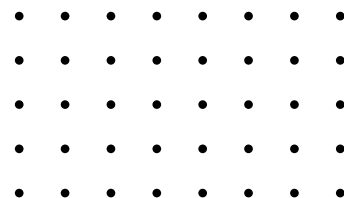




Outcomes for persons with Prader-Willi syndrome in full-time services

*Findings from international archival data
and future research directions*

Professor Brian Hughes, PhD, University of Galway, Ireland
& the IPWSO Research on Specialist Services Steering Group



RESEARCH

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Body weight, behaviours of concern, and social contact in adults and adolescents with Prader-Willi syndrome in full-time care services: Findings from pooled international archival data

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Abstract

Background Prader-Willi syndrome (PWS) is a complex genetic neurodevelopmental condition characterised by a range of debilitating and lifelong symptoms. The many physical and behavioural challenges that arise with adults with PWS often necessitate full-time (i.e., 24-hour) professional care support. However, despite the fact that many clinicians regard full-time PWS-specific care to represent best practice, relatively few studies have directly examined the benefits of such services. The purpose of this paper is to use archival data to investigate the impact of full-time care services on people with PWS, and to assemble a large statistical dataset on which robust analyses of improvements in weight, BMI, and behavioural outcomes can be based.

Methods Information collated by the International PWS Organisation (IPWSO), an international non-profit membership organisation supporting national PWS associations around the world, was combined into a single anonymised dataset for statistical analysis. Data were supplied by service-providers from several countries who provide full-time support to people with PWS. The dataset included details on the specific services provided, basic demographic information on service recipients, including weight, body mass index (BMI), and observational records relating to behaviours of concern (BOC; consisting of temper outbursts, skin-picking, egocentrism, inflexibility, and striving for dominance).

Results A total of 193 people with PWS (ranging in age from < 10 yrs to > 50 yrs; 93% of whom were > 18 yrs), residing in 11 services across 6 countries, were represented in the dataset. On average, people with PWS showed significant reductions in weight and BMI after joining a full-time care service, with improvements within one year of entering, which were cumulative over time and independent of age or initial weight at entry. Similar cumulative improvements over time were seen for BOC within one year and were unrelated to age or severity of BOC at entry.

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Hughes BM, Holland A, Hödebeck-Stuntebeck N, Garrick L, Goldstone AP, Lister M, Moore C, Hughes M. Body weight, behaviours of concern, and social contact in adults and adolescents with Prader-Willi syndrome in full-time care services: Findings from pooled international archival data. *Orphanet J Rare Dis.* 2024 Feb 7;19(1):48. doi: 10.1186/s13023-024-03035-x

