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ORIGINAL ARTICLE

Dietary intake in youth with Prader-Willi syndrome

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Dietary management is important to prevent severe obesity in individuals with Prader-Willi syndrome (PWS); however, few studies have examined dietary intake and quality in youth with PWS. Our objective was to estimate intake of essential nutrients and diet quality in youth with PWS compared to those without PWS. Three-day food records were used to estimate intake of energy, nutrients, nutrient-density, foods, and adherence to healthy eating guidelines. Data were presented as medians and interquartile ranges with Mann–Whitney *U* and Fisher's test used to compare between groups with $p < .05$ considered significant. Youth with ($n = 23$) and without ($n = 23$) PWS were similar in age and sex distribution. The PWS group had a lower energy intake ($p \le .001$), higher nutrient density ($p = .003$), and better adherence to guidelines ($p = .007$) compared to the control group. The proportion with nutrient intake from food below Estimated Average Requirement or Adequate Intake were similar between groups. Fiber, vitamin D, calcium, and potassium intake were below recommendations in 50% or more in both groups. The inclusion of supplement intake lowered the proportion below recommendations, except for fiber and potassium. Youth with PWS had a similar nutrient intake as those without PWS despite a lower energy intake, which could be attributed to higher diet quality. However, more than half of youth with PWS were at risk of inadequate fiber, vitamin D, calcium, and potassium intake. A greater emphasis on nutrient-dense foods would improve nutrient intake, but supplements may be warranted in youth with PWS who do not meet recommendations.

KEYWORDS
childhood obesity, diet quality, nutrient intake, Prader-Willi syndrome

1 | INTRODUCTION

Prader-Willi syndrome (PWS) is a neurodevelopmental disorder that occurs in approximately 1 in 10,000 to 30,000 births as a result of an alteration of the active paternally expressed genes from the chromosome 15q11-q13 region. PWS is the most common syndromic form of childhood obesity characterized by neonatal hypotonia, failure to thrive, developmental and cognitive delay, behavioral problems, and hyperphagia (Cassidy, Schwartz, Miller, & Driscoll, 2012). Hyperphagia in PWS is associated with a marked food preoccupation and lack of satiety (Cassidy et al., 2012; Martinez Michel, Haqq, & Wismer, 2016; Miller et al., 2011). In addition to hyperphagia, total daily energy expenditure is estimated to be 20%–50% lower for individuals with PWS compared to individuals without PWS (Alsaif et al., 2017).

Dietary management is important in the treatment of PWS to prevent obesity and associated health risks such as insulin resistance, Type 2 diabetes, and cardiovascular disease. Various types of diets have been developed for individuals with PWS (Altman, Bondy, & Hirsch, 1978; Balko, 2006; Bistrian, Blackburn, & Stanbury, 1977; Kriz & Cloninger, 1981; Miller, Lynn, Shuster, & Driscoll, 2013). Although diets recommended for individuals with PWS differ in macronutrient composition and/or foods, a common feature is limiting energy intake to less than what would be recommended for healthy youth of the same age.

It is assumed that essential macronutrient and micronutrient intake of youth with PWS is similar to healthy youth. However, with diets of lower energy content, the importance of selecting nutrient-dense foods to ensure diets meet recommendations for essential nutrients is challenging. Few studies have examined nutrient intake

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(Lindmark, Trygg, Giltvedt, & Kolset, 2010; Nordstrom, Paus, Andersen, & Kolset, 2015; Rubin et al., 2015) or diet quality in youth with PWS. The purpose of this study was to estimate intake of essential nutrients from foods and diet quality in youth with PWS compared to those without PWS.

2 | MATERIALS AND METHODS

2.1 | Study population

Dietary records obtained from multiple studies were used for this analysis. Participants with PWS were recruited through the pediatric endocrinology clinic at the Stollery Children's Hospital (Edmonton, Alberta, Canada) and PWS organizations through advertisements in newsletters, as well as word of mouth. PWS diagnosis was confirmed by DNA methylation and fluorescence in situ hybridization genetic analysis results within medical records.

The control group was partially composed of siblings or acquaintances of PWS participants, whereas others were recruited through advertisements in a local newspaper. Parents were asked to complete a medical history questionnaire that included information about participants' medication use. All studies were approved by the Health Research Ethics Board at the University of Alberta.

2.2 | Anthropometrics

Weight and height were measured by trained individuals following an overnight fast (~10 hr). Body weight was measured in duplicate to the nearest 0.1 kg using a digital scale (Pelstar, Bridgeview, IL), and height was measured in duplicate to the nearest 0.1 cm with a Digi-kit stadiometer (Measurement Concepts and Quick Medical, North Bend, WA). Participants wore light clothing and removed their shoes for the measurements. Self-reported weight and height measurements were obtained from some participants (n = 1 PWS and n = 8 controls). Body mass index (BMI) z-scores were calculated using EpiInfo (CDC, Atlanta, GA).

