A New Resource: 'Practical Portions for Prader-Willi Syndrome' Evelien Gevers

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Barts Health NHS Trust

William Harvey Research Institute



Background

- 1 out of every 15,000 births
- Hypotonia
- Low muscle mass
- Feeding difficulties
- Faltering growth
- Developmental and cognitive delay
- Behavioural problems
- Sleep disorders
- Neuroendocrine abnormalities
- Hyperphagia



Genotype – Phenotype relation

• mUPD:

Higher maternal age (31.4 ± 3.4 vs 27.8 ± 3.8 years), more anxiety (64.29% vs 26.09%) and autistic traits (57.14% vs 26.09%), and less hypopigmentation (42.11% vs 68.24%) and skin picking (42.86% vs 71.01%) than the deletion group. More change of psychosis.

• Type I deletion:

Diagnosed at earlier age (3.7 ± 3.3 vs 6.2 ± 3.2 years) and more common speech delay (95.45% vs 63.83%) than type II. Slightly higher BMI than mUPD.



Natural progression



Tauber M, Lancet Diab and Endocrinol 2021

Nutritional phases

- **Phase 0**: intrauterine growth is restricted.
- **Phase 1** (at birth): the infant is hypotonic
 - subphase 1a: difficulty feeding with failure to thrive (aged 0–9 months)
 - subphase 1b: grows steadily along a growth curve and weight increases at a normal rate (9 mo-2 years).
- Phase 2: excessive weight gain
 - subphase **2a** (aged 2–4 years): without a substantial change in appetite or caloric intake
 - Subphase **2b** (aged 4-6 years): with a concomitant increased interest in food.
- Phase 3: marked hyperphagia, typically accompanied by food seeking and insatiable appetite
- [Phase 4 (in some adults): no longer insatiable appetite and ability to feel full]





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Prader-Willi syndrome: guidance for children and transition into adulthood. Shaikh MG, Barrett TG, Bridges N, Chung R, Gevers EF, Goldstone AP, Holland A, Kanumakala S, Krone R, Kyriakou A, Livesey EA, Lucas-Herald AK, Meade C, Passmore S, Roche E, Smith C, Soni S. Endocr Connect. 2024 Jul 10;13

Diet and weight management

- Aim to prevent obesity rather than reduce obesity
- Provide safe food environment
- Three pillars: Growth hormone, healthy diet, exercise.
- Recognise the reduced energy requirement from approx. age 2









1) To develop a national pictorial resource defining and illustrating correct portion sizes for children with PWS.

2) To assess the nutritional adequacy of these proposed intakes.

Methods

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Used previously defined national portion sizes for children (Public Health England, 2016)

Proportionally reduced by 30%

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Resource used to produce a sample 24hour meal plan

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Applied the defined portion sizes for age

2-3years	11-13years
4-6years	14-18years
7-10years	19-64years

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2-3years	11-13 years	
4-6years	14-18 years	
7-10years	19-64 years	
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Analyse	d the diet	

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'Practical Portions for Prader Willi Syndrome'

Practical Portions for Prader-Willi Syndrome

- Aimed for use by carers and adult patients with supervision
- Photographs of 60 foods for 6 age categories
- Includes undesired foods, but labelled with a red dot
- Freely available on line and printed from PWSA-UK

2) To assess the nutritional adequacy of these proposed intakes.

'Practical Portions for Prader Willi Syndrome'

Cutlery and crockery

The cutlery and crockery shown here are the size of those used in the photographs. You can use similar household utensils to help measure your own food portions. This circle is the same size as the plate

The bowl used in the photos is 17cm (61/2 inches) diameter.

Teaspoon (tsp.) Dessert spoon (dsp.)

All tablespoons are heaped unless otherwise stated.

CLO OF SIGES ~ 200ml used in the photos (22cm/81/2 inches diameter). Serving food on a small plate like this, instead of a large plate, can make portions look bigger. Plate - 22cm/81/2 inches

Tablespoon (tbsp.)