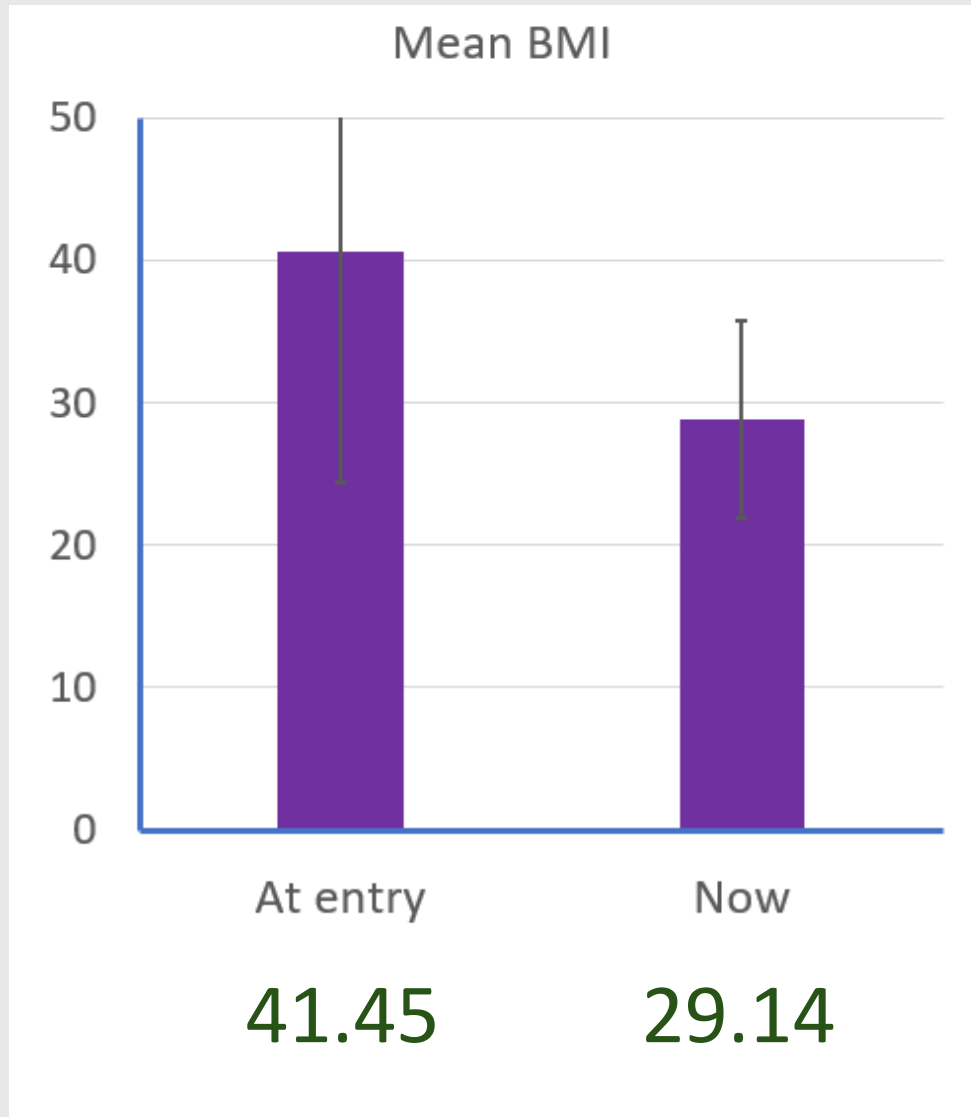


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The study

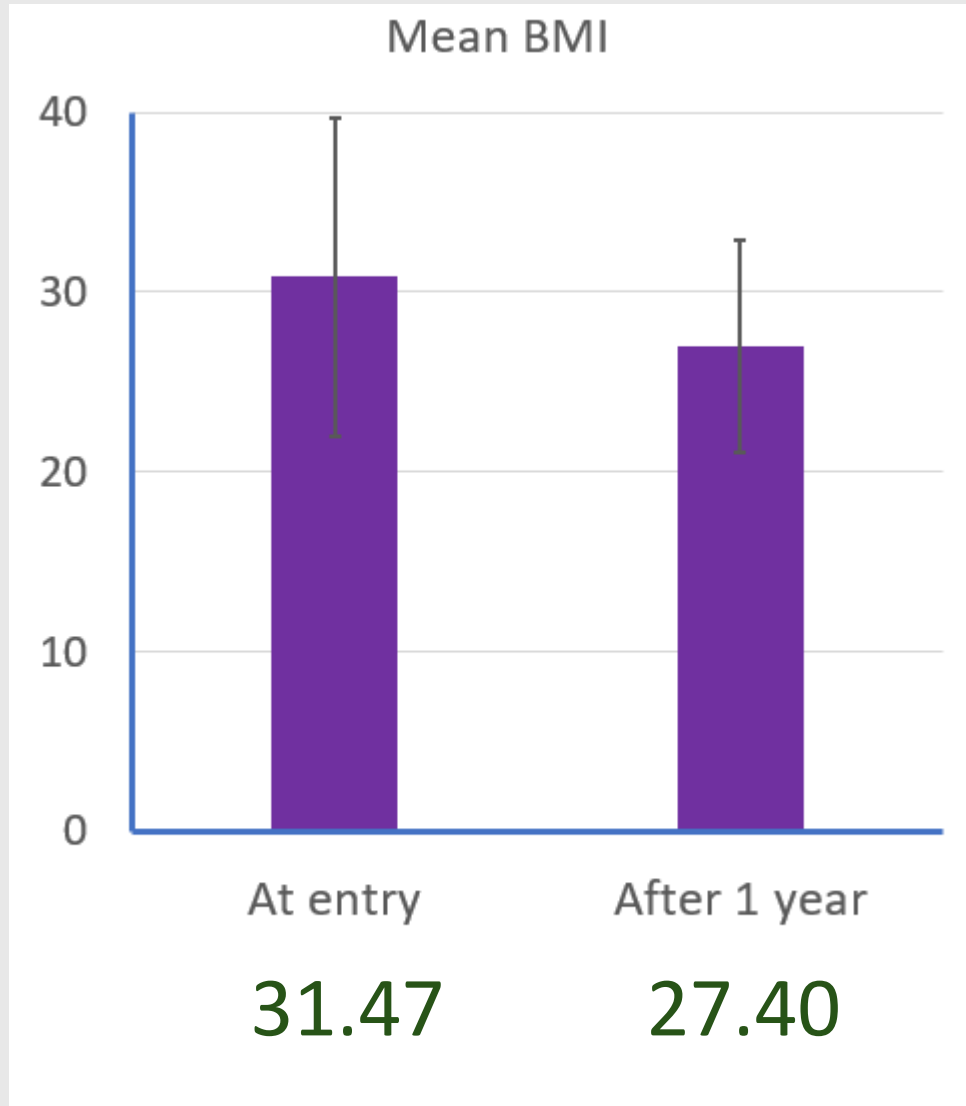
- Data supplied by 11 full-time care services from across six countries
 - Denmark, Germany, Ireland, Switzerland, United Kingdom, United States
- At least partial data: n = 193 individuals with PWS
 - In service for 10.26 yrs (range <5 to >40)
 - Average age of entry = 26.67 yrs (range <10 to >50)

