

DIETARY MANAGEMENT OF PRADER-WILLI SYNDROME INDIVIDUALS



**ASSOC. PROF. DR. ROSLEE BIN
RAJIKAN** DIETETICS PROGRAM
FACULTY OF HEALTH SCIENCES
UNIVERSITY KEBANGSAAN MALAYSIA
KUALA LUMPUR, MALAYSIA



THE PWS RESEARCH TEAM



Dr Shazli Ezzat Ghazali
Lecturer
Clinical Psychologist
Faculty of Health Sciences UKM



Mr Azhar Talib
President
Malaysian PWS



Ms Chua Ee Lin
UKM Dietetics 3rd Year



Dr Nur Hana Hamzaid
Lecturer and Dietitian
Faculty of Health Sciences UKM



Ms Nurul Aqilah Zamri
UKM Dietetics 3rd Year



Mr Zaridah Zainuri
Clinical Dietitian
Hospital Kuala Lumpur



Ms Nur Afiqah Johari
UKM Dietetics 3rd Year



Ms Farhani Khalid
UKM Dietetics 3rd Year



Content

1

Introduction

2

Statistics

3

Characteristics

4

Nutritional Phase

5

Nutritional Status

6

**Objectives of
Dietary
Management**

7

**Growth
Monitoring**

8

Calories

9

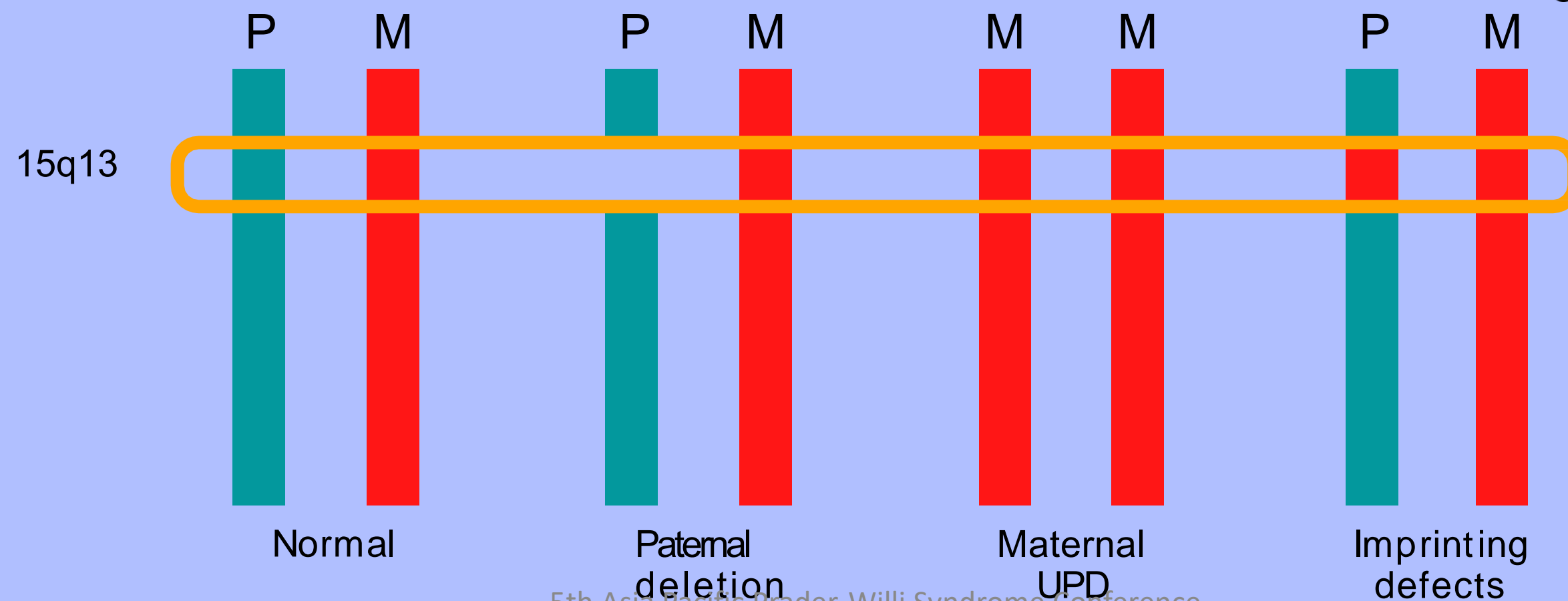
**Macronutrient
&
Micronutrient**

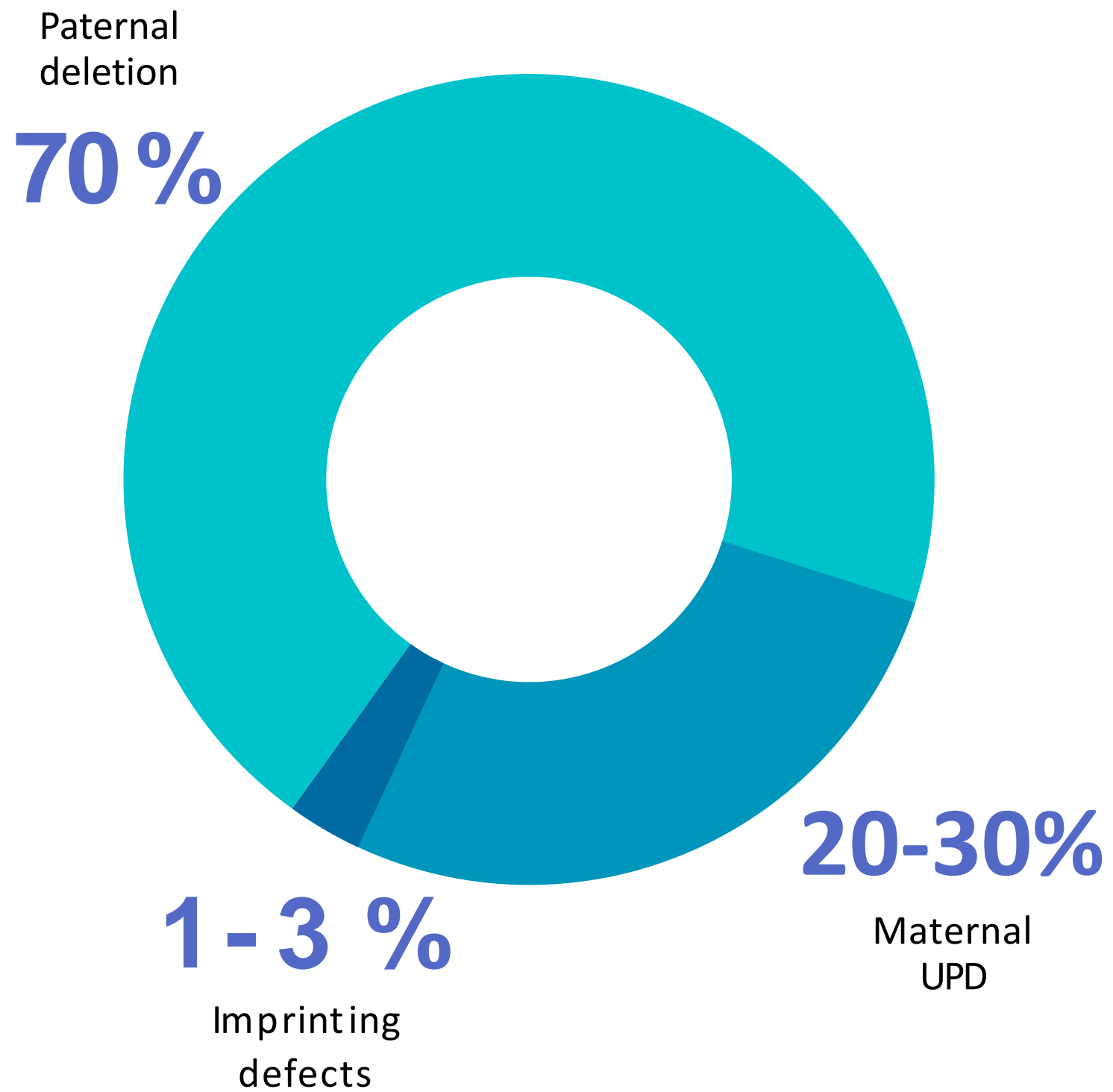
10

Physical Activities

What is Prader-Willi Syndrome?

Prader-Willi syndrome (PWS) is a **neuro-developmental genetic disorder** due to lack of expression of genes inherited from the paternal chromosome 15q11-q13 region (Butler et al. 2016)



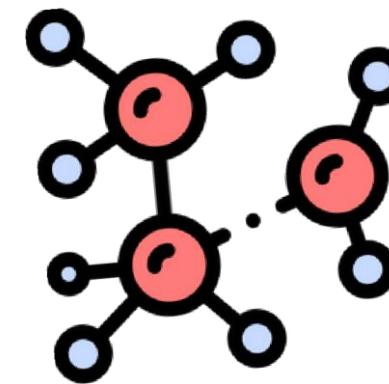


(Angulo et al. 2015; G Butler et al. 2016)

RISK FACTORS



Advanced maternal age



Father's exposure to the hydrocarbons at work



Assisted reproductive technology (ART)

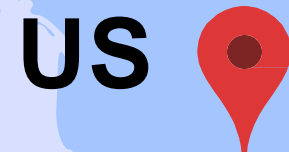
(Butler et al. 2009; Butler et al. 2016; Gold, 2014)

Statistics

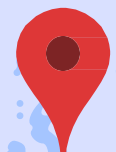
World Prevalence
Affected 350,000 to 400,000 individuals worldwide
(Butler et al. 2006)

Birth incidence
1/10,000 to 1/30,000

Estimated Population
10,000 to 20,000
(Cassidy et al. 2012; Lioni et al. 2015)



US



UK

1 in 45,000 (Whittington et al. 2001)



WESTERN JAPAN

1 in 16,000 (Ehara et al. 1995)



MALAYSIA

There are **118 active cases** of PWS individuals in 2020 (Malaysian Prader-Willi Syndrome Society 2020)



VICTORIA

Victorian Prader-Willi Syndrome Register showed that the birth prevalence was estimated to be **1:15,830** for 2003-2012 (Lioni et al. 2015)



PWS IS A RARE GENETIC DISORDER

- With no distinguishing sex, race or social status association
(Cassidy et al. 2012; Lioni et al. 2015)

CHARACTERISTICS

Infantile hypotonia



- Almond-shaped eyes
- Short upturned nose
- Thin upper lip
- Down turned corners of the mouth