2.3 | Dietary records

One to three 3-day food records were completed by the participant or the parent of the participant. Participants with multiple 3-day food records completed each record at least 2 weeks apart within a 6-month period. Instructions were provided to the participant and/or parent for the completion of the food records, including the description of all foods and beverages consumed, the serving size, the time of each meal or snack, and any supplements taken. Picture examples of serving sizes were also provided to help estimate serving size. For each of the 3 days, level of physical activity was self-reported from the list of five levels ranging from minimum (such as watching TV, reading, or computer work for most of the day) to high activity (such as walking fast, running, or playing sports for more than 90 min on a day). The food records included a section that asked about type, brand name and amount of supplements used each day. Upon the return of each food record, a member of the research team reviewed the information recorded with the participant and/or parent to obtain more details about items and portion sizes if needed.

Reported intake of food and beverages were entered into ESHA Research's Food Processor, version 10.6 (Salem, OR). The Health Canada Canadian Nutrient File database (Health Canada, 2015) was used as the primary source for food nutrient content. Manufacturer food labels were used for the nutrient content of supplements reported. The database was incomplete for some milk products (higher protein yogurts and alternative milk beverages) reported in the dietary records, and adjustments for the nutrient content of these products obtained from the United States Department of Agriculture (USDA) Nutrient database or manufacturer food labels were used.

2.4 | Energy and nutrient intake

Average daily energy, macronutrient and micronutrient intake was calculated from all food records for each participant. The micronutrients examined were vitamin A, vitamin B6, vitamin B12, vitamin D, folate, calcium, iron, magnesium, phosphorus, potassium, sodium, and zinc. These micronutrients were selected as they are the micronutrients with dietary intake levels below recommendations in greater than 5% of Canadian children and adolescents (Health Canada, 2012a, 2012b). Average daily energy and nutrient intake was assessed using age and gender appropriate dietary reference intakes (Institute of Medicine, 2006, 2011) including estimated energy requirement (EER), acceptable macronutrient distribution ranges (AMDR), estimated average requirement (EAR) or adequate intake (AI), and tolerable upper intake level (UL). Nutrient density was determined using the average of daily intake of nutrients per 1,000 kcal intake. Nutrient intake from supplements was not included in the comparison of nutrient intake relative to recommendations and the proportion meeting recommendations between groups. However, a separate analysis compared the proportion in the PWS group meeting nutrient recommendations with and without the inclusion of supplements.

2.5 | Food intake and healthy eating index

The first 3-day food record was used to estimate the intake of specific foods and diet quality in all participants. Food group classification and serving sizes were based on *Eating Well with Canada's Food Guide* (Health Canada, 2007). The number of servings from each of the four food groups were adjusted for energy intake (servings/1,000 kcal). The intake of "other foods" was also adjusted for energy intake (percent total kcal). "Other foods" were foods not within any of the food groups; these foods have low-nutrient density and are high in energy, fat, sodium, and/or added sugar such as desserts, sweets, potato chips, and soft drinks (Health Canada, 2007). Diet quality was assessed using an adaptation of the 2005 American Healthy Eating Index with recommendations from the Eating Well with Canada's Food Guide integrated (Garriguet, 2009). The index score was based on eight adequacy and three moderation components with a maximum score of 100 (Table 1). The adequacy components were scored based on meeting recommendations for servings of total vegetables and fruit, whole vegetables and fruit, dark green and orange vegetables, total grain products, whole grains, milk and alternatives, meat

TABLE 1 Scoring for components of the Healthy Eating Index—Canadian adaptation

Component	Range of score	Score criteria
Total vegetables and fruit	0–10	Maximum score: servings at or above recommendation[b] Minimum score: no servings
Whole vegetables and fruit	0–5	
Dark green and orange vegetables	0–5	
Total grain products	0–5	
Whole grain products	0–5	
Milk and alternatives	0–10	
Meat and alternatives	0–10	
Unsaturated fat intake	0–10	Maximum score: ≥18% of total energy Minimum score: 0% of total energy
Saturated fat intake	0–10	Maximum score: ≤ 7% of total energy Score of 8: 10% of total energy Minimum score: ≥15% of total energy
Sodium	0–10	Maximum score: ≤ AI[c] Score of 8: UL[d] Minimum score: ≥ twice the UL
Other foods	0–20	Maximum score: ≤5% of total energy Minimum score: ≥40% of total energy

[a] Proportional reduction in score if intake in between maximum and minimum intake.
[b] Age- and gender-specific serving recommendations in Healthy Eating with Canada's Food Guide (Health Canada, 2007).
[c] Age- and gender-specific AI in dietary reference intakes (Institute of Medicine, 2006).
[d] Age- and gender-specific UL in dietary reference intakes (Institute of Medicine, 2006).

and alternatives, and percentage of total energy from unsaturated fats. The three moderation components were scored based on limiting intake of saturated fats, sodium, and "other foods." Diet quality was classified as good diet quality (>80 points), diet that requires improvement (50–80 points), and poor diet (<50 points).