Overall BMI reduction



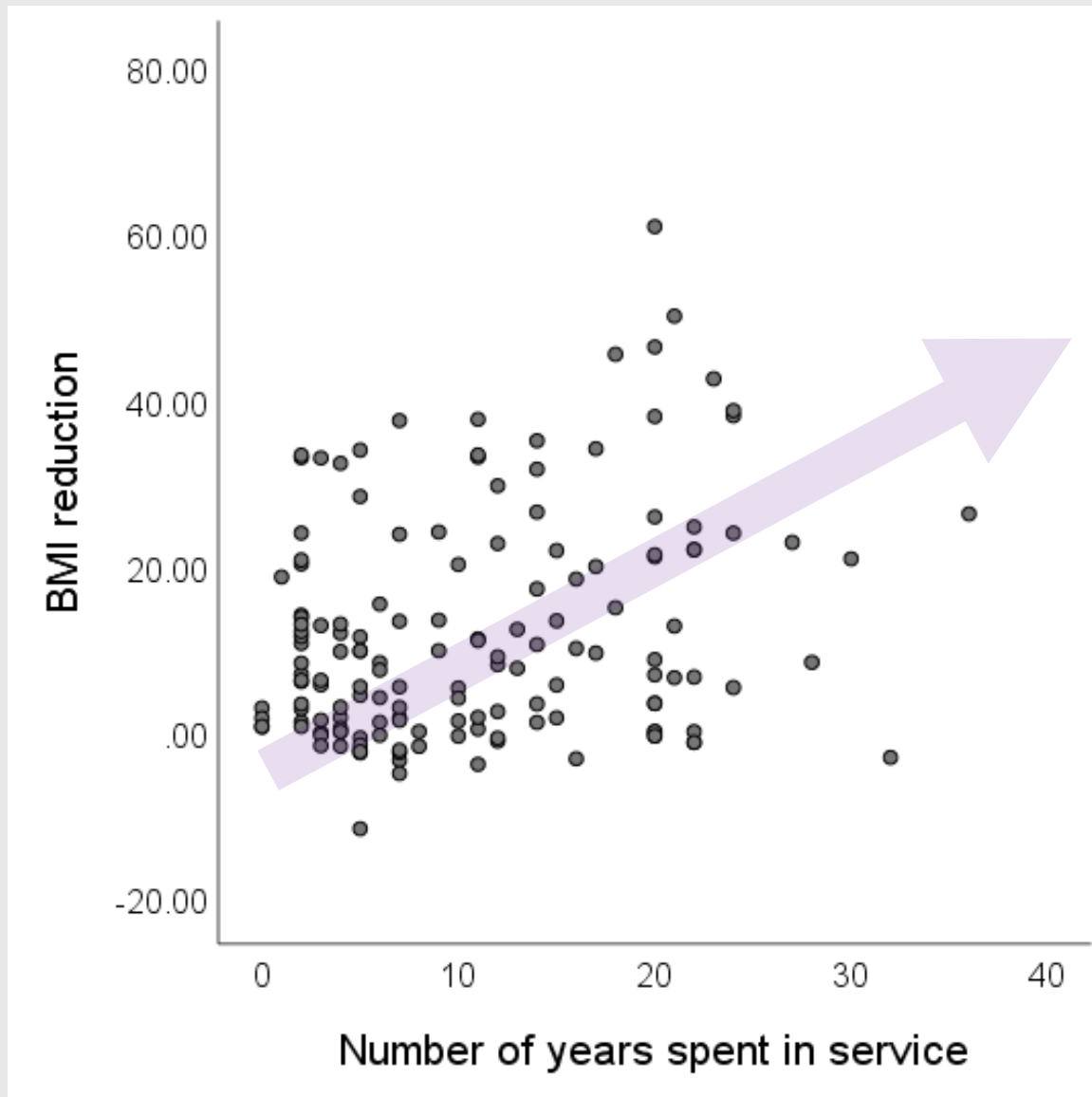
- Data sample: $n = 141$
- Mean BMI reduction
 - 12.31 kg/m^2
- Statistically significant change
 - $t(140) = 19.65$,
 $p < .001$