Poor suck



Hyperphagia - leading to early childhood obesity

Mental deficiency; behavioral problems

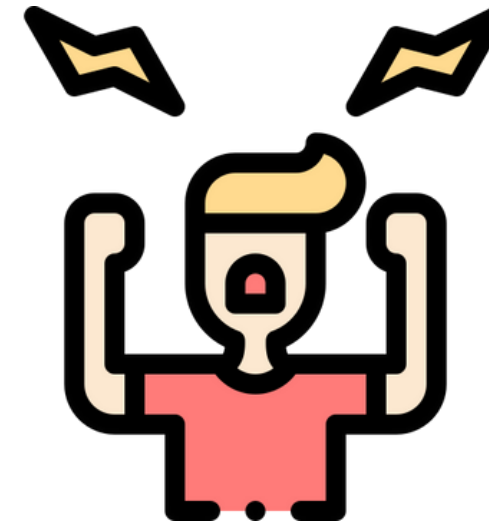


Hypogonadism/ Hypogonadism

(Butler et al.2009)

5th Asia Pacific Prader-Willi Syndrome Conference

BEHAVIOURAL PROBLEMS



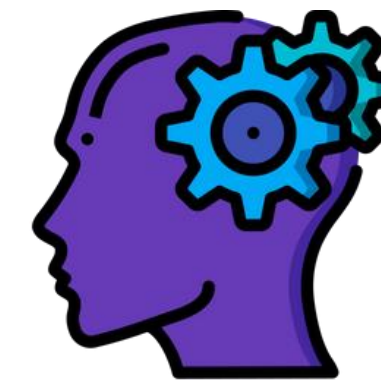
Temper tantrums



Excessive interest in food



Skin picking

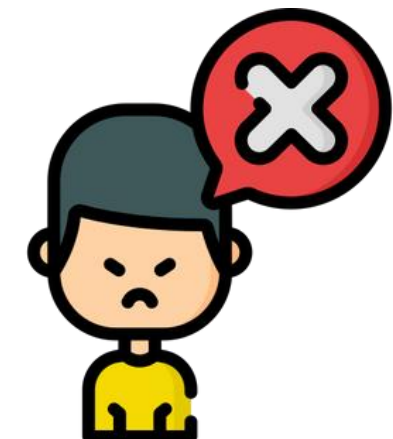


Obsessive & compulsive behaviour



Mood fluctuations

(Ho et al.2010)

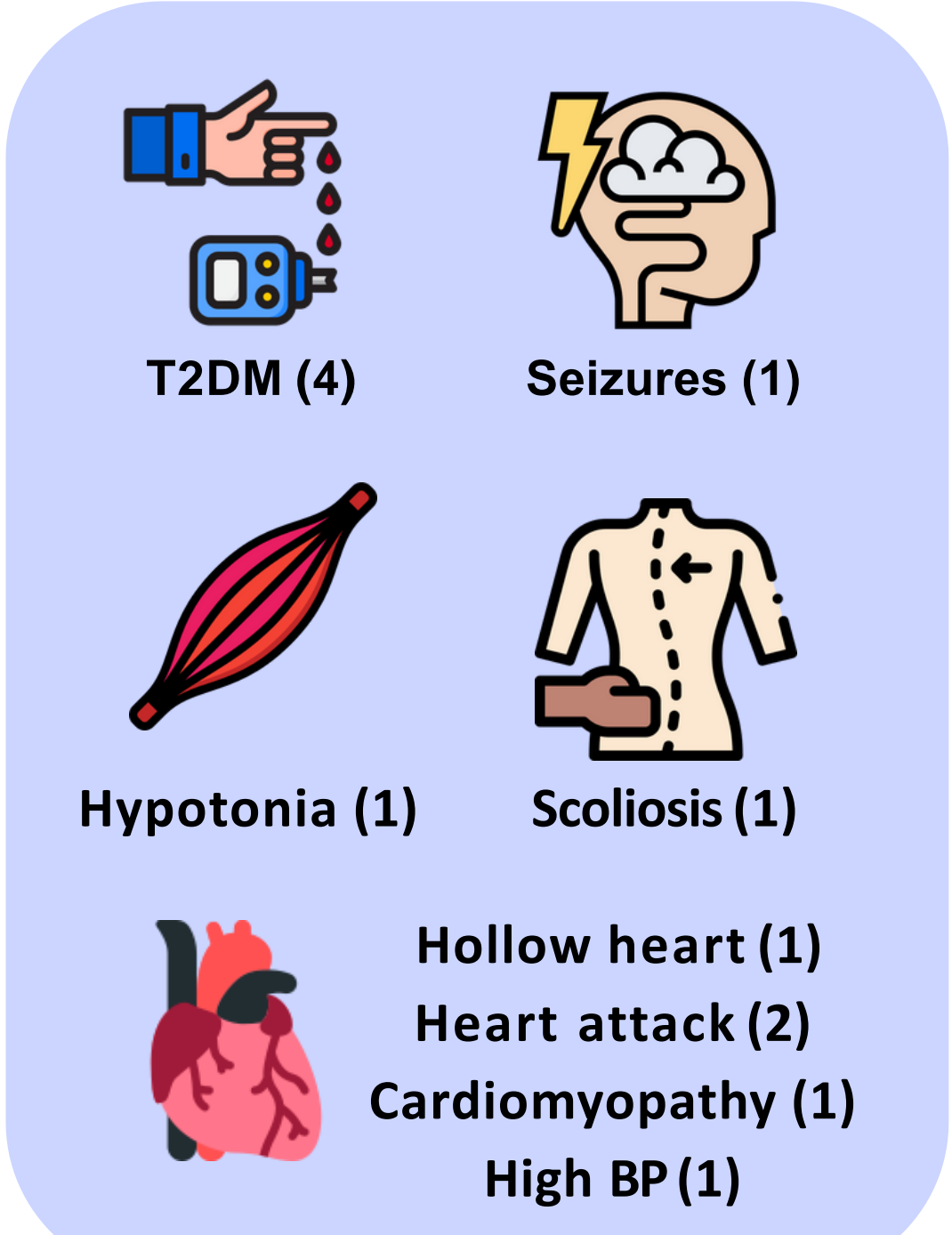
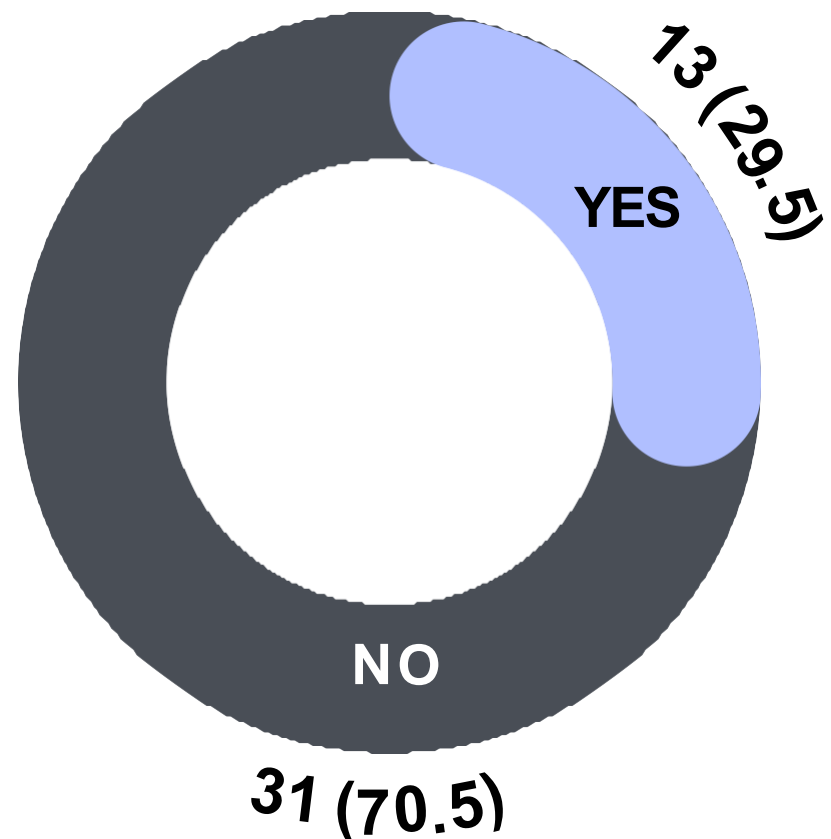


Difficulty changing routine

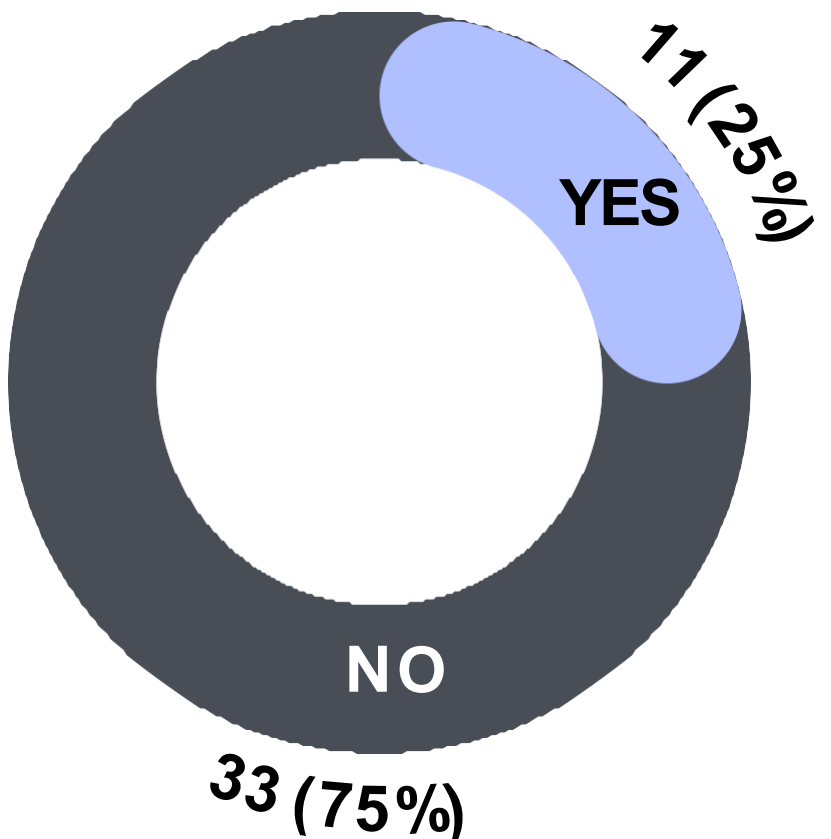
COMPLICATION RELATED TO PWS (N=44)

Majority patients not receiving GH compared to patients that receiving GH therapy.
(Tanaka et al. 2012)

Underlying disease or Symptoms, N (%)



Growth Hormone Therapy, N (%)



NUTRITIONAL PHASE IN PWS



9 KODAK 5062 PX

Phase	Median ages	Clinical characteristics
0	Prenatal to birth	Decreased fetal movements and lower birth weight than sibs
1a	0–9 months	Hypotonia with difficulty feeding and decreased appetite
1b	9–25 months	Improved feeding and appetite and growing appropriately
2a	2.1–4.5 yr	Weight increasing without appetite increase or excess calories
2b	4.5–8 yr	Increased appetite and calories, but can feel full
3	8 yr to adulthood	Hyperphagic, rarely feels full
4	Adulthood	Appetite is no longer insatiable

9 9A

^a Modified from J. L. Miller *et al.*: Nutritional phases in Prader-Willi syndrome. *Am J Med Genet A* 155A:1040–1049, 2011 (5), with

- Aggressive food seeking
- Constantly looking for opportunities to obtain food
- Sneak food and hide or hoard it
- Eat non-edible items,
- Steal food from stores or other people,
- Will steal money to buy food

(Miller & Tan 2020, Miller et al. 2011, Goldstone et al. 2012)

Consume unbelievably large quantities of food extremely quickly, even if it is spoiled, uncooked, or frozen.

