

Prader- Willi Syndrome: When Client's Rights and the Medical Necessity for Food Security Conflict

Introduction: Among the achievements to be celebrated for those providing care to persons with cognitive challenges is the improved quality of life for those so challenged, coupled with the commitment to "client's rights" for living in the least restrictive environment and making their own choices when appropriate. The now common concepts of "community inclusion" and "supported living" have opened new avenues of opportunity and enjoyment previously denied affected adults: At the same time, research regarding genetic and behavior phenotypes has documented that for certain genetic disorders, one size does not fit all. Recognizing that specific medical etiologies for any given disability may not always be of overriding importance in determining services and supports, it is nonetheless clear that specific genetic syndromes and recognizable neurobehavioral patterns present serious considerations that must be addressed in the development of a service plan. For these individuals, the uncritical application of "rights" without regard to the consequences resulting from failure to adhere to medical needs may lead to tragic outcomes. *There is no more tragic example of this than the horribly painful death from rupture of the stomach of adults with Prader-Willi syndrome when a misunderstanding of the proper application of "client's rights" results in a complete disregard of their medical needs for food security.*

The Right to Decide Not to Diet: The issue of adults with Prader-Willi syndrome deciding whether they "want to diet, or not" is just such an issue. The dialog raising this issue is framed by the concept "least restrictive environment" and "client rights." The argument generally is that strict dietary management is "too restrictive" or that locking food abrogates "rights." 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 programs have been ordered to increase client access to food, to move clients into less restrictive settings, and to give clients decision making control of their access to food. 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, invoking such rights and the resulting overeating have led to medical emergencies and premature deaths. This growing trend is both alarming and tragic. For those with Prader-Willi syndrome, 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. 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.* Individuals with Prader-Willi syndrome have a genetically based inability to sense satiety (they cannot tell when they are full), combined with a genetically driven markedly decreased caloric need results in an elevated production of fat tissue. The brain based failure to experience satiety (know that they are full), combined with a decreased ability to feel pain means that affected individuals do not experience a volume induced discomfort. That is, their brain fails to

tell them that their stomachs are too full. This inability to know when they have eaten beyond the capacity of their stomach to physiologically handle that amount of food has resulted in medical emergencies and, for too many, death from stomach rupture. Other medical emergencies have arisen from choking while "quickly stuffing down" food that is not on their diets and the access to food was not appropriately restricted. 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. ***The client's rights issue for persons with Prader-Willi syndrome is a right to appropriate protection from the abnormal physiologic drive to eat that, when not appropriately provided for, will ultimately lead to death. This right is termed the right to "food security".***

A second issue is whether restricting spending money (to limit ability to buy food) violates the personal rights of adults with Prader-Willi syndrome. Because of the characteristics of the syndrome, the responsibility to appropriately limit food access and intake also requires restriction of available money and monitoring of how the money is spent. It is just as medically neglectful to allow free purchase of food outside the home as it is to allow unrestricted access in the living setting. When spending is not appropriately monitored, and the affected person has been "caught" with unauthorized food, serious medical emergencies have occurred from choking while "quickly stuffing down" their purchase.

In addition to the short-term consequences that can lead to death from unrestricted food access, there are long-term medical consequences that also lead to an early death. It is well established that individuals with Prader-Willi syndrome gain weight on 1/2 the calories allowed for an unaffected individual. It just takes a few short weeks of increased caloric intake to lead 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.

The Right to Decide Revisited: Bioethicists dictate that informed consent requires the capacity to consider, and fully understand, the pros and cons of both sides of an 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.

The Least Restrictive Environment for Persons with Prader-Willi Syndrome: Developing an appropriate social milieu for individuals with Prader-Willi syndrome means creating an environment where the least restraints are present, remembering that environments of least restraint do not maximize freedom in an unbridled sense, but are designed to help individuals achieve their fullest possible potential. 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 to achieve food security, they show fewer needs for support in other aspects of their lives. Indeed, many persons with Prader-Willi syndrome show numerous 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. 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***. For individuals with Prader-Willi syndrome, failure of the care-giving environment to maintain a rigidly managed diet or to supervise food access, thus providing food security, inevitably leads to the previously described consequences that can in the short term lead to stomach rupture and death, or in the somewhat longer term will lead to rapid weight gain, cardiopulmonary compromise and death. There is nothing "least restrictive" about a person who is so morbidly obese they are in a wheelchair and on oxygen.

In a medical setting, failure to provide the appropriate dietary and food access limitations would lead to charges of malpractice. Such a failure in a certified living environment can, and has, led to equally serious legal consequences based on medical neglect.

Adopted by the Prader-Willi Syndrome Association Board of Directors April 2016

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