BMI reduction after one year



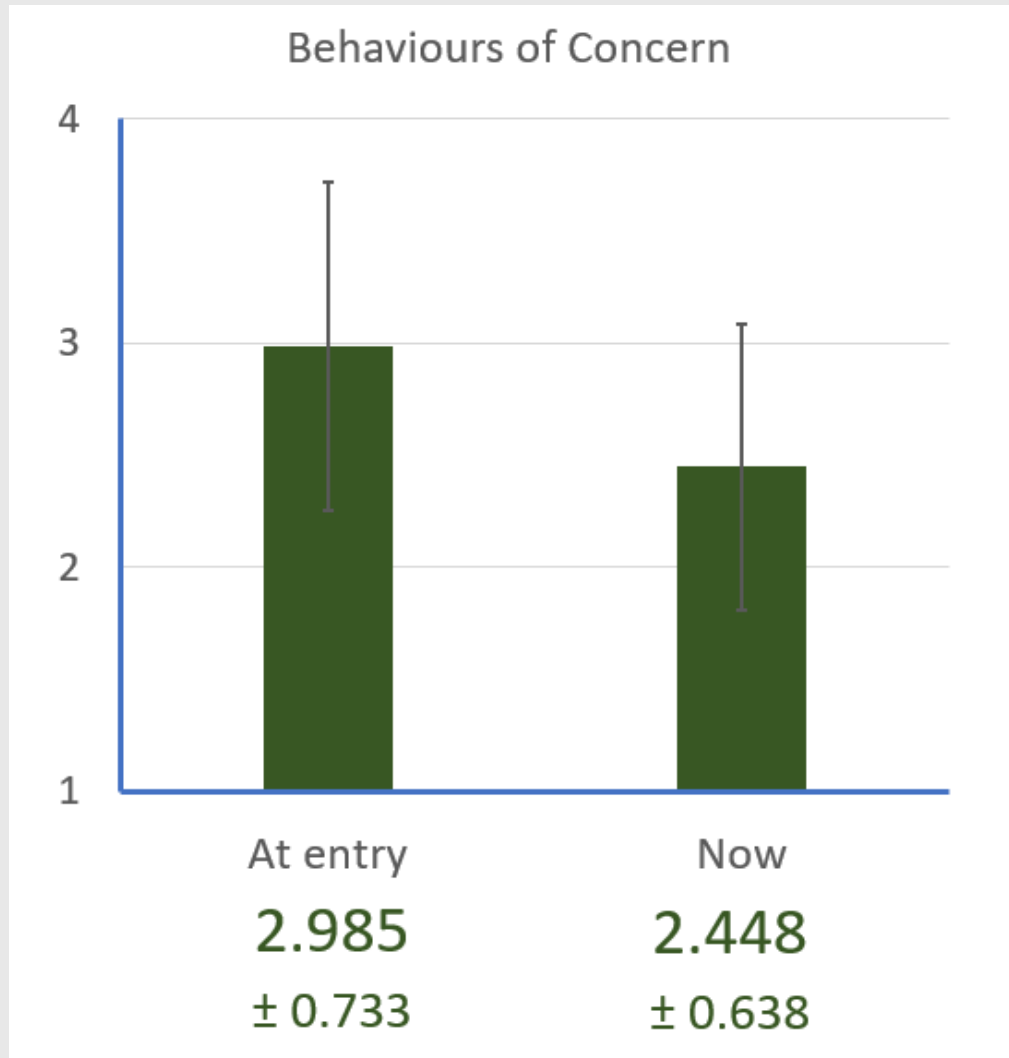
- Data sample: $n = 34$
- Mean BMI reduction
 - 4.07 kg/m^2
- Statistically significant change
 - $t(33) = 5.32,$
 $p < .001$