2.6 | Statistical analysis

Data were analyzed using the SPSS Statistics version 23.0 (IBM, Armonk, NY). Data were summarized as median and percentiles (25th, 75th). Mann–Whitney *U* test was used to assess differences between the PWS and control groups. Fisher's test was used to compare the proportion of participants meeting the nutrient intake recommendations between groups. Subanalysis within the PWS group was completed to explore the potential impact of genetic subtype and use of growth hormone (GH) treatment. Mann–Whitney tests (nonparametric) were used to assess differences in BMI, BMI *z*-score, energy intake and Healthy Eating Index score between those with deletion compared to uniparental disomy (UPD) and those receiving GH treatment compared to those not receiving GH treatment at the time of the study. Kruskal–Wallis tests were used to assess differences in BMI *z*-score between groups (either meeting, below, or above AMDR for fat). Statistical significance was at $p < .05$.

3 | RESULTS

3.1 | Participant characteristics

Twenty-three youth with PWS and 23 youth without PWS (control) were included in the study. The study participants included three PWS and control sibling pairs. The PWS group was composed of $n = 15$ youth with deletion subtype and $n = 8$ youth with UPD subtype.

Age, sex distribution, and weight *z*-score were not different between the PWS and control groups; however, height *z*-score was lower in the PWS group compared to the control group and there was a trend towards higher BMI *z*-score in the PWS group (Table 2). Within the PWS group, BMI *z*-score was not different between the participants with deletion subtype compared to UPD subtype as well as between participants receiving GH at the time of the study compared to those not receiving GH at the time of the study (data not shown).

3.2 | Food records

One 3-day food record was completed by 46% of participants ($n = 13$ PWS, $n = 8$ control), 24% completed two 3-day food records ($n = 2$ PWS, $n = 9$ control), and 30% completed three 3-day food records ($n = 8$ PWS, $n = 6$). The average distribution of percentage weekday: weekend days reported in the food records was 63:37 in the PWS group and 75:25 in the control group.

TABLE 2 Characteristics of youth with PWS and controls

Characteristic	PWS (*n* = 23)	Control (*n* = 23)	*p* value*
Age (years)	8.5 (6.1, 14.2)	11.9 (8.4, 14.7)	.127
Male:female (*n*)	8:15	14:9	.070
Weight *z*-score	0.3 (−0.5, 1.0)	0.6 (0.4, 1.1)	.184
Height *z*-score	−1.1 (−2.0, −0.1)	0.6 (−0.1, 1.0)	<.0005
BMI *z*-score	1.1 (0.3, 1.3)	0.4 (−0.1, 0.9)	.051

Notes. BMI, body mass index; PWS, Prader-Willi Syndrome. Data presented as median (25th and 75th percentiles). *z*-scores were calculated using EpiInfo (CDC, Atlanta, GA).
*Independent Mann–Whitney *U* test, statistical significance $p < .05$.

TABLE 3 Energy and macronutrient intake in youth with PWS and controls

	PWS (*n* = 23)	Control (*n* = 23)	*p* value*
Energy			
Intake (kcal/day)	1,523 (1,291, 1,763)	2,000 (1,726, 2,130)	<.001
Intake (kcal/day/cm)	11.3 (9.9, 13.5)	13.4 (11.0, 16.1)	.012
EER (kcal/day)	1,754 (1,298, 1,968)	2,106 (1,800, 2,524)	.003
Intake as %EER	91 (78, 109)	94 (85, 111)	.560
Carbohydrate Intake			
g/day	201 (170, 233)	250 (212, 276)	.001
Below EAR (*n*)	1	0	0.999
% kcal	58 (52, 60)	53 (50, 55)	.003
Below/above AMDR (*n*)	2/2	1/0	1.00/.489
Fiber, g/1,000 kcal	11.5 (9.8, 13.9)	9.3 (8.4, 10.7)	.005
Below AI (*n*)	18	22	.187
Protein Intake			
g/day	67 (55, 81)	88 (78, 100)	.002
g/kg/day	2.3 (1.5, 3.3)	2.0 (1.6, 2.9)	.645
Below EAR (*n*)	1	1	1.00
% kcal	19 (17, 20)	17 (17, 20)	.266
Below/above AMDR (*n*)	0/0	0/0	–
Fat Intake			
g/day	41 (38, 61)	69 (56, 83)	<.001
Total % kcal	27 (23, 30)	31 (28, 34)	.003
Below/above AMDR (*n*)	7/2	0/2	.015/1.00
Saturated % kcal	8.2 (6.5, 9.9)	10.8 (9.7, 12.4)	<.001
MUFA % kcal	8.4 (7.6, 11.2)	10.0 (6.0. 12.1)	.038
PUFA %kcal	4.5 (3.8, 6.8)	4.6 (3.9, 6.4)	.965

Notes. AI, adequate intake; AMDR, acceptable macronutrient distribution range; kcal, kilocalories; EAR,: estimated average requirement; EER, estimated energy requirements; PWS, Prader-Willi Syndrome. Energy and macronutrient intake is the average daily intake for each participant based on all dietary records obtained. dietary reference intakes (Institute of Medicine, 2006, 2011) for age and sex used for recommendations were EAR and AMDR. Data presented as median (25th and 75th percentiles).
*Independent Mann–Whitney *U* test or Fisher's test, statistical significance $p < .05$.