(Martinez et al. 2016)

Hyperphagia Scores and sociodemographic variables (n=39)

Variables		Behaviour		Drive		Severity	
		Mean ± SD	P-value	Mean ± SD	P-value	Mean ± SD	P-value
Age	2-9 y/o (n=22)	11.55 ± 4.71	0.565 ^c	10.09 ± 2.37	0.273 ^c	5.05 ± 2.13	0.623 ^c
	10-18 y/o (n=14)	10.57 ± 3.37		9.43 ± 1.83		4.64 ± 1.99	
	19-23 y/o (n=3)	9.00 ± 4.58		12.33 ± 3.51		5.67 ± 2.52	
Gender	Male (n=20)	11.30 ± 4.01	0.655 ^b	10.05 ± 2.14	0.948 ^b	5.20 ± 2.19	0.411 ^b
	Female (n=19)	10.68 ± 4.51		10.00 ± 2.58		4.68 ± 1.95	
Races	Malay (n=29)	11.93 ± 4.20	0.017^b	10.00 ± 2.24	0.987 ^b	5.34 ± 1.86	0.079 ^b
	Non-Malay (n=10)	8.30 ± 3.06		10.10 ± 2.73		3.80 ± 2.30	

^bIndependent Samples *t* Test/Mann-Whitney U test

^cOne-way ANOVA/ Kruskal-Wallis

Malay



Non-malay

There is a **significant difference** of behaviour domain between malay and non-malay PWS ($p < 0.01$).

Hyperphagia Scores and sociodemographic variables (n=39)

Variables		Behaviour		Drive		Severity	
		Mean ± SD	P-value	Mean ± SD	P-value	Mean ± SD	P-value
Education Level	Does not go to school (n=11)	8.82 ± 3.37	0.099 ^c	9.45 ± 1.86	0.237 ^c	4.82 ± 2.48	0.672 ^c
	Primary+Kindergarten (n=18)	11.44 ± 5.09		9.83 ± 2.60		4.72 ± 1.97	
	Secondary (n=10)	12.60 ± 2.12		11.00 ± 2.21		5.50 ± 1.84	
Other disease & symptoms	Yes (n=12)	11.25 ± 3.11	0.778 ^b	9.25 ± 1.96	0.223 ^b	5.25 ± 1.77	0.642 ^b
	No (n=27)	10.89 ± 4.68		10.37 ± 2.44		4.81 ± 2.20	
Receiving growth hormone	Yes (n=11)	9.36 ± 3.14	0.131 ^b	9.18 ± 1.66	0.160 ^b	3.64 ± 1.91	0.017^b
	No (n=28)	11.64 ± 4.46		10.36 ± 2.50		5.46 ± 1.92	

^bIndependent Samples *t* Test/Mann-Whitney U test

^cOne-way ANOVA/ Kruskal-Wallis

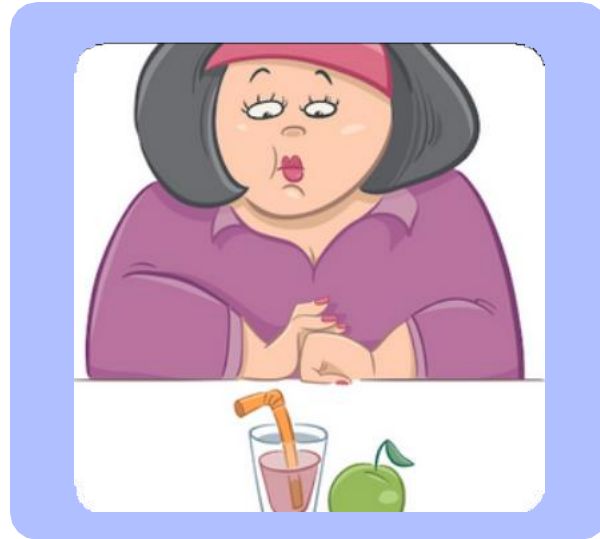
Not receive



Receive

There was a **significant difference** of severity domain between those receiving and not receiving GH (**p<0.01**).

OBESITY AMONGST PWS



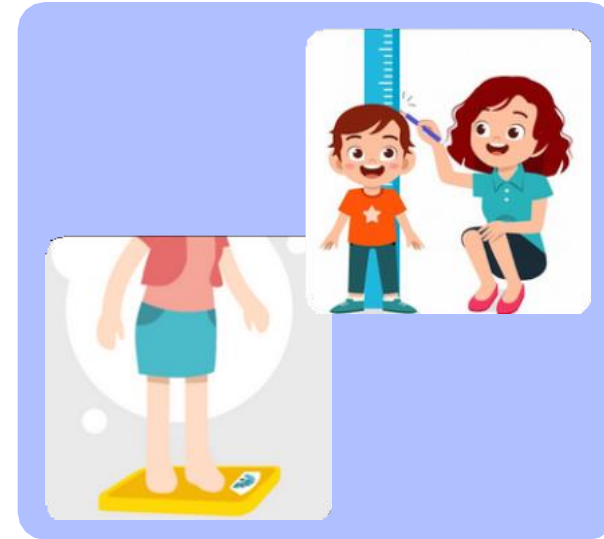
(Felix et al. 2020)

↑ Age, ↑ Weight



(Vijayakumar et al. 2018)

↑ BMI



(Jill V Butler et al. 2009)

↑ Age, ↓ Height



(Jill V Butler et al. 2009)

Inconsistence growth

Prevalence Obesity

40%

Children & Adolescent

(Damen et al. 2020)



>80%

Adult

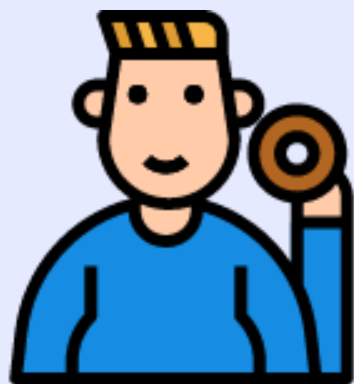
(Tan et al. 2019)



DIETARY INTAKE



PWS consume 14% fewer calories than obese; 80% parents reported PWS on restricted diet
(Rubin et al.2015)



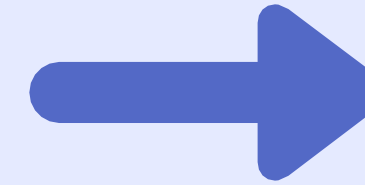
PWS with T2DM consume more carbs from high intake of white rice; >300g carb contribute to T2DM in PWS
(Vijayakumar et al. 2018)

Still did not meet recommendation, except of protein
(Rubin et al.2015)

PWS

Have low intake of fat, some parents restrict intake of fat
(Lindmark et al. 2010)

FEEDING PROBLEM



- Poor sucking
- Severe hypotonia
- Disinterest in food

Increase of obsessive of food-related behaviour with evidence of hyperphagia

(Mcallister et al.2011)



Hyperphagic behaviours such as food foraging and stealing were more prevalent

(Foerste et al. 2016)

OBJECTIVE OF DIETARY MANAGEMENT

- i. Control weight**
- ii. Support for Optimal Growth**



WEIGHT AND GROWTH MONITORING

Infant growth failure, early childhood obesity, absent pubertal growth spurt, adolescent short stature and inherent altered body composition are hallmark of PWS