More time, more BMI reduction



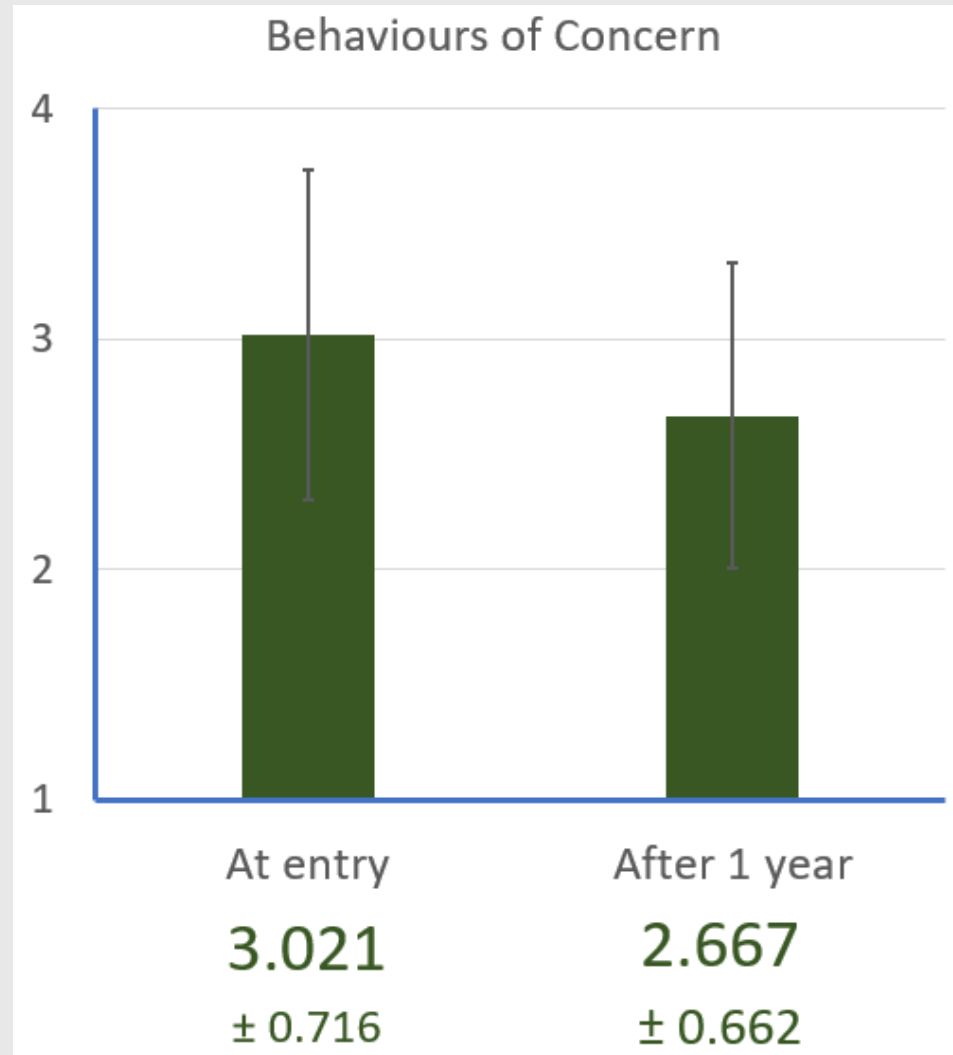
- Data sample: $n = 141$
- Bivariate correlation
 - $r = +.272, p = .001$
- Controlling for age
 - $r_p = +.284, p = .001$
- Excluding outlier
 - $r_p = +.267, p = .002$

Overall change in Behaviours of Concern



- Data sample: $n = 41$
- Mean reduction
 - 0.538 ± 0.576
- Statistically significant change
 - $t(40) = 5.98,$
 $p < .001$

Change in Behaviours of Concern after one year



- Data sample: $n = 39$
- Mean reduction
 - 0.354 ± 0.446
- Statistically significant change
 - $t(38) = 4.96,$
 $p < .001$

