

# PRADER-WILLI SYNDROME ASSOCIATION

*Still hungry for a cure.*

## FOR THOSE WANTING TO START A SUPPORTIVE LIVING HOME FOR PRADER-WILLI SYNDROME

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While we appreciate your interest in starting a home for PWS, it will save both you and us some time by assuring that you first read the following before we talk:

- Become a members of PWSA (USA) in order to keep updated with the latest information through the newsletter
- Buy the Management book – 3rd edition (most important is Chpt 16, “Residential Care of Adults with PWS”)
- Buy the DiCosomo book & CD, “Supportive Living Care Plan for Adult with PWS in Placement”
- Buy the diet book for adolescents & adults.
- Read our advice printed below

If you get to the implementation stage, we can recommend consultants you can pay to come and meet with you to work out the details. Note: Much of the information below has been provided by Mary Kay Ziccardi, Regional Director of REM Consulting of Ohio. Mary Kay has over ten years experience in the development and implementation of PWS homes and is available for consultation and training.

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### OUTLINE OF RECOMMENDATIONS FOR BASIC RESIDENTIAL PROGRAM STANDARDS FOR PRADER-WILLI SYNDROME

Individuals with Prader-Willi Syndrome require a great many environmental controls, a structured and predictable schedule, and high levels of supervision.

<ul style="list-style-type: none"> <li>• <b>Environment</b> <ul style="list-style-type: none"> <li>Locked kitchen door/lowered rods</li> <li>Separate, locked from the outside bedrooms</li> <li>Ample space</li> <li>Alarm system</li> <li>Sturdy, heavy furniture</li> <li>Limited food access neighborhood</li> </ul> </li> <li>• <b>Food</b> <ul style="list-style-type: none"> <li>Pre-planned and posted menus</li> <li>Food notebooks/contacts with dietician/selection of three dislikes</li> <li>No participation in food shopping preparation/menu selection</li> <li>Goal weight privileges</li> </ul> </li> <li>• <b>Exercise</b> <ul style="list-style-type: none"> <li>Schedule for individualized needs</li> <li>One hour a day</li> <li>Utilize community sites</li> <li>Exercise physiologist for staff training and oversight</li> </ul> </li> <li>• <b>Medical</b> <ul style="list-style-type: none"> <li>Nursing oversight</li> <li>Community-based services</li> <li>Educating physicians</li> </ul> </li> <li>• <b>Behavior Management</b> <ul style="list-style-type: none"> <li>Individualized plans based on needs</li> <li>Token Economy- concrete expectations and consequences</li> <li>Team involvement and approval</li> </ul> </li> </ul>	<ul style="list-style-type: none"> <li>• <b>Staff Training</b> <ul style="list-style-type: none"> <li>Cornerstone of successful program</li> <li>Initial training (10 hours)</li> <li>PW training (4-5 additional hours)</li> <li>Continuous empowerment, support and problem-solving</li> <li>Adequate staff ratios based on needs</li> </ul> </li> <li>• <b>Vocational/Educational Contacts</b> <ul style="list-style-type: none"> <li>Training/Education</li> <li>Variety of settings as an option</li> </ul> </li> <li>• <b>Community Involvement/Activities</b> <ul style="list-style-type: none"> <li>Small groups or individualized</li> <li>Planned by individuals with PWS, coordinated by staff</li> <li>Natural opportunities, active lifestyle</li> </ul> </li> <li>• <b>Staff Ratio</b> <ul style="list-style-type: none"> <li>During busy time after work until bedtime – ratio of 3/5 or 2/4</li> <li>Overnight – ratio of 2/5 awake staff. In a home with 4 people with PWS, the ratio would vary between 1 and 2 depending on the situation.</li> <li>During the day 1 staff to drive to work and appointments, be home if sick, shop, cook, etc.</li> </ul> </li> </ul>
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# RECOMMENDATIONS FOR SUPPORTIVE LIVING FACILITIES FOR PRADER-WILLI SYNDROME

