EATING THEMSELVES TO DEATH:

Have "Personal Rights" Gone Too Far in Treating People With PWS?

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In contrast to even a decade ago, giant steps have been made toward increased community inclusion, choice, and self-determination for persons with mental retardation. As workers put these goals into practice, they are sometimes faced with uncomfortable exceptions to the rule: persons for whom increased decision-making leads to unhealthy and even life-threatening consequences. Prader-Willi syndrome is one such exception.

Prader-Willi syndrome is a genetic disorder resulting in mild to moderate levels of mental retardation and distinctive features (Cassidy, 1992; Dykens & Cassidy, 1996; Dykens, Hodapp, Walsh & Nash, 1992a, 1992b). One of the hallmark features of this syndrome is hyperphagia, overeating and marked interests in food (Holm et al., 1993). Contrary to popular belief, hyperphagia in individuals with Prader-Willi syndrome is not due to a weak character or to a lack of willpower around food. Rather, this hyperphagia in individuals with Prader-Willi syndrome stems from altered function of the hypothalamus (Swaab, Purba, & Hofman, 1995), the part of the brain that controls appetite and feelings of satiety. Though hyperphagia's exact cause is unknown, research shows that people with Prader-Willi syndrome do not have normal feelings of fullness and have reduced metabolic rates (Holland, Treasure, Coskeran & Dallow, 1995). These features lead to overeating and to high risks of obesity in affected individuals. Even today, Prader-Willi syndrome remains a life-threatening disorder, with most deaths related to complications of obesity, including cardiopulmonary compromise, hypertension, and Type II diabetes (Hanchett et al., in press).

Although not curable, hyperphagia can be successfully managed through a reduced calorie diet, a regular exercise program, and frequent weigh-ins. Appetite suppressants and behavior modification techniques aimed at reducing overeating have not proven particularly helpful, though pharmacotherapy and behavioral techniques are often successfully used to treat psychiatric and other problems in people with Prader-Willi syndrome (e.g., Dykens et al., 1992a; Dykens, Leckman, & Cassidy, 1996). Widely used approaches for managing hyperphagia in Prader-Willi syndrome rely on external controls, such as locking the refrigerator and other food sources; supervising clients in cafeterias, grocery stores; and community settings; and limiting client access to spending money because money is often used to buy food (e.g., Alexander & Greenswag, 1995).

Does it violate the rights of clients with Prader-Willi syndrome to lock up their food and restrict visits to friends, community travel, or spending money, especially given the relatively high mean IQ of 70 in this population? Many states have answered yes to this question. In these states, agencies and group homes that specialize in Prader-Willi syndrome are increasingly criticized as being too restrictive and have been cited for violating client rights. Further, these group home practices are viewed by many states as being overly protective and as denying people with Prader-Willi syndrome opportunities to learn from the "natural consequences" of their behavior (Goff, 1995). Subsequently, program personnel have been ordered to increase client access to food, move clients into less restrictive settings, and give clients control of their diets (Cormier, 1995: Goff, Greenswag et al., 1995).

Unfortunately, the natural consequences that stem from lesser restrictions include increased health risks and premature deaths related to complications of obesity. To date, we know of at least a dozen cases of deaths related to state policies and mandates to relax the food restrictions or living situations of clients with Prader-Willi syndrome. Other natural consequences include job losses due to food-seeking or poor work performance secondary to obesity, repeated arrests for stealing, physical harm while panhandling for food or money, and even trading sexual favors for food (Cormier, 1995; Goff, 1995). Indeed, an individual's drive for food can easily overcome their good judgments about the high risks of stealing, pan-handling, or becoming more obese.

Future tragedies such as premature death or exploitation can be avoided by appreciating certain unequivocal rules in treating hyperphagia in people with Prader-Willi syndrome. One unequivocal rule is that hyperphagia has a physiologic as opposed to a motivational basis. As such, external controls are needed to curb obesity, such as locked food sources and close supervision. Though people can learn specific food techniques that are helpful to them, interventions should not rely solely on this learning or on strategies such as increasing willpower or self-control around food. A second unequivocal rule is that food restrictions in Prader-Willi syndrome need to be life-long. Hyperphagia does not disappear if people achieve their ideal weight, though the drive for food may wax and wane in any particular individual over time. Long-term dietary restrictions in Prader-Willi syndrome are just as necessary as the life-saving diets used to treat other chronic medical disorders, such as phenylketonuria or diabetes.

Other treatment approaches to hyperphagia in Prader-Willi syndrome are less clear-cut. Specific food practices, for example, often differ between as well as within people over the course of time. One person may tolerate an unsupervised

coffee break at work, another may use this same opportunity to steal food. Some individuals do very well helping plan the group home menu; others become overly anxious doing so. One person may readily handle making his bag lunch with minimal supervision, another may not. The same person who successfully bags his lunch may forage through the garbage at work. Food supervision may need to increase in new environments and during transitions or stressful times and return to baseline when activities and places are more routine and familiar. Most professionals readily agree that specific food practices need to be tailored to the changing needs of each individual with Prader-Willi syndrome.

In spite of this agreement, however, the hyperphagia issue remains controversial among professionals, policy-makers, families, and clients. This is because many "best practices" in treating Prader-Willi syndrome conflict with mandates for increased choice and personal rights and the movement to place people in least restrictive environments. As workers strive to increase the independence of their clients with Prader-Willi syndrome on the job or in the community, they run into the immediate obstacle--food is everywhere. As people with Prader-Willi syndrome often exploit this fact with remarkable ingenuity, careful planning and coordination are needed across all settings where people live, work, and play.

To date, no single or simple solution exists to the autonomy versus food control controversy in Prader-Willi syndrome. Though we do not take issue with ideas of independence or personal rights, persons with Prader-Willi syndrome need to be uniquely considered as workers put these ideas into practice. In particular, we ask professionals and policymakers to keep in mind that hyperphagia in Prader-Willi syndrome has a physiological as opposed to a motivational basis that results in a life-long need for external controls. Appreciating these simple facts will go a long way toward meeting our most fundamental goal-the prevention of even more unnecessary deaths.

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