thus, the broad application of LRE principles to living environments and residential care settings for PWS are more likely to precipitate severe physical disability, suffering and premature death. The responsible application of LRE principles to residential care settings requires significant adaptation and accommodation for disease-specific requirements of care and cannot merely open access to self-destructive behaviors in vulnerable populations such as those with Prader-Willi syndrome.

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