Review weight regularly

Need to use PWS specific growth chart

Non GH

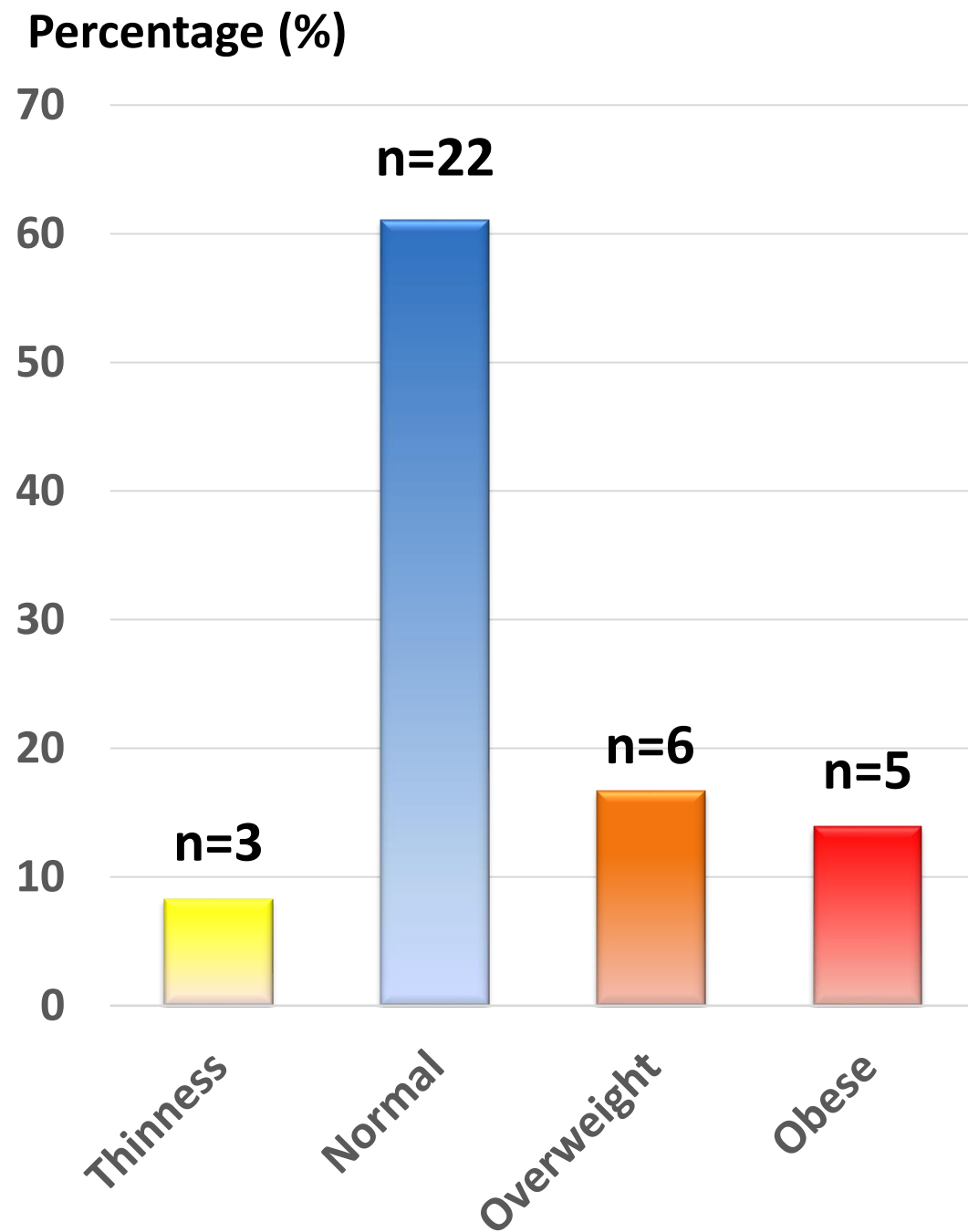
GH

(Butler et al. 2014, Mongkollarp et al. 2016)

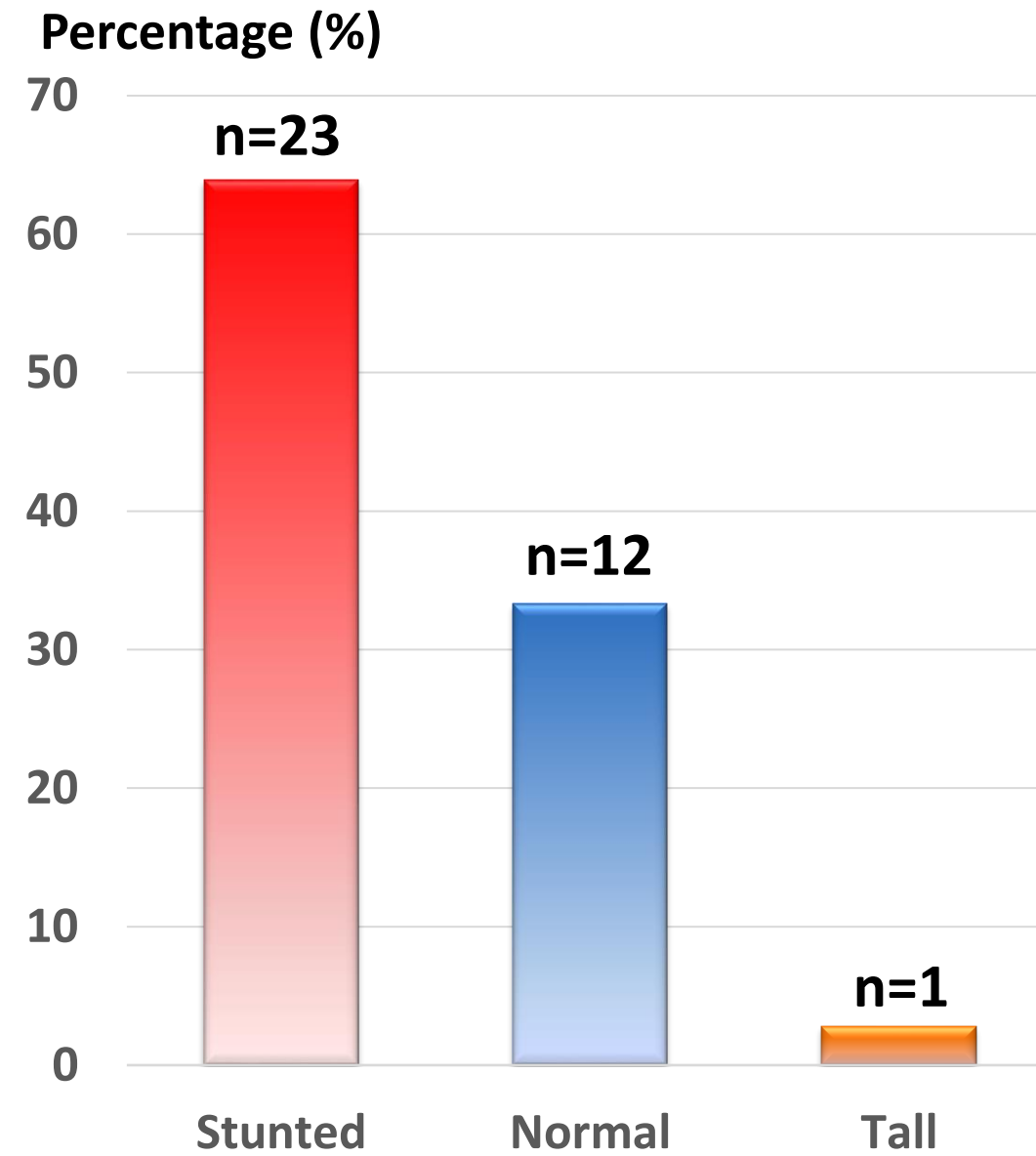
(Butler et al. 2016)

Anthropometry Data of PWS individual (n=36)

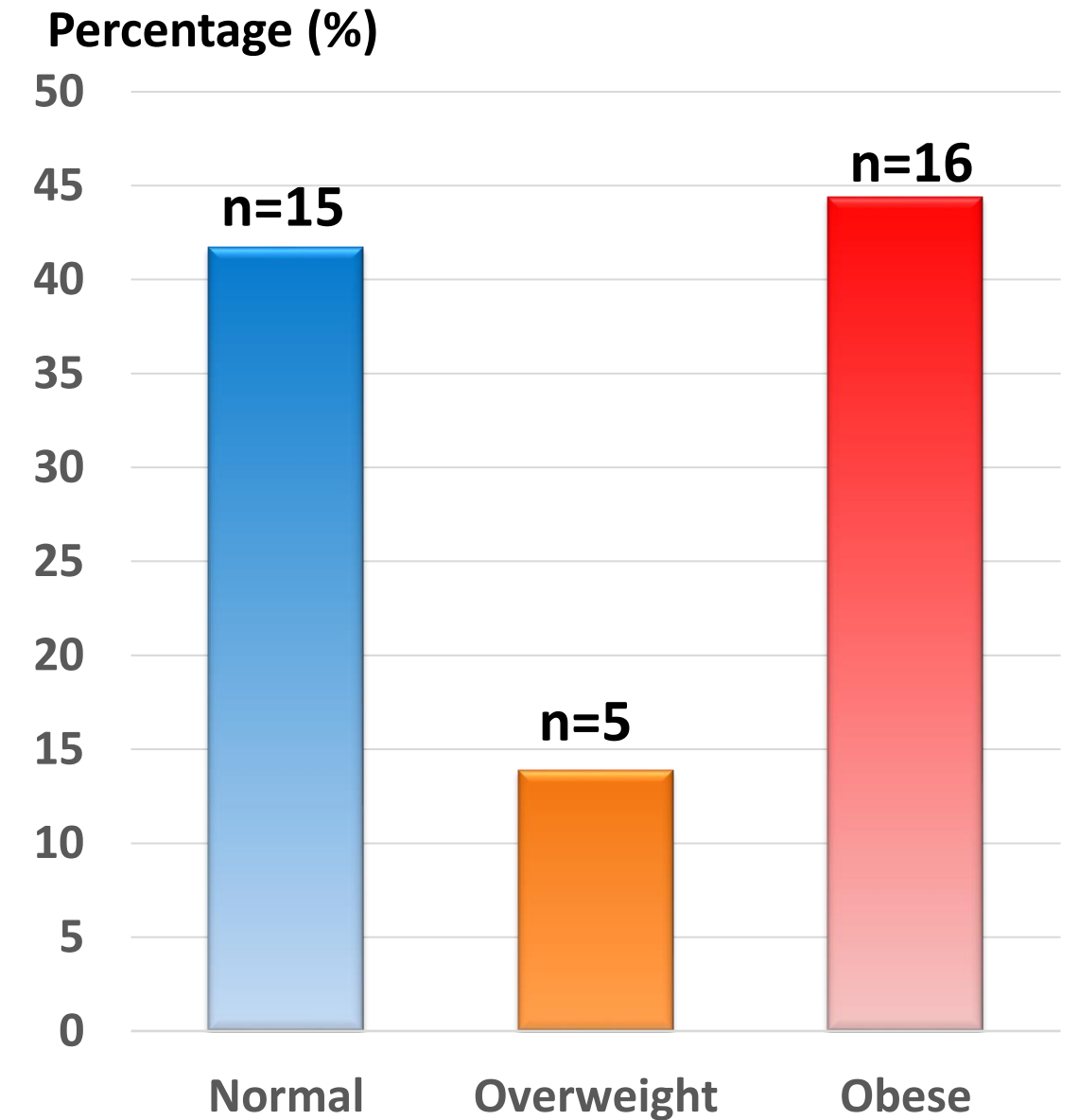
Weight-for-age



Height-for-age



BMI-for-age



DIETARY MANAGEMENT

“A reduced energy intake, well balanced diet improved weight control in children with PWS”

(Miller et al. 2013)



CALORIE INTAKE

Once excessive weight gain and hyperphagia begin, caloric intake must be significantly restricted to prevent obesity. However significant calorie restriction subsequently increases the hunger drive and behavioural problems.

(Miller and Tan 2020)



Monitor daily nutritional intake and consider calorie adjustment (60-80% from typical intake – infant and children.

