



# 6th International Prader-Willi Syndrome Caregivers' Conference

**21-23 May 2024, Berlin, Germany**

## Operationalized List of Infrastructure and Staff Skills Required for Specialized Care of PWS as Rated by Residential Care Providers in the USA

Janice Forster<sup>1</sup>, Lynn Garrick<sup>2</sup>, Kim Tula<sup>3</sup>, Linda Gourash<sup>4</sup>, Stacy Ward<sup>5</sup>, Ann Manzardo<sup>6</sup>

<sup>1</sup>Developmental Neuropsychiatrist, Pittsburgh Partnership, Pittsburgh, PA, USA

<sup>2</sup>Medical and Research Coordinator, PWSA I USA, Brandon, FL, USA

<sup>3</sup>Family Support Counselor, PWSA I USA, Brandon, FL, USA

<sup>4</sup>Developmental & Behavioral Pediatrician, Pittsburgh Partnership, Pittsburgh, PA, USA

<sup>5</sup> Interim CEO, PWSA I USA, Brandon, FL, USA

<sup>6</sup> Associate Professor of Pharmacology, Department of Psychiatry and Behavioral Sciences, University of Kansas Medical Center, Kansas City, KS, USA

# Background

- Residential placement crisis for persons with PWS in the USA
- Etiology is multiply determined
  - Not enough PWS placements available
    - People in placement are living longer because of improved health
  - Failure to meet eligibility criteria for funding
    - IQ < 70
    - IQ > 70 with deficits in adaptive function measured before age 18
  - Impact of growth hormone on the next generation of persons with PWS
    - Increased IQ scores
    - Not necessarily improved adaptive function or behavior

# Aims

- Identify better tools to assess maladaptive behaviors that determine eligibility for residential placement
- Define the characteristics that distinguish PWS residential care from other IDD residences
- Operationalize the essential infrastructure, personal care, and staff skills required for successful PWS-specific residential care
- Match the needs of the person with PWS to the environmental structure that provides the best “goodness of fit”

# Methods - 1

- Survey construction
  - 5 experts (the authors) with extensive experience
  - Informed by the Best Practice Guidelines for Standard of Care of Adults with PWS in Residential Living (2010)
  - Obtained consensus on items essential for the residential care of adults with PWS
    - infrastructure
    - personal care
    - staff skills/attributes
- Survey structure
  - 49 items
    - Respondents were asked to indicate “yes” or “no” if items were associated with the *specialized care and safety management of adults with PWS*

# Methods - 2

- Survey validation
  - 2 online focus groups
  - Email invitation with survey link
  - Respondents were asked to indicate how important the item was  
1(not important)-10 (very important)
    - 1) Experienced providers of residential care for PWS (n=7)  
85% Agreement
    - 2) Parents managing adult children with PWS at home (n=7)  
89% Agreement

# Methods - 3

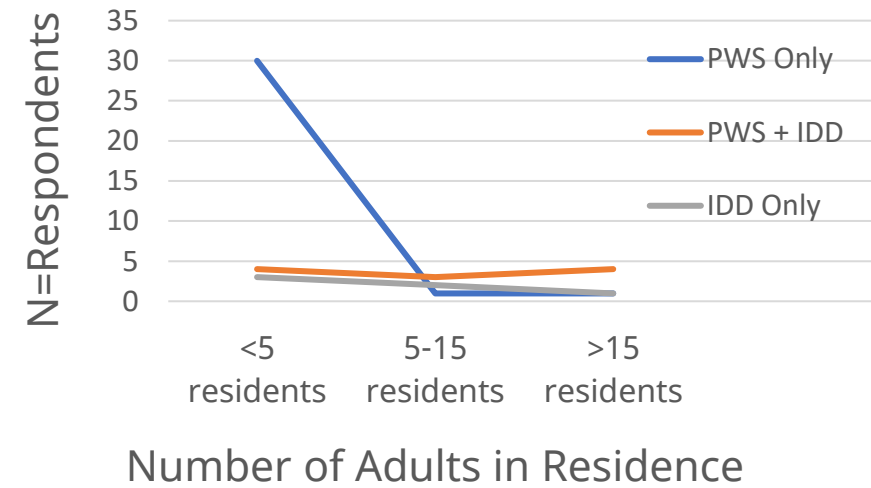
- “PWS Residential Care Survey” (RCS/PWS)
  - Electronic survey developed using the REDCAP platform
  - 49 items plus 5 demographic questions
    - Respondents were asked to indicate the number of years they worked with PWS, their level of employment experience, and characteristics of the residential setting in which they work
- Survey solicitation
  - Systems of care for PWS in USA
  - Provider associations in USA
  - Survey link was open between June 22-November 15, 2023
  - Surveys were submitted electronically and anonymously
- Survey analysis
  - Data was analyzed using SAS version 9.4 by Ann Manzardo, PhD at KUMC

# Results

## Survey completion and demographics

Group Home Type	Respondents Total N=50	Years of Experience
PWS Only	N=38, 76%	7.4±6 (1-22)
PWS + IDD	N=6, 12%	9.0±7 (3-23)
IDD Only	N=6, 12%	11.3±7.8 (3-20)

- Staffing: 24 hours per day (>82%) in all groups
- 78% Agreement among respondents