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In supportive living homes, many modifications need to be considered during the construction phase of the homes. For instance, the kitchen door needs to be secured, and closet rods should be lowered due to the individual's short statures – unless the person has been on growth hormone. Individuals who have Prader-Willi Syndrome function best when privacy and space is ample. To this end, all individuals should have their own bedrooms, and carry a key to lock their door. This simple environmental strategy can reduce their paranoia about others stealing their possessions, and greatly minimize the incidents of stealing and property destruction which often occurs among individuals with PWS. The front door and windows should have an alarm system to avoid a person with the syndrome sneaking out in the night to access food, and furniture should be very heavy and sturdy to avoid breakage during a tantrum. The home should not be near easy food access such as a restaurant or grocery store.

Program plans to meet the needs of individuals with PWS are equally extensive. Basic considerations of a carefully monitored program of dietary management and exercise are essential. Because people with PWS gain weight on very few calories, a registered dietician should carefully supervise the caloric intake. Menus should be planned in advance, and include three meals and two snacks. Each day's menu should be posted to assist in alleviating the intense anxiety and preservative behaviors that many people with PWS display in anticipation of the "unknown" regarding food. A multitude of other dietary strategies should be employed to ensure correct portion sizes and appetizing appearance and taste of the meals. Special considerations for added daily calories and occasional dining out can be added to an individual's plan once a goal weight is achieved and maintained.

Individual exercise plans should be created and overseen by a consulting exercise physiologist and implemented on a daily basis by direct care staff. Walking and swimming are the most popular and effective choices. Once a goal weight is obtained, strengthening and conditioning routines can be added.

Daily management of the challenges of PWS is also best monitored through the use of behavioral interventions. Well-defined expectations and rewards and consequences applied with consistency can positively impact an individual's opportunity for successful outcomes. Any formal behavioral strategies should be identified based on individual's needs and developed with interdisciplinary team and Human Rights Committee consensus. The expertise of a psychologist and staff trainer are invaluable in helping to meet these needs.

Another aspect in the daily management of PWS is medical oversight. Medical needs need to be monitored by a nurse, who will communicate with the individuals' physicians. There are major medical issues that need to be addressed such as skin-picking, medication use, temperature instability, high pain threshold, osteoporosis, and the life threatening complications of obesity. A thorough understanding of the complex medical issues of PWS is essential.

In addition to the above identified items, a consideration in a successful program needs to be highly-trained, supported, and empowered direct care staff. Staff needs to be trained extensively upon their hiring in regards to the basic areas of working with individuals with developmental disabilities. Further initial training should be provided specifically to address the unique needs of supporting individuals with PWS. Continuous training in the areas of behavioral interventions, assisting the individuals to establish a community presence and make choices within the scope of the issues faced, and value-based services should be provided.

Staff ratio should consist of the following:

- **After work** until bedtime 3/5 or 2/4
- **Overnight** -- ratio of 2/5 awake staff. In a home with 4 people, the ratio would vary between 1 and 2 depending on situation.
- **During the day** 1 staff to drive to work and appointments, be home if sick, cook, etc.

The use of materials from the nation office of the PWSA (USA), competency-based follow-up to behavioral plan training, and role-playing scenarios with staff have been found to be highly effective training methods. Successful staff must be able to provide consistent and caring responses to the many challenges presented on every shift.

Supports and structure need to be available in all of the individual's environments to assist with maximum opportunities for success and self-growth. To this end, extensive training and supports need to be provided to medical providers, community and sheltered work environments and school systems, as well as in other situations where an individual with PWS may have access. This has proven successful as it can reduce the amount of misunderstanding and miscommunications.

We realize that almost all individuals with PWS will require some level of structure and supervision throughout their lives. The challenges that they face can only be managed through successful program strategies and will not ever be entirely alleviated.