3.3 | Energy and macronutrient intake from food and beverages

Total estimated energy intake and EER were lower in the PWS group compared to control group; however, estimated energy intake relative to EER were similar (Table 3). Within the PWS group, energy intake, and EER were not different between those receiving GH treatment compared to those not receiving GH treatment. The lower intake of energy was due to a lower intake of all of the macronutrients (carbohydrates, protein, and fat) in the PWS group compared to controls. Macronutrient contribution to total energy intake was different between groups, with a higher percentage of energy from carbohydrates and lower percentage of energy from fat (total, saturated, and monounsaturated) in the PWS group compared to control group. The percent of energy from protein was similar between groups.

The intake of carbohydrates and protein (on a g/kg basis) was above the EAR for nearly all participants. Despite a higher intake of fiber per 1,000 kcal in the PWS group compared to control group, a similar proportion in each group were below the AI for fiber. Most participants had macronutrient intake within the AMDR. The proportion of participants that did not meet the AMDRs were similar between groups, except for a higher proportion in the PWS group who were below the AMDR for fat compared to the control group. Fat intake below AMDR was not associated with differences in BMI *z*-score within the PWS group.

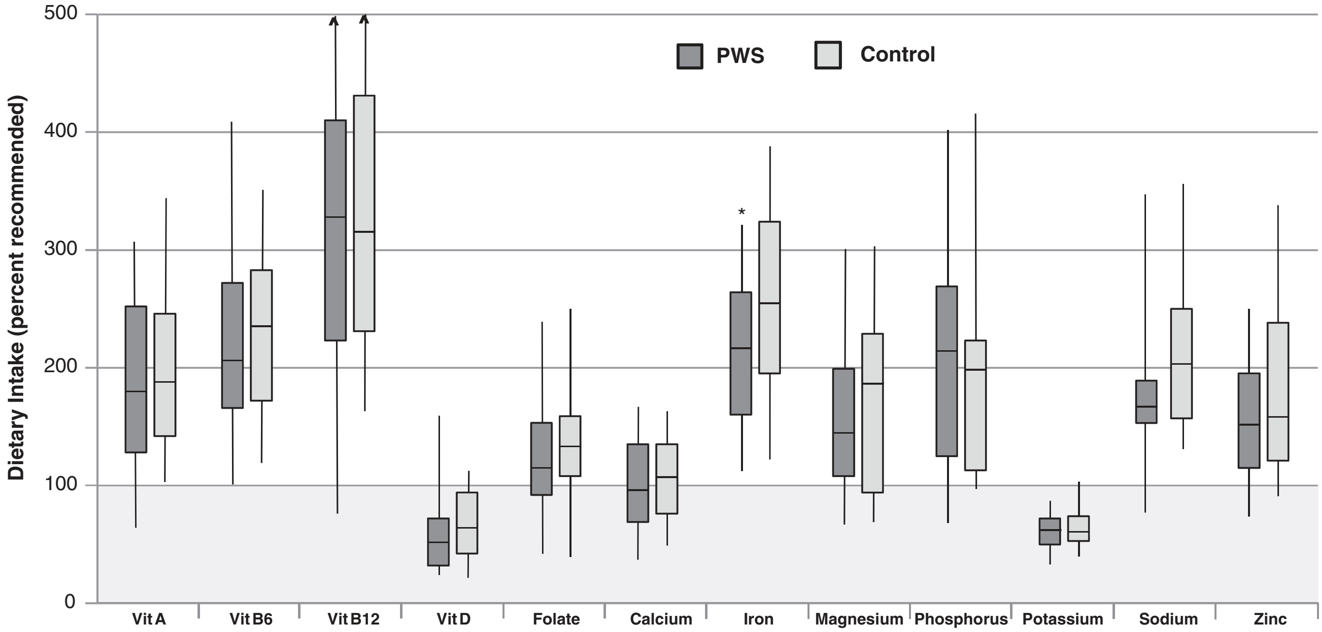
3.4 | Micronutrient intake from food and beverages

Median intake was above 100% of EAR or AI for most micronutrients assessed (Figure 1), except for vitamin D (both groups), calcium (PWS group), and potassium (both groups). Nearly all participants had intakes below EAR for vitamin D and AI for potassium. Half of the participants had intakes below the EAR for calcium. Iron intake relative to recommendation was lower in the PWS group compared to controls; however, no participants had intakes less than the EAR in either group. None of the participants had dietary intakes below the EAR for vitamin B6 and less than 11% total had intakes below recommendations for vitamin A, vitamin B12, phosphorus, sodium, and zinc. Most participants (*n* = 17 PWS, *n* = 18 control) had sodium intake above the UL, and one participant in the control group had zinc intake above the UL. The proportion of participants not meeting recommendations for the micronutrients was not different between groups.