(Miller 2012, Alsaif et al. 2017)

Typical person

1500-2500 kcal/day

PWS

Children and adolescent

10-11 kcal/cm for maintaining growth velocity or 8-9 kcal/cm for slow weight loss or support linear growth

(Butler et al. 2006, Lima et al. 2016, Crino et al. 2018)

Adults

1000-1200 kcal/day

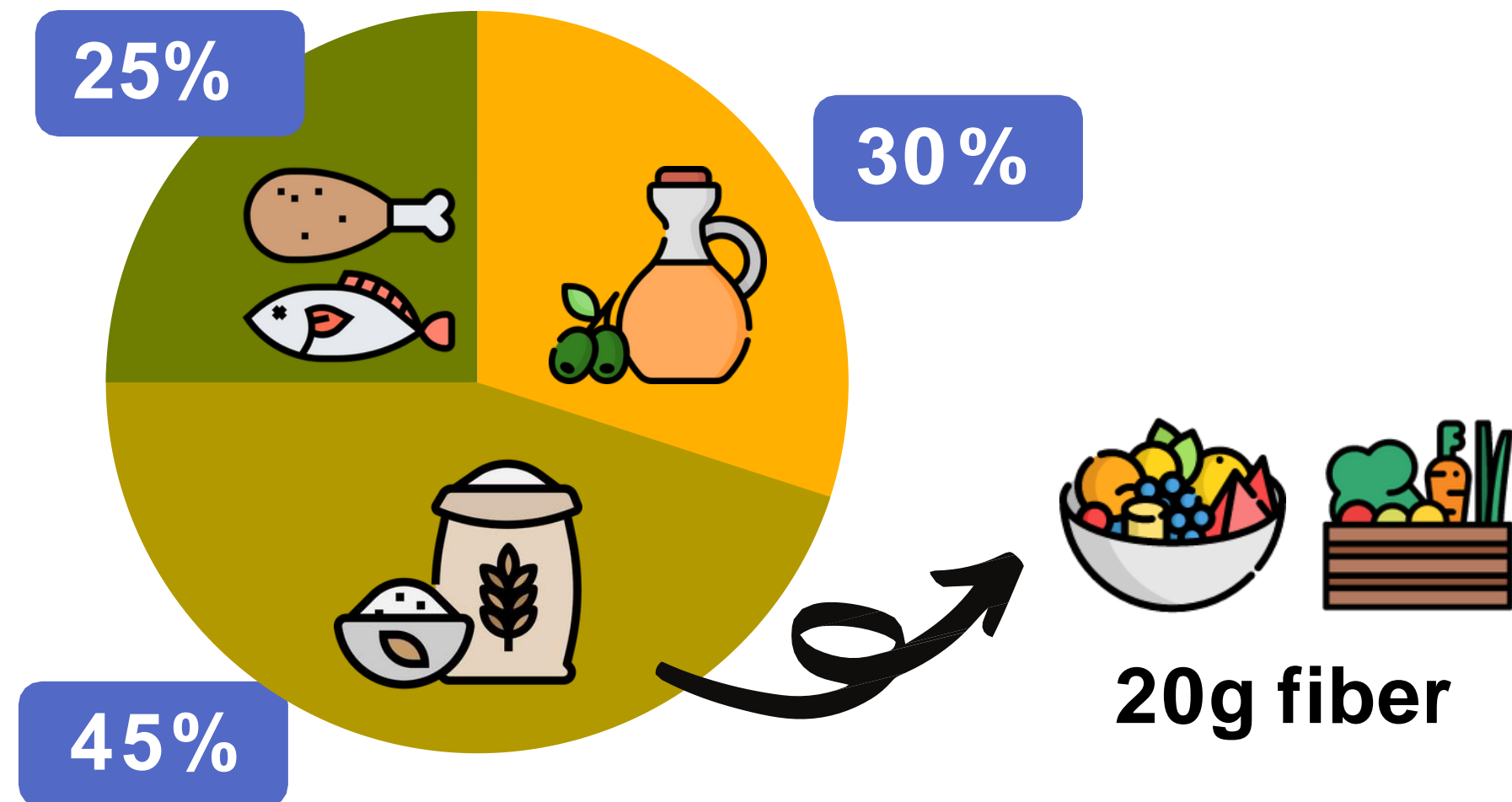
Dietary Management



1000-1200 kcal/day
(Cassidy et al. 2012)



Low-calorie and well-balanced diet in concordance with regular exercise (Ho et al. 2010)



Energy intake for PWS individuals

		Male (n=17)	Female (n=11)	p value*
Energy				
Intake (kcal/day)	TOTAL	1066 (852, 1398)	1118 (925, 1560)	.438
	with GH male (n=5) female (n=4)	1050 (806, 1328)	1370 (1146, 2372)	.086
	without GH male (n= 12) Female (n=7)	1075 (865, 1403)	943 (850, 1560)	.673
Below RNI N(%)		14 (82.4)	9 (81.8)	-
Equal and above RNI N(%)		3 (17.6)	2 (18.2)	

Data presented as median(25th, 75th percentiles). RNI, Recommended Nutrient Intake for Malaysia 2017. Energy intake is the average daily intake for each participant based on all dietary records obtained. Average energy intake was compared to the RNI for age and sex of recommendation

*Mann–Whitney U test statistical significance p < .05

Baseline energy intake of children with PWS was significantly lower compared to daily energy requirements for age- and sex-matched healthy children

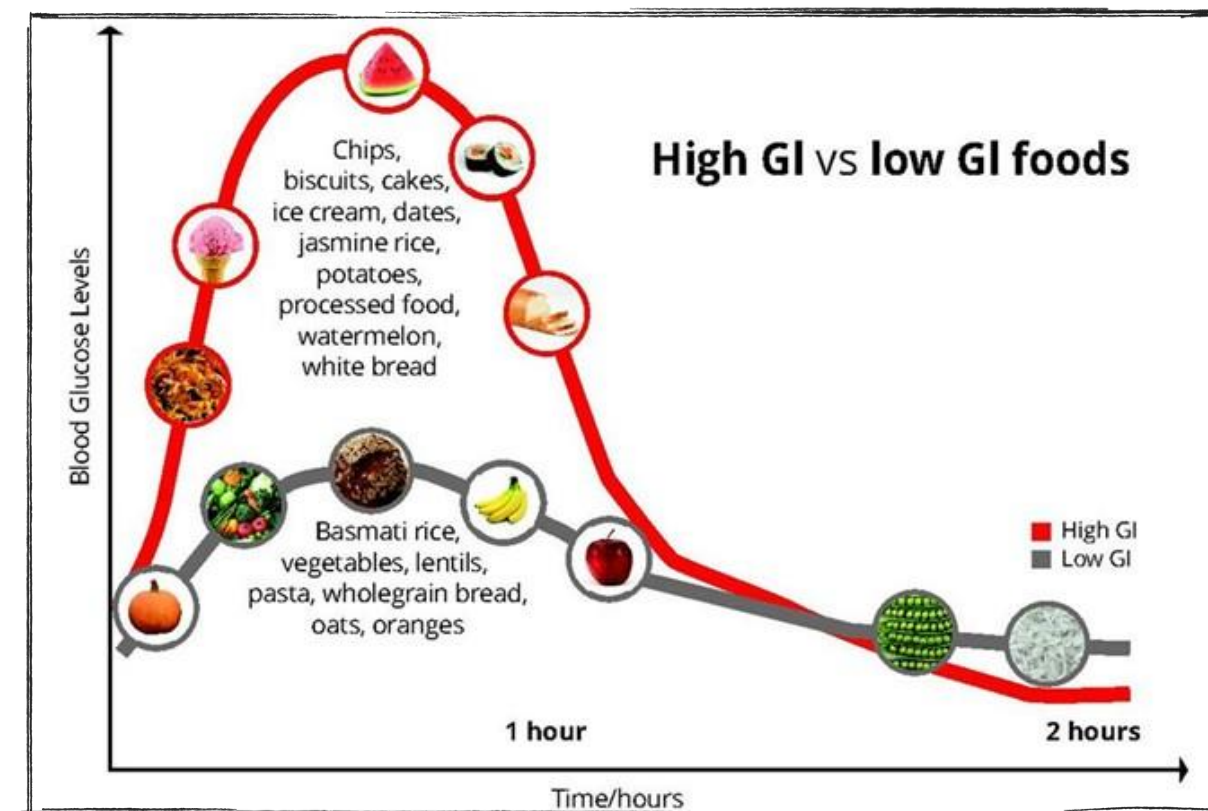
(Bakker, Siemensma, Koopman, et al. 2015)

Youth with PWS consumed 14% fewer calories than the obese group.

(Rubin et al. 2015)

MACRONUTRIENT INTAKE

- Well balanced but reduced CHO intake -->25- 30%protein, 40-45% complex CHO, 30% Fat (Miller et al. 2013)
- Avoid empty calories, Eliminate sugar, reduced processed food particularly processed CHO. Avoid high glycemic food because risk of diabetes.
- High quality Protein



Macronutrient intake for PWS individuals

	Male (n=17)	Female (n=11)	p value*
Carbohydrate Intake			
g/day	144 (129, 179)	143 (129, 219)	.557
Below recommendation/ above recommendation RNI (n)	4/2	1/0	-
% kcal	54 (50, 59)	56 (51, 59)	.869
Protein Intake			
g/day	43 (33, 55)	47 (35, 64)	.621
Below recommendation/ above recommendation RNI (n)	0/1	0/1	-
% kcal	16 (14, 19)	15 (13, 17)	.410
Fat Intake			
g/day	36 (24, 48)	30 (26, 55)	.589
Below recommendation/ above recommendation RNI (n)	4/7	3/4	-
% kcal	29 (25, 33)	29 (25, 32)	.655