# Results

Food-Related Infrastructure Items	Total, N=50		PWS-Only	Not PWS-Only	p-value
	Yes	No	N, % Yes	N, % Yes	
1. Daily calorie count or ideal body weight determined by PCP	100%				NA
2. Dietary plan determined by dietician or nutritionist	96%	4%	N=38, 100%	N=10, 83%	0.01
3. Weight measured at least weekly	98%	2%			NA
4. Predetermined plan for managing weight fluctuations	96%	4%			NA
5. Psychological Food Security "I know, I accept, I am content"	96%	4%			NA
6. Direct supervision of meals and snacks	98%	2%			NA
7. Use of Pace and Chase	88%	12%	N=36, 95%	N=8, 67%	<0.01
8. Predetermined plan to address unauthorized food access	94%	6%	N=37, 97%	N=10, 83%	0.07
9. Food access managed by locks	98%	2%			NS
10. Food access managed by door alarms	74%	26%	N=31, 82%	N=6, 50%	0.03
11. Food access managed by motion detectors	42%	58%	N=21, 55%	N=0	<0.001
12. Food access managed by cameras	16%	84%	N=8, 21%	N=0	NS
13. Eyesight supervision in the community	96%	4%			NA



# Results

Non-food Related Infrastructure Items	Total, N=50		PWS Only	Not PWS Only	p-value
	Yes	No	N, % Yes	N, % Yes	
14. Carer available at night	100%				NA
15. Physical safety managed by locks on doors and windows	88%	12%			NS
16. Physical safety managed by door/window alarms	92%	8%			NS
17. Physical safety managed by motion detectors	48%	52%	N=22, 58%	N=2, 17%	0.01
18. Physical safety managed by cameras	26%	74%			NS
19. Monitor/manage personal safety on internet	84%	16%	N=35, 92%	N=7, 58%	<0.01
20. Monitor/manage modesty	88%	12%	N=36, 95%	N=8, 67%	<0.01
21. Monitor/manage romantic relationships	92%	8%	N=35, 92%	N=8, 67%	0.02
22. Monitor/manage media/entertainment for sex or violence	76%	24%	N=33, 87%	N=5, 42%	<0.001
23. Monitor/manage use of money or equivalent	94%	6%			NS
24. Monitor/manage time on phone, screens	66%	34%	N=28, 74%	N=5, 42%	0.04
25. Monitor/manage organization of belongings	92%	8%	N=38, 100%	N=8, 67%	<0.01

# Results

Personal Care Items	Total, N=50		PWS Only	Not PWS Only	p-value
	Yes	No	N, % Yes	N, % Yes	
26. Collect data on refusals or disruptive behavior	100%				NA
27. Predetermined daily plan for hygiene and grooming	94%	6%			NS
28. Predetermined daily plan for work/school/leisure	94%	6%			NS
29. Predetermined daily plan for social interaction with peers	86%	14%	N=36, 95%	N=7, 58%	<0.01
30. Predetermined weekly plan for contact with family	72%	28%			NS
31. Predetermined daily plan for community activities	94%	6%	N=38, 100%	N=9, 75%	<0.01
32. Predetermined plan for religious practice	72%	28%	N=31, 82%	N=5, 42%	<0.01
33. Supervised exercise at least 1 hour/day	86%	14%			NS
34. Supervised/assisted oral care	90%	10%			NS
35. Supervised/assisted hygiene and grooming	92%	8%			NS
36. Body checks head to toe for skin picks or injury	86%	14%	N=36, 95%	N=7, 58%	<0.01
37. Monitor frequency of BMs	82%	18%			NS

# Results

Personal care; Staff attributes/skills Items	Total, N=50		PWS Only	Not PWS Only	p-value
	Yes	No	N, % Yes	N, % Yes	
38. Monitor amount of sleep day and night	70%	30%			NS
39. Monitor daily fluid intake	84%	16%	N=36, 95%	N=6, 50%	<0.001
40. Monitor budget/use of money	94%	6%			NS
41. Carer knows/uses low expressed emotion	80%	20%			NS
42. Carer knows/uses language adapted for IQ and PWS	82%	18%			NS
43. Carer knows/uses cues for task shifting	92%	8%	N=37, 97%	N=9, 75%	0.01
44. Carer knows/uses verbal redirection	100%				NA
45. Carer knows how to say "No"	98%	2%			NA
46. Carer knows how to provide choices	98%	2%			NA
47. Carer knows how to identify/manage poor judgment	92%	8%			NS
48. Carer knows/uses de-escalation techniques	98%	2%			NA
49. Carer knows/models appropriate social skills, boundaries, conflict resolution	98%	2%			NA

# Conclusions

- The Residential Care Survey for PWS (RCS/PWS) is a consensus document that defines the infrastructure, personal care, and skills and attributes that carers need to successfully manage adults with PWS in the residential care setting.
- Environmental control over food access is an essential management tool, but there are many other aspects of infrastructure, personal care, and skills and attributes of the carers that define and operationalize specialized care for PWS in the residential setting.
- A second survey using the (RCS/IDD) is planned to allow a comparison with the current data to better define a continuum of residential environmental supports for adults with PWS from least to most restrictive.
- Moving forward, we will use an assessment tool (PWS Acuity Scale) to define the phenotypic and behavioral characteristics of each adult with PWS on a waiting list prior to placement and then re-evaluate 6 months after admission to assess status.



# Thank you

janiceforstermd@aol.com

## 6th International Prader-Willi Syndrome Caregivers' Conference

21-23 May 2024, Berlin, Germany