3.5 | Supplement intake

Supplement use data were available for 76% of participants (*n* = 22 PWS, *n* = 13 control). Of these participants, 23% in the PWS group and 38% in the control group did not take any supplements. The most common supplement reported in both groups was a multivitamin/mineral (*n* = 15 PWS, *n* = 7 control). Other common single nutrient supplements reported were fish oil (*n* = 9 PWS, *n* = 2 control), calcium with vitamin D (*n* = 4 PWS), vitamin D (*n* = 1 PWS, *n* = 2 control), iron (*n* = 2 PWS), vitamin C (*n* = 1 PWS, *n* = 1 control), carnitine (*n* = 3 PWS), and coenzyme Q10 (*n* = 6 PWS). Use of fiber, magnesium, probiotic, N-acetyl cysteine, and vegetable/fruit extract supplements were also reported in the PWS group (*n* = 1 each).

The proportion of participants that meet nutrient recommendations with and without the inclusion of supplements was examined only in the PWS group (Table 4) because of the high number of missing information on supplement use in the control group. The inclusion of supplemental nutrient intake resulted in a lower number of PWS participants with an intake below the EAR for vitamin A, vitamin D, folate, calcium, and magnesium. The number below EAR or AI for vitamin B12, phosphorus, potassium, sodium, zinc, and fiber were not reduced with the inclusion of supplemental intake. The majority that remained below EAR or AI with the inclusion of supplemental intake did not report supplemental intake for the specific nutrients. However, some participants that remained below recommendations for vitamin D (*n* = 5), calcium (*n* = 2), and fiber (*n* = 1) did report the use of multivitamin/mineral supplement or single supplement containing the



n below EAR or AI	Vit A	Vit B6	Vit B12	Vit D	Folate	Calcium	Iron	Magnesium	Phosphorus	Potassium	Sodium	Zinc
PWS	4	0	1	22	7	12	0	5	4	23	2	2
Control	0	0	0	21	3	11	0	6	1	22	0	3

FIGURE 1 Micronutrient intake from food in youth with PWS (n = 23) and controls (n = 23). Micronutrient intake as a percent recommendation is presented as median, interquartile range, minimum, and maximum. Micronutrient intake is the average daily intake for each participant based on all dietary records obtained. dietary reference intakes (Institute of Medicine, 2006, 2011) used for recommendations were the Estimated Average Requirement (vitamins A, B6, B12, and D, folate, calcium, iron, magnesium, phosphorus, and zinc) or Adequate Intake (potassium and sodium) for age and sex. Table at bottom provides the number of participants in each group with intake below EAR or AI. ↑Maximum for vitamin B12: PWS 566%, control 731%. * Independent Mann–Whitney *U* test, p = .036. PWS: Prader-Willi Syndrome

TABLE 4 Number of youth with PWS not meeting nutrient recommendations with supplements excluded and included

	Intake excluding supplements PWS n = 22		Intake including supplements PWS n = 22	
	Number below EAR or AI	Number above UL	Number below EAR or AI	Number above UL
Vitamin A	4	0[a]	2	3[a]
Vitamin B6	0	0	0	1
Vitamin B12	1	No UL	1	No UL
Vitamin D	22	0	11	1
Folate	7	b	4	b
Calcium	12	0	8	0
Iron	0	0	0	0
Magnesium	5	No UL	3	No UL
Phosphorus	4	0	4	0
Potassium	22	No UL	22	No UL
Sodium	2	16	2	16
Zinc	2	0	2	3
Fiber	17	No UL	17	No UL

Notes. AI, adequate intake; EAR, estimated average requirements; PWS, Prader-Willi Syndrome; UL, tolerable upper intake level. Micronutrient intake with and without supplements is the average daily intake for each participant based on all dietary records obtained. Dietary reference intakes (Institute of Medicine, 2006, 2011) for age and sex used for recommendations were EAR (vitamins A, B6, B12, and D, folate, calcium, iron, magnesium, phosphorus, and zinc), AI (potassium and sodium), and UL.
[a] UL includes only preformed vitamin A.
[b] UL includes only synthetic forms of folate, separate intake for synthetic, and natural forms of folate not available.

specific nutrient. Supplement intake increased the number of participants in the PWS group with intake above the UL for vitamin A, vitamin B6, vitamin D, and zinc.