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Potatoes, bread, rice, pasta and other starchy foods (6-8 portions per day)

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How much food should a person with PWS eat every day?

The table below shows the recommended number of portions from each food group a person with PWS needs every day. Depending on individual needs, your healthcare professional may suggest something different.

Food Group	Number of portions each day
Vegetables	At least 3
Fruit	2
Potatoes, bread, rice, pasta and other starchy carbohydrates	6-8
Beans, pulses, fish, eggs, meat and other proteins. Include at least 1 portion of oily fish such as salmon, herring or mackerel every week.	2-3
Dairy and alternatives	3
Oils and spreads	3
Foods and drinks high in fat, salt or sugar	Not recommended

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Design of 24 hr meal plan

		Monday	
Meal/ snack	Food	Food group	Number of portions
	1½ wheat biscuits	Potatoes, bread, rice, pasta and other starchy carbohydrates	1
Breakfast	210ml skimmed milk	Dairy and alternatives	1
	1½ plums	Fruit	1
Spack	3 breadsticks	Potatoes, bread, rice, pasta and other starchy carbohydrates	1
SHACK	2 tbsp. hummus	Beans, pulses, fish, eggs, meat and other proteins	1
	2 slices wholegrain bread	Potatoes, bread, rice, pasta and other starchy carbohydrates	2
	1½ tsp. reduced-fat spread	Oils and spreads	1
Lunch	3 slices roast beef	Beans, pulses, fish, eggs, meat and other proteins	1
	5cm cucumber and 6 cherry tomatoes	Vegetables	1
	²/ ₃ banana	Fruit	1
Snack	² / ₃ small matchbox sized piece of cheese	Dairy and alternatives	1
	³ / ₄ wrap	Potatoes, bread, rice, pasta and other starchy carbohydrates	1
	½ salmon fillet, oven cooked	Beans, pulses, fish, eggs, meat and other proteins	1
	2 egg-sized potatoes	Potatoes, bread, rice, pasta and other starchy carbohydrates	1
Dinner	3 heaped tbsp. carrots	Vegetables	1
	3 heaped tbsp. broccoli	Vegetables	1
	1 tsp. olive oil	Oils and spreads	1
	1 fromage frais (85g)	Dairy and alternatives	1

12 yr old girl

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Protein %	288	288	354	354	302	302	231	236	208	258	192	236
Fat %	61	66	62	67	60	64	57	61	51	62	51	64
Carbohydrate (mse) %	40	44	44	49	41	44	41	44	36	44	31	40
Energy (kcal) %	59	64	63	69	61	66	58	62	52	64	49	62
Energy (kJ) %	60	64	63	69	62	66	58	62	52	64	49	62
NSP (Englyst) %											61	61
Saturated fats %	62	67	49	54	49	53	48	51	41	50	40	51
Mono-unsat.fats %	47	50	37	40	39	42	34	36	29	36	28	36
Poly-unsat.fats %	41	45	34	37	36	39	33	35	28	35	25	32
Trans.fats %	13	14	10	11	10	11	9	10	8	10	8	10
Sodium (Na)	121	121	130	130	84	84	81	81	97	97	78	78
Potassium (K)	157	157	196	196	122	122	90	90	91	91	84	84
Calcium (Ca)	120	120	121	121	108	108	77	96	86	108	109	109
Magnesium (Mg)	125	125	157	157	104	104	89	89	97	97	83	92
Phosphorus (P)	213	213	247	247	231	231	161	200	191	237	234	234
Iron (Fe)	64	64	134	134	105	105	99	76	115	88	131	77
Copper (Cu)	94	94	119	119	109	109	115	115	106	106	77	77
Zinc (Zn)	85	85	110	110	127	127	112	112	130	176	119	161
Chloride (Cl)	114	114	118	118	77	77	71	71	81	81	64	64
Selenium (Se)	100	100	113	113	112	112	81	81	66	77	50	62
lodine (I)	76	76	69	69	76	76	80	80	86	86	80	80
Vitamin D	36	36										
Thiamin	141	153	166	182	152	163	147	158	126	154	118	151
Riboflavin	124	124	139	139	128	128	137	149	142	167	135	160
Niacin	112	120	130	142	144	155	128	137	116	143	105	134
Vitamin B6	120	120	141	141	128	128	124	124	119	119	126	126
Vitamin B12	673	673	557	557	694	694	632	632	630	630	550	550
Folate	151	151	189	189	131	131	121	121	135	135	123	123
Vitamin C	115	115	225	225	226	226	202	202	183	183	182	182
Vitamin A (TRE)	264	264	494	494	397	397	342	342	294	342	293	342
Protein % of energy	22	22	21.7	21.7	25	25	23.3	23.3	24.2	24.2	24.6	24.6
Carbohydrate % of energy	40.3	40.3	41.9	41.9	38.9	38.9	40.5	40.5	39.8	39.8	37.3	37.3
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2) To assess the nutritional adequacy of these proposed intakes.

