

Adults With Prader-Willi Syndrome and Decisions Regarding Least Restrictive Environment and the Right To Eat

Policy Statement:

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The quality of life for persons with mental retardation has improved significantly over the past two decades. This can be attributed, at least in part, to changes in societal attitudes and standards of care, often accompanied by legislative supports. Thus the concepts "inclusion," "least restrictive environment," and "the dignity of risk" in the context of community-based "supported living arrangements" have opened many new avenues of opportunity previously denied these adults. When applying the concept of "rights" to persons with developmental disabilities, however, it must be remembered that current disability rights laws were formulated primarily in consideration of those with physical disabilities. Uncritical application of these "rights" to persons with cognitive disabilities without regard to possible consequences may lead to tragic outcomes. While specific medical etiologies for any given disability may not always be of overriding importance in determining services and supports, there are, nonetheless, specific genetic syndromes and recognizable neurobehavioral patterns that present serious considerations that must be addressed in the development of a service plan. Thus, for instance, the type and level of support services for persons with Prader-Willi syndrome are relatively independent of their level of mental retardation and are much more related to the medical and neurobehavioral implications of the specific syndrome.

Developing an appropriate social milieu for individuals with Prader-Willi syndrome means creating an environment where the least restraints are present; however, environments of least restraint do not maximize freedom in an unbridled sense but are designed to help individuals achieve their fullest possible potential. Environments must become more restrictive when lesser restraints fail to protect the physical or emotional well-being of the person or to protect the person from doing avoidable harm to themselves or to others.

The issue of adults with Prader-Willi syndrome deciding whether they "want to diet or not" is just such an issue. The dialog that raises this issue is framed by the concept "least restrictive environment" or "client rights." The argument generally is that strict dietary management is "too restrictive" or that locking food abrogates "rights." A second issue is whether restricting spending money (to limit ability to buy food) violates the personal rights of adults with Prader-Willi syndrome. In many states, the agencies and group homes that specialize in Prader-Willi syndrome are increasingly criticized as being too restrictive, and as violating consumer rights. Many of these programs have been ordered to increase client access to food, to move clients into less restrictive settings, and to give clients control of their diets (Cormier, 1995; Goff, 1995; Greenswag et al., 1995). Although easier access to food may be a strong desire for individuals with Prader-Willi syndrome, it is a dangerous and medically neglectful practice. In too many cases, such practices have led to medical emergencies or to premature deaths related to complications of obesity. This growing trend is both alarming and tragic.

Failure to restrict access to food is tantamount to medical neglect. To illustrate, let us draw a parallel with diabetes. Diabetes results from a failure of the pancreas to produce adequate insulin. Thus, the person with diabetes must maintain a calorie- and carbohydrate-restricted diet while taking supplemental insulin. Failure to rigidly follow this regimen leads to elevated blood sugars and, ultimately, death. No caregiver home would think of telling diabetics that their diet was "too restrictive" or that restricting access was an abrogation of rights.

The management of the eating behaviors in persons with Prader-Willi syndrome is based on similar physiologic failures and is equally medically critical. In this instance, there is a genetically based inability to sense satiety, combined with a decreased utilization of calories, resulting in an elevated production of fat tissue. A failure to experience satiety leads the person to continue eating far beyond physiologic or nutritional needs. This overeating combined with elevated fat tissue production leads to rapid and morbid obesity. This rapid obesity overtaxes the heart and leads to complications that can include sleep apnea, diabetes, hypertension, and cardiopulmonary compromise. This physiologically driven eating behavior is no more under cognitive control, nor amenable to cognitive remediation, than is the failure of the pancreas to produce insulin in diabetes. Further, there are, to date, no medical, pharmacologic, or behavioral treatments that fix or cure this biological malfunction. Bioethicists dictate that informed consent requires the capacity to consider, and fully understand, the pros and cons of both sides of a decision issue prior to making a decision. Since by their own physiology, persons with Prader-Willi syndrome cannot decide "not to eat," therefore they cannot responsibly decide the converse: "to eat, or not to diet." Thus, to allow such decisions under the guise of "restriction of rights" is both medically and ethically unsound. Failure of the care-giving environment to maintain a rigidly managed diet or to supervise food access leads to the previously described rapid weight gain and can easily result in cardiopulmonary compromise and death. Such a failure in a medical setting would lead to charges of malpractice. Such a failure in a certified living environment may arguably lead to equally serious legal consequences.

Nonetheless, in the past three years, several persons with Prader-Willi syndrome have been placed in less restrictive environments under the argument of "rights." To date, several have died and the rest have been placed in more restrictive settings or rushed to critical care due to cardiopulmonary crises. Most have gained over 100 pounds in less than six months with the attendant acute medical complications. Clearly, restricting food is not an abrogation of rights; it is the standard of care for a person with Prader-Willi syndrome. Failure to restrict food and allowing a person to eat themselves to death is, in fact, a removal of "rights" to a protected environment.

Is it not a paradox that we would allow someone with Prader-Willi syndrome the "right" to eat themselves to death, but if someone without such cognitive limitations were to threaten suicide, the caregiver that failed to provide suicide restrictions would be found guilty of lack of protection? So we will protect those who are cognitively normal from their own self-destructive impulses, but argue that someone who has cognitive limitations and has physiologically driven eating behaviors has the right to eat themselves to death.

In planning the care-giving environment for persons with Prader-Willi syndrome, some contradictions are evident. While persons with Prader-Willi syndrome need extensive food support, they show fewer needs for support in other aspects of their lives. Indeed, many persons with Prader-Willi syndrome show competencies and decision-making abilities outside the food arena. Nonetheless, until there are medical or pharmacologic interventions for this physiologically driven eating behavior, structured environments with restricted access to, and intake of, food must be standard care for persons with Prader-Willi syndrome.

Some will argue that these recommendations conflict with concerns with choice, personal rights, and least restrictive environment. We do not take issue with these philosophical goals. Instead we assert that the appropriate frame of reference is the "least restrictive environment," given that the individual has Prader-Willi syndrome. Indeed the concept "least restrictive environment" is meant to imply "as normal a life as possible within the framework of a given disability." Too often it is translated as: "Despite your disability, you will live as though you are normal". Society's efforts to undo a previously created "social disability" may ultimately lead to a completely restricted environment when appropriate limit setting is insufficient. Persons with Prader-Willi syndrome must be uniquely considered as least restrictive goals are put into practice in order to prevent further deaths and to promote a fuller quality of life.

*Approved by the PWSA (USA) Board of Directors
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