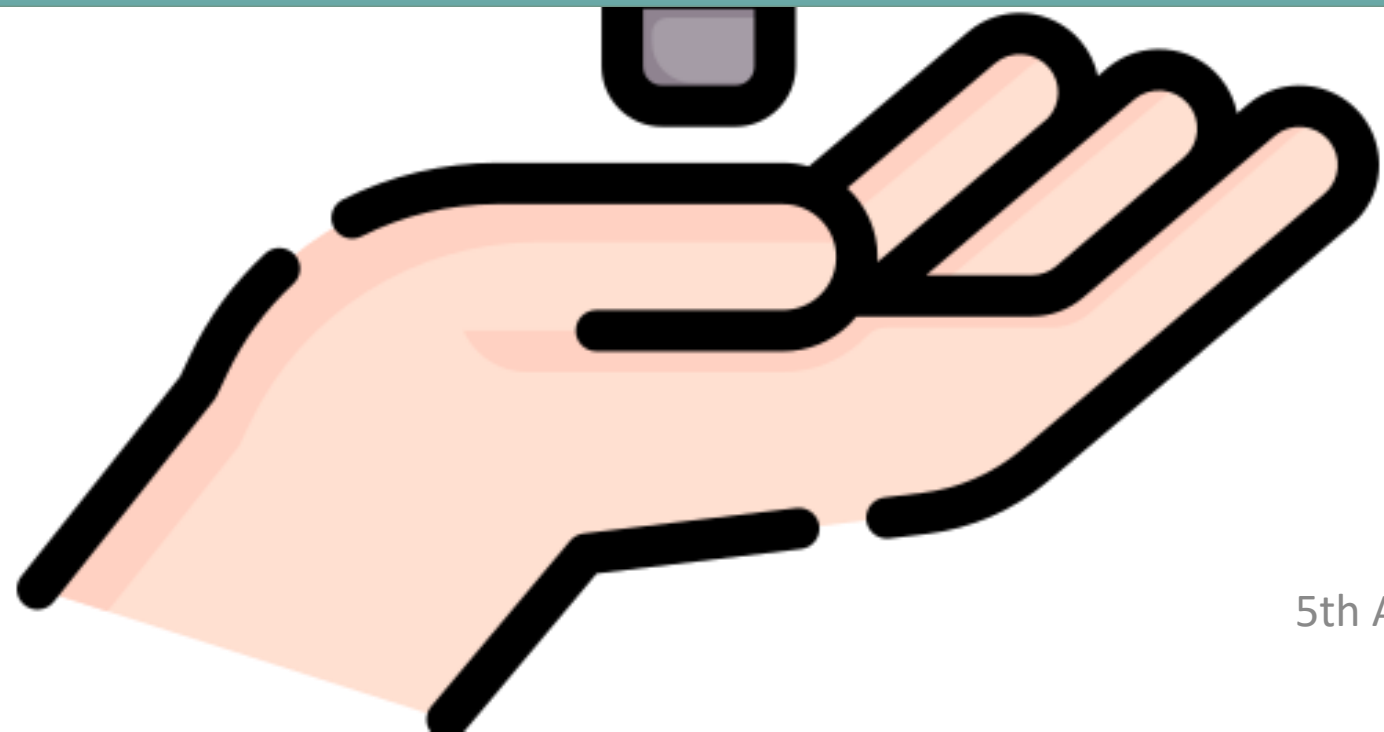
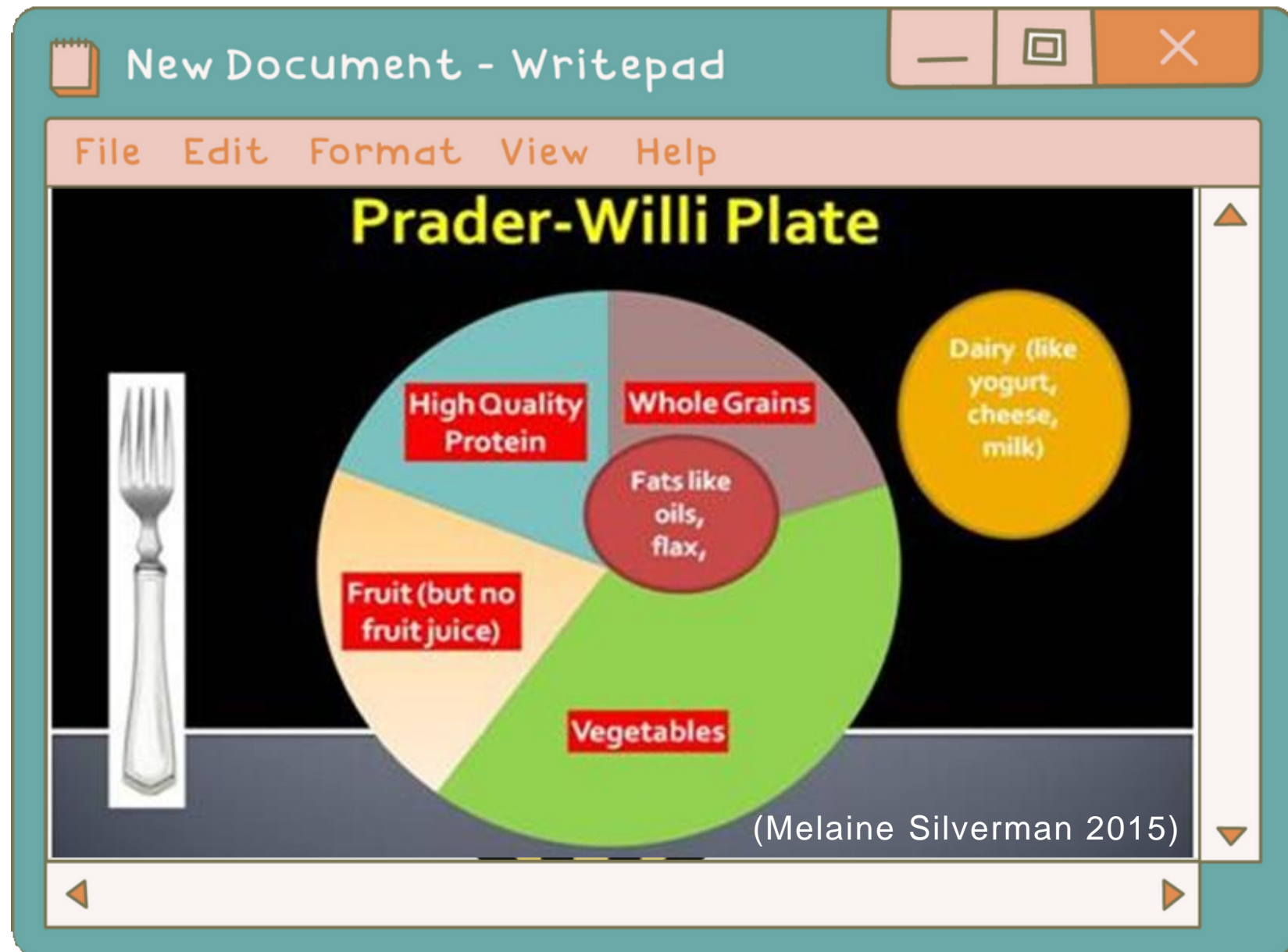
Data presented as median(25th, 75th percentiles). RNI, Recommended Nutrient Intake for Malaysia 2017. Macronutrient intake is the average daily intake for each participant based on all dietary records obtained. Normal range carbohydrate (50-65%), protein (10-20%), fat (25-30%)
*Mann-Whitney U test statistical significance p < .05

5th Asia Pacific Prader-Willi Syndrome Conference

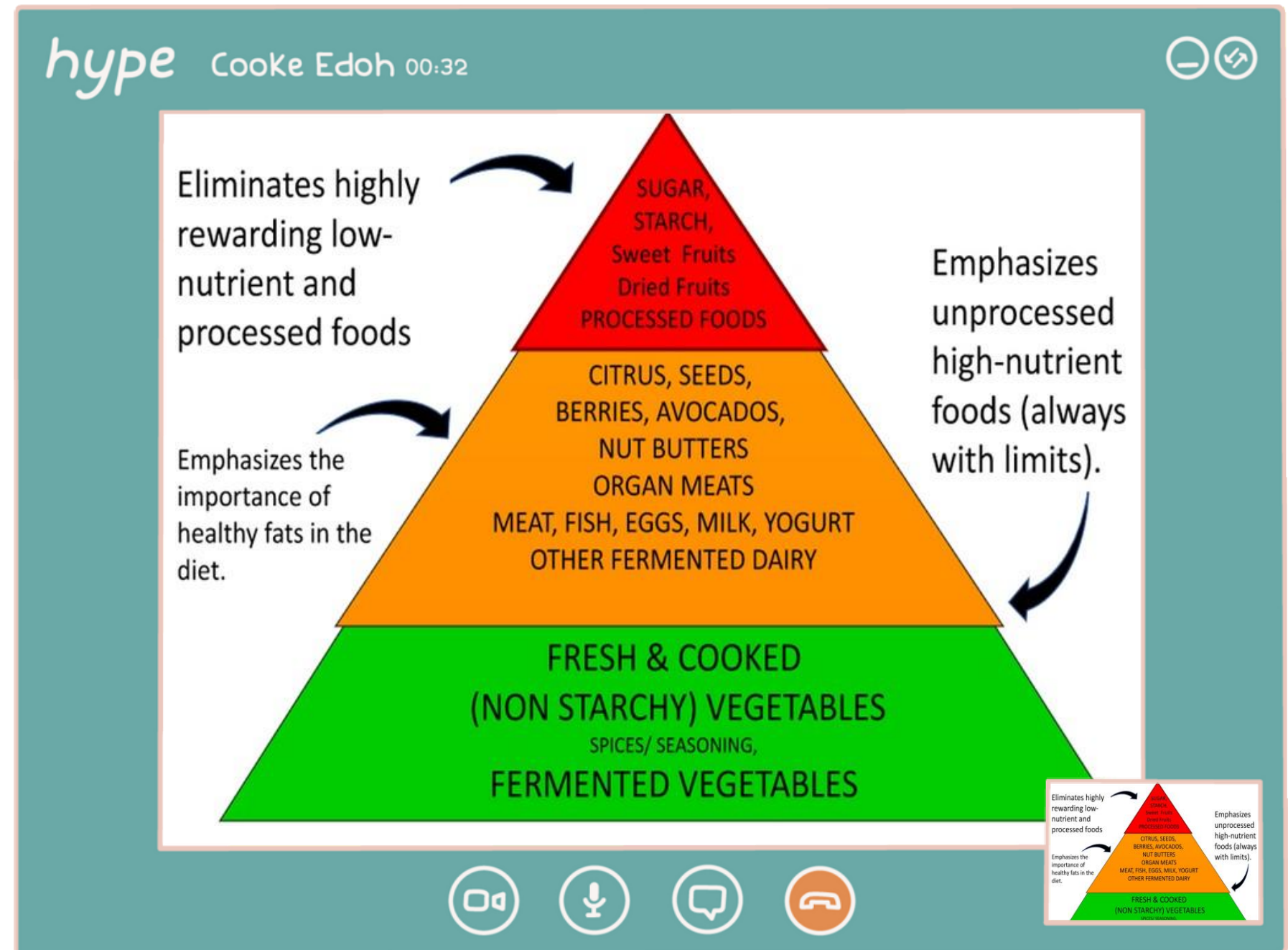
PWS appeared to consume adequate amounts of protein and the recommended proportions of carbohydrate and fat

(Rubin et al. 2015)





Proposed PWS Food Pyramid



(Linda Gourash Pittsburgh Partnership 2017)

MICRONUTRIENT INTAKE

Micronutrient requirement similar with normal child

However 3 studies (Lindmark et al. 2010, Ruben et al. 2015, Smith et al. 2017) indicate low micronutrient intake including ours



Low calorie and portion intake

HOW to improve?