Conclusion nutritional analysis of 24 hr meal plan

- **Appropriate reduction of calorie content** to approx. 65-70% for most age groups
- Calorie restriction maybe more than intended for adult males
- Vitamin requirement generally met, apart from Vit D
- Mineral requirement is not met for several minerals (Zn, iodine, iron, calcium) in all or specific age ranges.
- Therefore, a mineral supplement is recommended

Additional information in the booklet

• General introduction about PWS

RFSULTS

- Who is it for? Anyone with PWS or carers of a child, young person or adult with PWS. It can help to explain PWS and show appropriate portion sizes to others.
- A message for people with PWS Ask for help if there is anything they do not understand
- Cooking methods Advise to use steaming, baking, grilling, boiling and microwaving rather than frying.

- Food security and food seeking tips, including advise for schools
- **Puberty** Boys in puberty may need up to an additional 250 kcal per day
- **Supplement** A vitamin and mineral supplement with iron, zinc, vitamin A and D is recommended
- Fluids Children aged 2-3 years should aim to drink 6-7 glasses a day. From 4 years onwards, 6-10 glasses a day. A glass is about 200ml.

1) To develop a national pictorial resource defining and illustrating correct portion sizes for children with PWS.

2) To assess the nutritional adequacy of these proposed intakes.

Strengths and limitations

STRENGTHS	LIMITATIONS
Visual guide	Only 60 foods
Contains commonly eaten foods	May not represent every full cultural diets
User involvement	Contain foods we wouldn't necessarily recommend
Unique	Not individualised
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USE OF RESOURCE

Use of the resource (Oct 2024-April 2025)

It is an absolutely brilliant resource, so clear and easy to understand.

I love it we use it when we do our daughters' meals, I love that there are tablespoons options really helpful for when dishing up dinner.

This would help me so I can provide myself with a balanded diet without putting myself at risk. (Adult with PWS)

PRAC	ΓICAL
P@RT	

Food Group	Number of portions each day
Vegetables	At least 3
Fruit	2
Potatoes, bread, rice, pasta and other starchy carbohydrates	6-8
Beans, pulses, fish, eggs, meat and other proteins. Include at least 1 portion of oily fish such as salmon, herring or mackerel every week.	2-3
Dairy and alternatives	3
Oils and spreads	3
Foods and drinks high in fat, salt or sugar	Not recommended

Readers around the wold

This will be a great resource to share

with the rest of the family as they never understand quantities that are suitable for our daughter Since October 2024 2,105Copies Ordered 1,413Online Reads 313Downloads

Top 10 countries UK- 545 USA - 446 Canada - 61 Ireland - 49 Australia - 41 South Korea - 38 France - 23 Finland - 19

New Zealand - 19

Netherlands - 18

CONCLUSION

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Important to get correct nutritional intake for people with PWS to support their acute and long-term health Important to get correct nutritional intake for people with PWS to support their acute and long-term health

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Unique and practical resource

- Advises portion size for PWS from 2 yrs af age
 that meets daily requirements
 - Care is required with micronutrients and individualisation remains best

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Young LR, Nestle M.

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Thank you !

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