3.6 | Nutrient density of food and beverage intake

Nutrient density (amount of micronutrient per 1,000 kcal consumed) was higher in the PWS group compared to control for seven micronutrients: calcium, magnesium, phosphorus, potassium, vitamin C, pantothenic acid, and copper (Table 5).

3.7 | Food group intake

The number of servings per 1,000 kcal of vegetables and fruit, and meat and alternatives was higher in the PWS group compared to the control group (Table 6). Servings of grains and milk alternatives were similar between groups. The percent energy from the intake of foods that are high in energy, sugar, fat, and/or sodium (other foods) was also similar between groups.

All participants in the PWS group and 21 in the control group consumed less than the minimum servings recommended for at least one of the food groups. More participants with PWS were below the minimum recommended servings of grains compared to control group; a similar number were below the minimum recommended servings of vegetables and fruit, milk and alternatives, and meat and alternatives.

TABLE 5 Micronutrient density of food and beverage intake in youth with PWS and controls

	Nutrient density		
	PWS	Control	*p* value*
Vitamin A (μg RAE/1,000 kcal)	236 (160, 302)	366 (296, 486)	.410
Vitamin B1 (mg/1,000 kcal)	0.8 (0.8, 1.1)	0.8 (0.6, 0.8)	.082
Vitamin B2 (mg/1,000 kcal)	1.1 (1.0, 1.4)	1.0 (0.9, 1.2)	.086
Vitamin B3 (mg NE/1,000 kcal)	19 (16, 21)	19 (16, 22)	.991
Vitamin B6 (mg/1,000 kcal)	0.9 (0.8, 1.1)	0.8 (0.7, 1.0)	.082
Vitamin B12 (μg/1,000 kcal)	2.8 (2.0, 3.2)	2.1 (1.8, 3.0)	.111
Vitamin C (mg/1,000 kcal)	60 (40, 85)	38 (31, 49)	.004
Vitamin D (μg/1,000 kcal)	3.4 (2.4, 5.1)	3.1 (2.2, 4.6)	.277
Vitamin E (mg/1,000 kcal)	2.7 (1.8, 3.4)	2.2 (1.7, 2.7)	.203
Folate (μg DFE/1,000 kcal)	170 (142, 225)	159 (133, 188)	.277
Vitamin K (μg/1,000 kcal)	40 (23, 78)	31 (13, 64)	.277
Pantothenic (mg/1,000 kcal)	3.2 (2.8, 3.5)	2.8 (2.4, 3.0)	.019
Calcium (mg/1,000 kcal)	653 (484, 796)	496 (452, 574)	.038
Copper (mg/1,000 kcal)	0.7 (0.6, 0.7)	0.6 (0.5, 0.7)	.048
Iron (mg/1,000 kcal)	7.3 (6.7, 8.2)	7.2 (6.4, 7.9)	.475
Magnesium (mg/1,000 kcal)	169 (152, 179)	140 (135, 162)	.004
Manganese (mg/1,000 kcal)	1.6 (1.3, 1.8)	1.4 (1.2, 1.9)	.410
Phosphorus (mg/1,000 kcal)	837 (727, 905)	693 (606, 744)	.003
Potassium (g/1,000 kcal)	1.7 (1.6, 1.9)	1.3 (1.2, 1.5)	<.001
Selenium (μg/1,000 kcal)	52 (43, 63)	55 (45, 65)	.792
Sodium (g/1,000 kcal)	1.5 (1.2, 1.8)	1.4 (1.2, 1.6)	.489
Zinc (mg/1,000 kcal)	5.8 (4.7, 6.2)	4.8 (4.4, 5.6)	.124

Notes. PWS, Prader-Willi Syndrome. Nutrient density is the average daily nutrient density for each participant based on all dietary records obtained. Data presented as median (25th and 75th percentiles).
*Independent Mann–Whitney *U* test, statistical significance $p < .05$.

3.8 | Healthy eating index

Median total healthy eating index score for all participants was 69 out of a maximum score of 100, with an interquartile range of 59–74. Total healthy eating index score was higher in PWS group compared to the control group and a higher number in the PWS group had scores in the highest diet quality category (Figure 2). The PWS group had a higher median score for limiting the intake of saturated fat (9.1 vs. 6.4, $p < .001$) and lower score for meeting minimum servings of grain products (3.0 vs. 4.5, $p = .003$) compared to the control group. The groups were similar in scores for meeting the minimum servings of vegetables and fruit (total, whole, dark green, and orange), whole grains, milk and alternatives, and meat and alternatives as well as meeting recommendations for unsaturated fats, and limiting intake of sodium and foods not within the food groups (other foods).