✓ Sufficient Micronutrient and ✓ fiber intake

Focus on Non-Starchy vegetables

Drink water and milk (nutrient densed)



Micronutrient intake for PWS individuals

	Male (n=17)		Female (n=11)		Total (n=28)	
	Equal and above RNI N(%)	Below RNI N(%)	Equal and above RNI N(%)	Below RNI N(%)	Equal and above RNI N(%)	Below RNI N(%)
Vitamin A (µg)	5 (29.4)	12 (70.6)	3 (27.3)	8 (72.7)	8 (28.6)	20 (71.4)
Vitamin B1 (mg)	7 (41.2)	10 (58.8)	1 (9.1)	10 (90.9)	8 (28.6)	20 (71.4)
Vitamin B2 (mg)	11 (64.7)	6 (35.3)	5 (45.5)	6 (54.5)	16 (57.1)	12 (42.9)
Vitamin B3 (mg)	7 (41.2)	10 (58.8)	1 (9.1)	10 (90.9)	8 (28.6)	20 (71.4)
Vitamin B6 (mg)	9 (52.9)	8 (47.1)	2 (18.2)	9 (81.8)	11 (39.3)	17 (60.7)
Vitamin B12 (µg)	7 (41.2)	10 (58.8)	2 (18.2)	9 (81.8)	9 (32.1)	19 (67.9)
Vitamin C (mg)	12 (70.6)	5 (29.4)	5 (45.5)	6 (54.5)	17 (60.7)	11 (39.3)
Vitamin D (µg)	1 (5.9)	16 (94.1)	1 (9.1)	10 (90.9)	2 (7.1)	26 (92.9)
Vitamin E (mg)	5 (29.4)	12 (70.6)	3 (27.3)	8 (72.7)	8 (28.6)	20 (71.4)
Folate (µg)	3 (17.6)	14 (82.4)	1 (9.1)	10 (90.9)	4 (14.3)	24 (85.7)
Vitamin K (µg)	12 (70.6)	5 (29.4)	3 (27.3)	8 (72.7)	15 (53.6)	13 (46.4)

Data presented as N(%). RNI, Recommended Nutrient Intake for Malaysia 2017.

Descriptive statistics

Micronutrient intake is the average daily intake for each participant based on all dietary records obtained.

Average nutrient intake were compared to RNI 2017 based on sex and age distribution.

Micronutrient intake for PWS individuals

	Male (n=17)		Female (n=11)		Total (n=28)	
	Equal and above RNI N(%)	Below RNI N(%)	Equal and above RNI N(%)	Below RNI N(%)	Equal and above RNI N(%)	Below RNI N(%)
Pantothenic acid (mg)	2 (11.8)	15 (88.2)	1 (9.1)	10 (90.9)	3 (10.7)	25 (89.3)
Calcium (mg)	3 (17.6)	14 (82.4)	1 (9.1)	10 (90.9)	4 (14.3)	24 (85.7)
Copper (µg)	3 (17.6)	14 (82.4)	3 (27.3)	8 (72.7)	6 (21.4)	22 (78.6)
Iron (mg)						
10%	12 (70.6)	5 (29.4)	6 (54.5)	5 (45.5)	18 (64.3)	10 (35.7)
15%	13 (76.5)	4 (23.5)	8 (72.7)	3 (27.3)	21 (75)	7 (25)
Magnesium (mg)	4 (23.5)	13 (76.5)	2 (18.2)	9 (81.8)	6 (21.4)	22 (78.6)
Manganese (mg)	0	17 (100)	0	11 (100)	0	28 (100)
Phosphorus (mg)	8 (47.1)	9 (52.9)	5 (45.5)	6 (54.5)	13 (46.4)	15 (53.6)
Potassium (g)	17 (100)	0	11 (100)	0	28 (100)	0
Selenium (ug)	10 (58.8)	7 (41.2)	6 (45.5)	5 (54.5)	16 (57.1)	12 (42.9)
Zinc (mg)	7 (41.2)	10 (58.8)	5 (45.5)	6 (54.5)	12 (42.9)	16 (57.1)
Sodium (mg)	12 (70.6)	5 (29.4)	7 (63.6)	4(36.4)	19 (67.9)	9 (32.1)

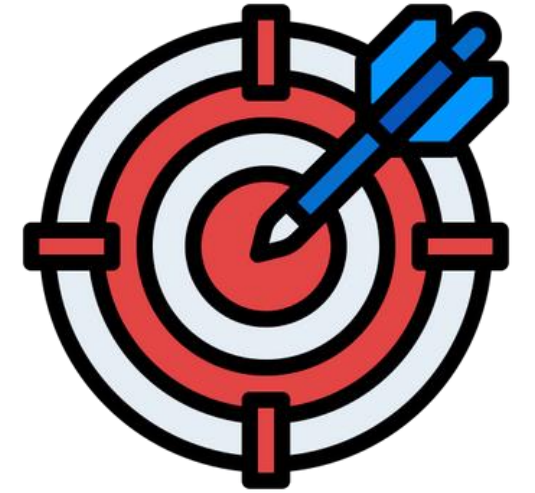
Data presented as N(%). RNI, Recommended Nutrient Intake for Malaysia 2017.

Descriptive statistics

Micronutrient intake is the average daily intake for each participant based on all dietary records obtained. Average nutrient intake were compared to RNI 2017 based on sex and age distribution.

Low intake of several essential nutrients is mainly due to the reduced intake of energy (Lindmark et al.2010)

CHALLENGES



BEHAVIOUR PROBLEMS

including
dysfunctional
satiation rather
than excessive
hunger

Preference for
rigid schedule

Resistance to
change

Food
scavenging

Ingestion of
inappropriate
food **GARBAGE**

Leftover
food

Stealing
food

Scraps off the
floor

Tips on good eating behaviors for Prader-Willi syndrome patients

Start early on healthy foods and regular mealtimes as soon as child eat solid

Plan and structure meals include other activities such exercise and breaks

BE AWARE OF SMELLS



DONT DELAY SNACK



Develop a small set of meals and snacks on rotation and become familiar with what the appropriate portion size looks like

Use smaller plates or portion containers

AVOID USING FOOD AS REWARDS OR INCENTIVES

Provide a wide choice of healthy foods, especially a large variety of vegetables, salads, fruits, and lean meat



Secure food accessibility by locking cabinets/fridge

Avoid giving extra treats



LIMIT THE VISIBILITY OF FOOD

Supervise food exposure



Post meal schedule/menu

No unsupervised access to food

AVOID GIVING MONEY

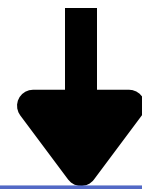


SPECIAL DIETS



Low Carbohydrate Diet

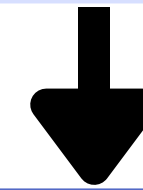
Carbohydrate-restricted diet (ketogenic diet) for weight control in adolescents with PWS (15% carbohydrates, 65% fat, and 20% protein)



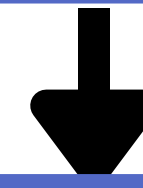
Effective short term in glycemic and weight control (Irizarry et al 2019) but deficiencies in vitamin D, calcium, and fiber and difficult to sustain in long term (Ferraris et al. 2019)



Modified Atkin Diets (Felix et al. 2020)



10–15 g of net carbohydrates with unrestricted or customized protein and fat in children + Supplement



Small sample size, compliance issue, weight stable/loss, elevation of LDL and TC



Physical Activity



At least **60 minutes** of moderate to vigorous activity per day.

(MDG 2013)



↑ Age ↓ Vigorous Activity

5-10 years old

Engaged in more vigorous PA than those 18+ years old

10-17 years old

Engaged in more moderate intensity PA per week than children 5-10 years old

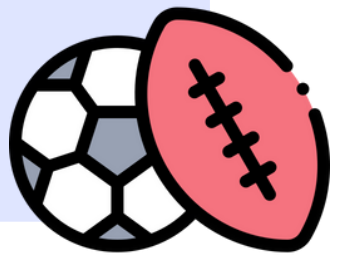
5th Asia Pacific Prader-Willi Syndrome Conference
(Elena et al. 2012)

(Rubin et al. 2012)

YOUTH WITH PWS < OBESSE YOUTH WITHOUT PWS

Youth with PWS engaged in less overall PA than obese youth without PWS

(Castner et al. 2014)



They may be at a disadvantage due to **poor stamina** as well as **cardiovascular fitness** and **low motor proficiency**.