Predictors of Current BMI

Data sample: $n = 91$

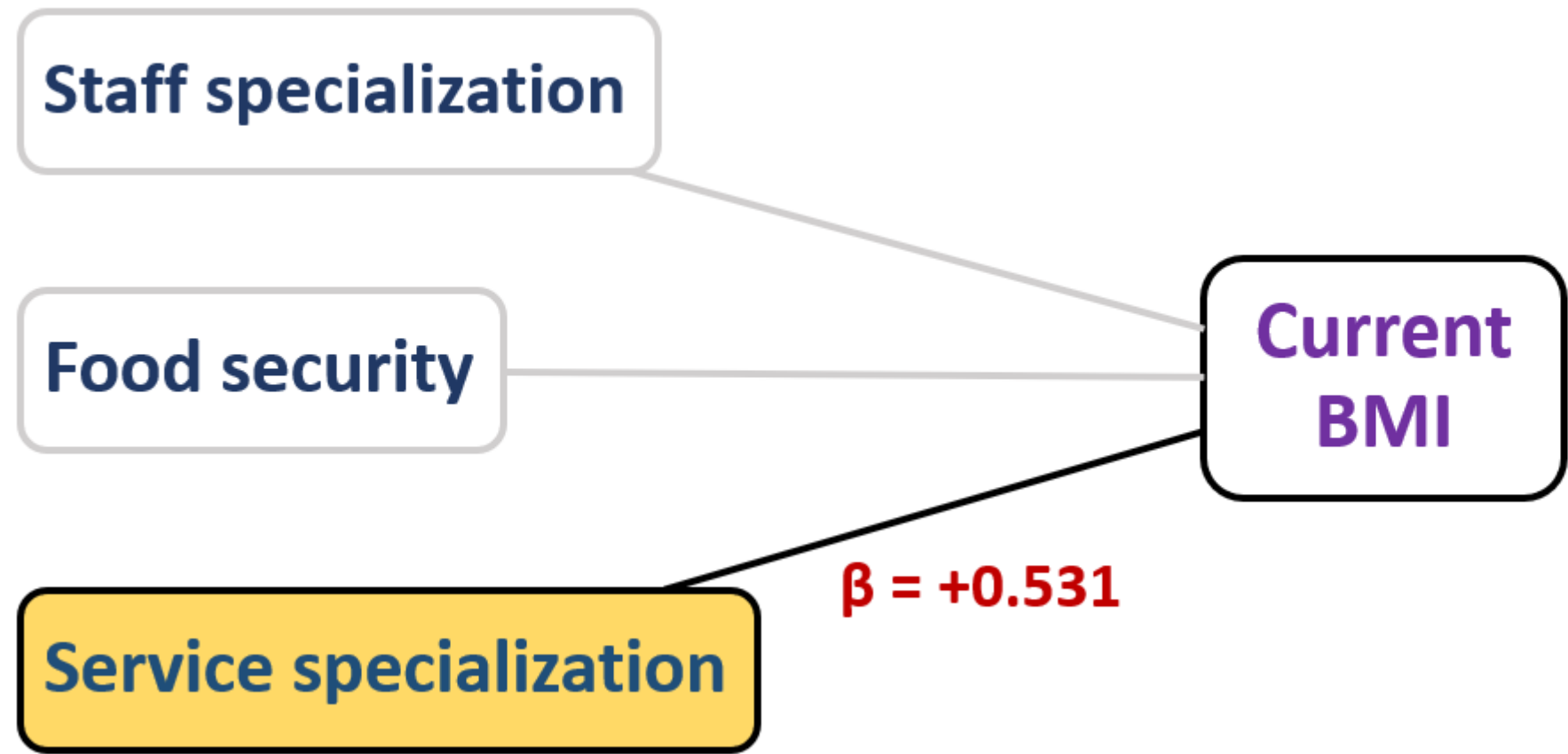
Staff specialization

Food security

Service specialization

Current
BMI

$\beta = +0.531$



Predictors of Behaviours of Concern

Data sample: $n = 91$

Staff specialization

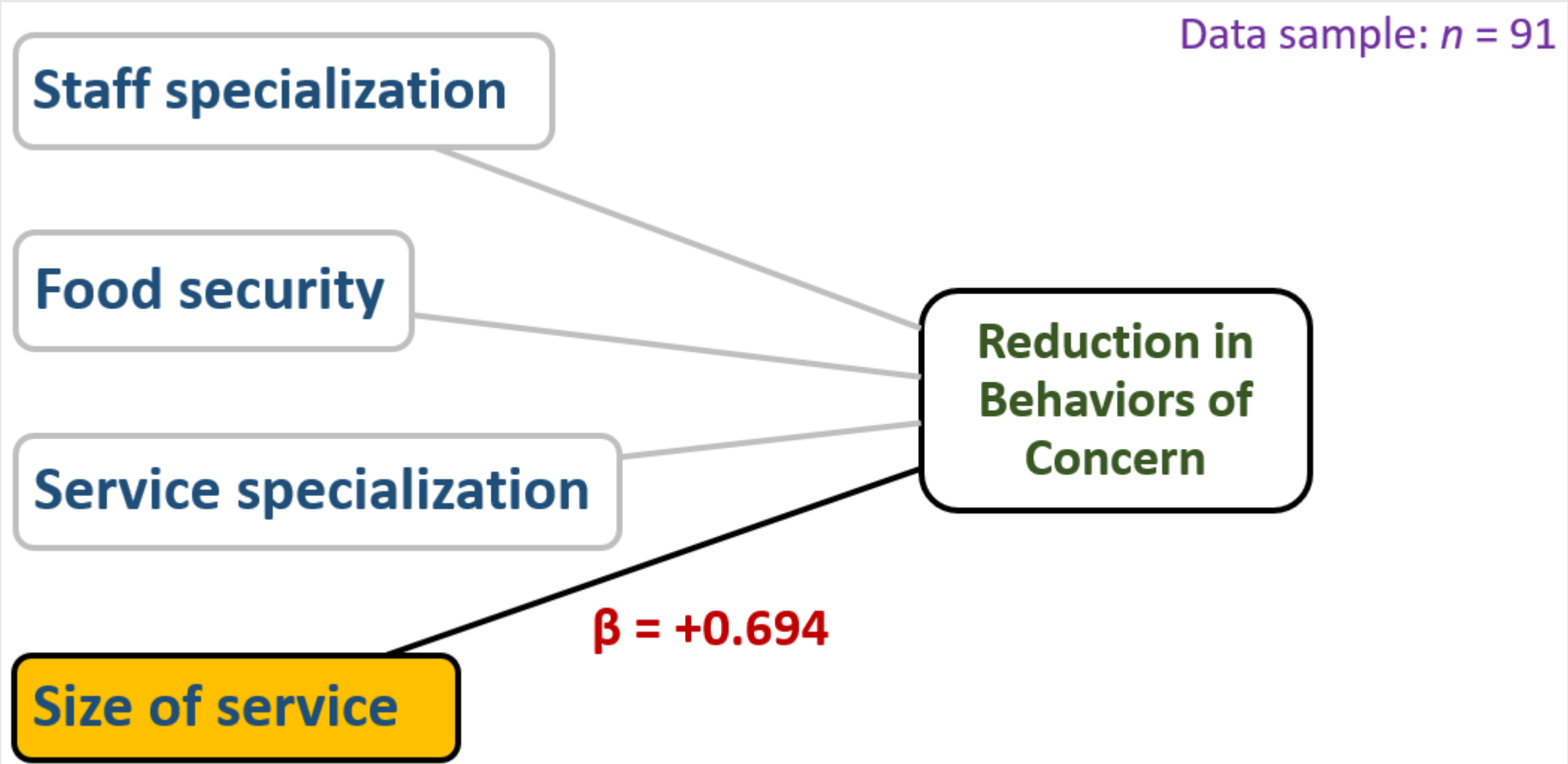
Food security

Service specialization

Size of service

Reduction in
Behaviors of
Concern

$\beta = +0.694$



Summary of Findings

- When in residential services, people with PWS
 - (a) lose significant weight, and
 - (b) exhibit significant improvements in behaviours of concern
- These improvements
 - (a) become apparent within one year, and
 - (b) increase over time
- Various service features are associated with improvements:
 - (a) Service specialization, staff specialization, food security & size of service all important
 - (b) Service specialization is especially important for BMI
 - (c) Size of service is especially associated with improvements in behaviours of concern

Future Research Directions?

- Archival data
 - (a) Broaden / deepen / greater cultural representation
- Primary data
 - (a) Standardised outcome measures
 - (b) Voices of persons with PWS
 - (c) Voices of families
 - (d) Voices of caregiving staff
- Processes vs. Structures
 - (a) Why are certain service features important?
 - (b) Why do outcomes improve over time?

FAM-PWS 2024

A Study of Families Living with PWS



Welcome to our study!

We are [psychology researchers based in the University of Galway in Ireland](#). One of us is a parent of a son who has PWS.

Our study will examine the experiences and well-being of families living with [Prader-Willi Syndrome](#).

If you are **a parent/guardian of a person who has PWS**, then please consider taking part in our study. The study involves a survey that you can complete online in around 20-25 minutes.

Click [here](#) to take part.

pws-research.com



Thank you!
Danke schön!

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

21-23 May 2024, Berlin, Germany

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