4 | DISCUSSION

Dietary management is a central component in the prevention of excess weight gain in individuals with PWS, but restrictive diets can increase the risk of inadequate intake of essential nutrients. The purpose of our study was to estimate intake of essential nutrients from foods and diet quality in youth with PWS compared to those without PWS. Despite lower energy intake in the youth with PWS, dietary intake of most essential nutrients was similar and diet quality was higher compared to those without the syndrome.

Compared to the general population, energy needs are lower in individuals with PWS, attributed to a smaller amount of lean body mass relative to body weight and less energy expended doing physical activity (Butler, Theodoro, Bittel, & Donnelly, 2007; Davies & Joughin, 1993; Orsso et al., 2017; Reus et al., 2011; Schoeller, Levitsky, Bandini, Dietz, & Walczak, 1988; van Mil et al., 2000). The PWS group in the current study had lower energy intake and EER compared to the control group, with similar energy intakes relative to energy expenditure. However, it is difficult to assess the adequacy of energy intake as energy requirements have not been empirically determined for individuals with PWS. Recommendations for the population were used in this study to assess dietary intake, but it is important to recognize that it is not known if these recommendations are optimal for individuals with PWS, specifically in regards to promoting maintenance of a healthy body weight. Current estimations for energy needs of youth with PWS are 8–11 kcal/cm (Holm & Pipies, 1976; Scheimann, Lee, & Ellis, 2006; Schmidt, Pozza, Bonfig, Schwarz, & Dokoupil, 2008). The median absolute energy intake was 24% lower in the PWS group compared to the control group and corresponded to a median intake of 11 kcal/cm; thus, some of the youth in the PWS group may be at risk for excessive weight gain.

A reversed sex composition was present, with a higher number of males in the control group. In general, it is expected males would have a higher body weight and height than females with a more evident

TABLE 6 Daily food group intake in youth with PWS and controls

	PWS (*n* = 23)	Control (*n* = 23)	*p* value*
Vegetables and Fruit			
Servings/1,000 kcal	3.6 (2.7, 4.9)	2.4 (1.5, 3.2)	.006
Below minimum recommended servings (*n*)	12	18	.120
Grains			
Servings/1,000 kcal	2.2 (1.5, 2.7)	2.5 (1.9, 2.7)	.317
Below minimum recommended servings (*n*)	21	12	.007
Milk and Alternatives			
Servings/1,000 kcal	1.6 (1.2, 2.1)	1.2 (0.8, 1.7)	.645
Below minimum recommended servings (*n*)	11	13	.768
Meat and Alternatives			
Servings/1,000 kcal	1.2 (1.0, 1.5)	1.0 (0.9, 1.5)	.003
Below minimum recommended servings (*n*)	5	2	.414
Other Foods			
Percent total kcal	22 (11, 28)	27 (19, 31)	.085

Notes. PWS, Prader-Willi Syndrome. Food intake is the average daily intake for each participant based on the first 3 days of dietary records obtained. Food group designations, serving sizes, and recommended servings from *Eating Well with Canada's Food Guide* (Health Canada, 2007). Data presented as median (25th and 75th percentiles).
*Independent Mann–Whitney *U* test or Fisher's test, statistical significance $p < .05$.

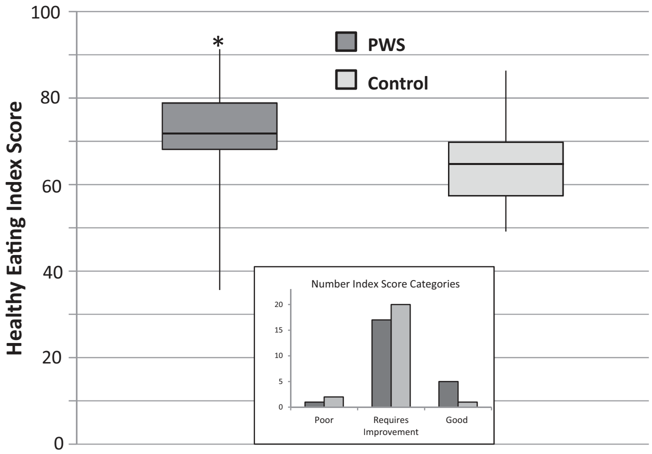


FIGURE 2 Healthy eating index score in youth with PWS (*n* = 23) and controls (*n* = 23). Healthy eating index score presented as median, interquartile range, minimum, and maximum. Insert is number of youth in each group within each score category. Healthy eating index was calculated using the average daily intake for each participant based on the first 3 days of dietary records. Scoring is based on Health Eating Index 2005—Canadian adaptation (Garriguet, 2009) with a higher score (maximum 100) indicating higher diet quality. Classification based on USDA classification for index score: *poor* (<50 points), *requires improvement* (50–80 points), and *good* (>80 points). *Independent Mann–Whitney *U* test, *p* = .007. PWS: Prader-Willi Syndrome

difference in the older youth. The impact of age and sex on the assessment of dietary intake was minimized with the use of intake relative to age- and sex-specific dietary reference intakes (Institute of Medicine, 2006, 2011), rather than absolute amounts. We also assessed weight, height and BMI using age and sex specific *z*-scores for comparison (EpiInfo, CDC, Atlanta, GA). Limitations to these, however, are the assumption of typical pubertal development for the age groups within the dietary reference intakes. In future studies, methods to estimate energy expenditure, such as doubly labeled water, and calculations based on measured lean body mass and calorimetry measured resting energy expenditure, could be used to provide a more accurate estimated energy expenditure. While pubertal stage would be expected to impact absolute nutrient intake, it is not clear if intake relative to recommendations or diet quality would be impacted.