(Castner et al. 2014)

PA & Sports are fundamental therapy for PWS patients

Improves physical functions

Promotes socialization

Helps improve caloric expenditure, together with diet

Is one of the best ways to limit access to food



PAL Scores and its comparison with sociodemographic variables (n=39)

PAL Categories		Low (≤1.44) (n=23)		Moderate (1.45 – 2.37) (n=13)		High (≥2.38) (n=3)		Overall PAL (n=39)	
		N (%)	P-value	N (%)	P-value	N (%)	P-value	Mean ± SD	P-value
Gender	Male (n=20)	11 (47.8)	0.807 ^a	7 (53.8)	0.807 ^a	2 (66.7)	0.807 ^a	2.02 ± 0.65 ✓	0.614 ^b
	Female (n=19)	12 (52.2)		6 (46.2)		1 (33.3)		1.91 ± 0.64	
Races	Malay (n=29)	19 (82.6)	0.362 ^a	8 (61.5)	0.362 ^a	2 (66.7)	0.362 ^a	1.85 ± 0.64	0.050 ^b
	Non-Malay (n=10)	4 (17.4)		5 (38.5)		1 (33.3)		2.30 ± 0.51 ✓	
Education level	Did not go to school (n=11)	7 (30.4)	0.856 ^a	4 (30.8)	0.856 ^a	0 (0)	0.856 ^a	1.87 ± 0.60	0.641 ^c
	Kindergarten/Primary (n=18)	10 (43.5)		6 (46.2)		2 (66.7)		2.07 ± 0.65 ✓	
	Secondary (n=10)	6 (26.1)		3 (23.1)		1 (33.3)		1.88 ± 0.69	
Other underlying disease/ syndrome	Yes (n=12)	7 (30.4)	0.428 ^a	5 (38.5)	0.428 ^a	0 (0)	0.428 ^a	1.80 ± 0.55	0.285 ^b
	No (n=27)	16 (69.6)		8 (61.5)		3 (100)		2.04 ± 0.67 ✓	
Growth hormone therapy	Yes (n=11)	5 (21.7)	0.156 ^a	6 (46.2)	0.156 ^a	0 (0)	0.156 ^a	2.08 ± 0.54 ✓	0.513 ^b
	No (n=28)	18 (78.3)		7 (53.8)		3 (100)		1.92 ± 0.67	
Weight status	Underweight / Normal (n=12)	9 (39.1)	0.331 ^a	2 (15.4)	0.331 ^a	1 (33.3)	0.331 ^a	1.79 ± 0.67	0.263 ^b
	Overweight / Obese (n=27)	14 (60.9)		11 (84.6)		2 (66.7)		2.04 ± 0.62 ✓	

^aChi-Square Test ^bIndependent Samples t Test ^cOne-way ANOVA test

Youth with PWS engaged in **less overall PA**
(Castner et al. 2014)



EXERCISE

- Aims for **30 minutes** of exercise daily
- Physical activity is recommended as an adjunctive therapy to maximize lean mass and efforts at body weight maintenance in PWS.



ASSOCIATION

Hyperphagia and BMI

Correlation between hyperphagia domains with BMI

Variables	Hyperphagia					
	Behaviour		Drive		Severity	
	r_s	P-value	r_s	P-value	r_s	P-value
BMI	.348*	.030	.258	.112	.379*	.017

r_s = Spearman correlation coefficient

*. Correlation is significant at the 0.05 level (2-tailed).

There was a **significant difference** of severity and behaviour domain between BMI.



Correlation between hyperphagia domain and BMI among adult shows **no significant result** (Jauregi et al. 2013)



Quality of Life (QOL)

Quality of life is an individual's perception of their place in life in the context of the culture and value system in which they live and is related to their goals, expectations, standards and concerns (WHO 2014)



Caregivers'

QOL PWS CAREGIVERS < QOL HEALTHY NORMS

(Mao et al. 2019)

PWS Individuals'

QOL CHILD WITH PWS < QOL CHILD WITHOUT PWS

(Wilson et al. 2016)

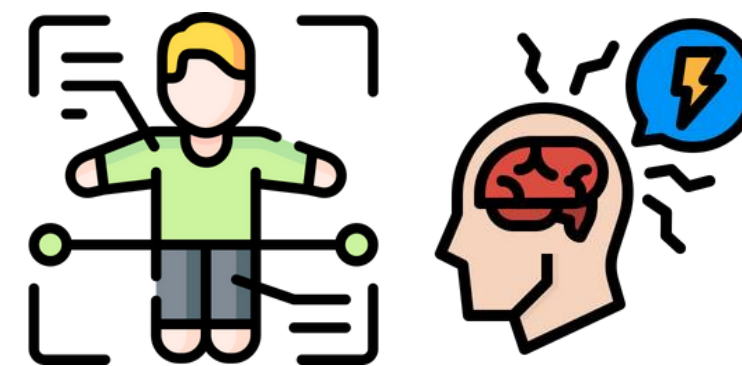
FACTORS AFFECTING QOL

Children's ages

Children with combined diseases or symptoms

Caregivers having concerns about children

Children's social adaptation ability



Physical and mental aspects of quality of life are impaired in Prader-Willi patients



QOL can be increased during growth hormone (GH) treatment

TAKE HOME MESSAGES

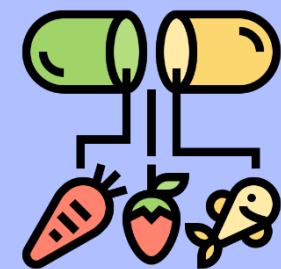
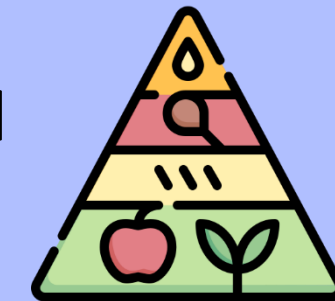


Review weight regularly

Low calorie



Well balanced diet

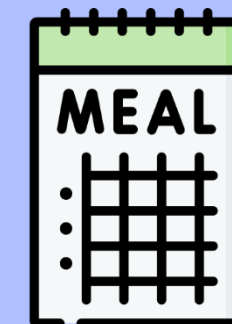


Focus on micronutrient

Practise healthy eating behaviour earlier



Structured and plan meals



Exercise regularly

**THANK
YOU**

REFERENCES

- Adams, D., Rose, J., Jackson, N., Karakatsani, E. & Oliver, C. 2018. Coping Strategies in Mothers of Children with Intellectual Disabilities Showing Multiple Forms of Challenging Behaviour: Associations with Maternal Mental Health. *Behav Cogn Psychother* 46(3): 257-275.
- Bakker, N., Siemensma, E., Koopman, C. & Hokken-Koelega, A. 2015. Dietary Energy Intake, Body Composition and Resting Energy Expenditure in Prepubertal Children with Prader-Willi Syndrome before and During Growth Hormone Treatment: A Randomized Controlled Trial. *Hormone research in paediatrics* 83(5):321-331.
- Butler, J. V., Whittington, J. E., Holland, A. J., Mcallister, C. J. & Goldstone, A. P. 2010. The Transition between the Phenotypes of Prader-Willi Syndrome During Infancy and Early Childhood. *Developmental Medicine Child Neurology* 52(6): e88-e93.
- G Butler, M., M Manzardo, A. and L Forster, J., 2016. Prader-Willi syndrome: clinical genetics and diagnostic aspects with treatment approaches. *Current pediatric reviews*, 12(2), pp.136-166.
- Caliandro, P., Grugni, G., Taruscio, D., Kodra, Y., & Padua, L. (2011). Quality of Life Assessment in Prader–Willi Syndrome. In *Handbook of Behavior, Food and Nutrition* (pp. 3153-3162). Springer, New York, NY.
- Castner, D.M., Tucker, J.M., Wilson, K.S. & Rubin, D.A. 2014. Patterns of Habitual Physical Activity in Youth With and Without Prader-Willi Syndrome. *Research in Developmental Disabilities* 35(11): 3081-3088.
- Crinò, A., Fintini, D., Bocchini, S. and Grugni, G., 2018. Obesity management in Prader–Willi syndrome: current perspectives. *Diabetes, metabolic syndrome and obesity: targets and therapy*, 11,p.579.
- Felix, G., Kossoff, E., Barron, B., Krekel, C., Testa, E. G. & Scheimann, A. J. O. J. O. R. D. 2020. The Modified Atkins Diet in Children with Prader-Willi Syndrome. *Orphanet Journal of Rare Diseases* 15(1): 1-5.
- Jauregi, J., Laurier, V., Copet, P., Tauber, M. & Thuilleaux, D. 2013. Behavioral Profile of Adults with Prader-Willi Syndrome: Correlations with Individual and Environmental Variables. *Journal of Neurodevelopmental Disorders* 5(1): 1-10.
- Lindmark, M., Trygg, K., Giltvedt, K. & Kolset, S. 2010. Nutrient Intake of Young Children with Prader–Willi Syndrome. *Food nutrition research* 54(1): 212.
- Mao, S. J., Shen, J., Xu, F., & Zou, C. C. (2019). Quality of life in caregivers of young children with Prader–Willi syndrome. *World Journal of Pediatrics*, 15(5), 506-510.

REFERENCES

- Miller, J. L., Lynn, C. H., Driscoll, D. C., Goldstone, A. P., Gold, J. A., Kimonis, V., Dykens, E., Butler, M. G., Shuster, J. J. & Driscoll, D. J. 2011. Nutritional Phases in Prader–Willi Syndrome. *American Journal of Medical Genetics Part A* 155(5): 1040-1049.
- Nayak, M. & Narayan, K. 2019. Strengths and Weakness of Online Surveys. *IOSR Journal of Humanities Social Science* 24(5): 31-38.
- Miller, J.L. and Tan, M., 2020. Dietary Management for Adolescents with Prader–Willi Syndrome. *Adolescent Health, Medicine and Therapeutics*, 11,p113.
- Rubin, D. A., Nowak, J., McLaren, E., Patiño, M., Castner, D. M. & Dumont-Driscoll, M. C. 2015. Nutritional Intakes in Children with Prader–Willi Syndrome and Non-Congenital Obesity. *Food nutrition research* 59(1): 29427.
- Shivers, C. M., Leonczyk, C. L. & Dykens, E. M. 2016. Life Satisfaction among Mothers of Individuals with Prader-Willi Syndrome. *Journal of autism developmental disorders* 46(6): 2126-2137.
- Tan, Q., Orsso, C.E., Deehan, E.C., Triador, L., Field, C.J., Tun, H.M., Han, J.C., Müller, T.D. and Haqq, A.M., 2020. Current and emerging therapies for managing hyperphagia and obesity in Prader-Willi syndrome: A narrative review. *Obesity Reviews*, 21(5), p.e12992.
- Vijayakumar, P., Narayanasamy, A. & Vellingiri, B. 2018. Excessive Carbohydrate Consumption and Body Mass Index: The Risk Factors for Type 2 Diabetes Mellitus in Patients with Prader-Willi Syndrome in Tamil Nadu Population. *The Journal of Basic Applied Zoology* 79(1): 54.
- Wilson, K. S., Wiersma, L. D., & Rubin, D. A. (2016). Quality of life in children with Prader Willi syndrome: Parent and child reports. *Research in developmental disabilities*, 57, 149-157.
- Wong, S. B., Wang, T. S., Tsai, W. H., Tzeng, I. S. & Tsai, L. P. 2020. Parenting Stress in Families of Children with Prader–Willi Syndrome. *American Journal of Medical Genetics Part A*.
- Zyga, O. & Dimitropoulos, A. 2020. Preliminary Characterization of Parent-Child Interaction in Preschoolers with Prader-Willi Syndrome: The Relationship between Engagement and Parental Stress. *American journal on intellectual developmental disabilities* 125(1): 76-84.