The absolute intake of all three macronutrients was lower in the PWS group compared to the control group contributing to the lower estimated energy intake. Additionally, the macronutrient distribution of energy was different in the PWS group in that the proportion of energy from carbohydrates was higher and fat lower. Children with PWS who adhered to a diet with 40%–50% of energy from carbohydrates, 20%–30% from protein, and 25%–30% from fat had a lower BMI *z*-score than children with PWS who followed a diet with more energy from carbohydrates and less energy from fat and protein (Miller et al., 2013). A higher energy intake from protein, such as 20%–30% of energy, might be beneficial for individuals with PWS by supporting lean body mass gain and promoting satiety (Baum, Gray, & Binns, 2015). More research is needed to determine if a specific macronutrient distribution would be beneficial in individuals with PWS, particularly as it relates to body composition and hyperphagia.

Despite lower energy intake, estimated intake of essential nutrients relative to recommendations from food and beverages was similar in the PWS and control group with median intakes of most essential nutrients examined above the EAR or AI. The similarities in essential nutrient intake between the PWS and control group suggest that the PWS group consumed a more nutrient-dense diet. Nutrient density was higher in the PWS group for fiber and seven micronutrients. Higher energy-adjusted intake of foods from the vegetable and fruit, and meat and alternatives food groups likely contributed to the higher nutrient density. Our results are consistent with other studies that have also found a higher intake of vegetables and fruit in individuals with PWS (Nordstrom et al., 2015; Rubin et al., 2015). Diet quality was examined further in our study using a healthy eating index to evaluate dietary intake in relation to guidelines for healthy eating, including meeting recommendations for servings from the food groups and unsaturated fats, and minimizing the intake of saturated fats, sodium, and foods with low nutrient density (Garriguet, 2009). Youth with PWS had a higher healthy eating index score, and the median score in the PWS group of 72 was above the average healthy eating index score for Canadian children and adolescents in a national survey (score ranged from 54 to 65 in different age groups) (Garriguet, 2009). The study participants included three PWS and control sibling pairs. Siblings of an individual with PWS may consume a different diet compared to peers in families not living with an individual with PWS. However, because of the small number of participants included as sibling controls, this is unlikely to have impacted the end results about diet quality. Additionally, we would have been biased to find no differences between PWS and controls by inclusion of sibling controls; therefore, if anything, this strengthens our findings.

Although the PWS group had a better diet quality as indicated by higher dietary nutrient density and adherence to healthy eating guidelines, there were some dietary components that could be improved. Median percent of total energy from "other foods" was similar between groups (~25%). "Other foods" are foods that are low in nutrients but high in energy, salt, sugar and/or fat. Meeting nutrient requirements without excessive energy intake is difficult with a high intake of "other foods." Median fiber, vitamin D, and potassium dietary intakes were below recommendations in both groups, and median calcium intake was below recommendations in only the PWS group. National surveys of dietary intake (without including the contribution of supplements) in Canadian children and adolescents have also identified fiber, potassium, vitamin D, and calcium as nutrients with a high percentage of inadequate intake from food and beverages (Health Canada, 2012b; Health Canada, 2012a). Nearly all participants in the current study had sodium intake higher than the Tolerable Upper Level of Intake, similar to Canadian surveys for all age groups (Health Canada, 2012b, Health Canada, 2012a). Fiber, potassium, and sodium intake have not been assessed in youth with PWS in other studies, but inadequate dietary intake of vitamin D and calcium in children with PWS have been previously reported (Rubin et al., 2015).

Risk of inadequate nutrient intake was assessed in our study but nutrient status was not measured. Thus it is not known if nutrient deficiencies are present; however, low dietary nutrient intakes are a concern beyond just the presence of nutrient deficiency due to associations with risk of chronic disease. Low intakes of fiber and potassium and high intake of sodium are associated with increased risk of cardiovascular disease, diabetes, and/or cancer (Micha et al., 2017; Threapleton et al., 2013; World Cancer Research Fund, 2007). Adequate intake of calcium and vitamin D also contribute to the development of higher peak bone density during adolescence (Weaver et al., 2016).

Supplements may be needed to meet micronutrient recommendations in youth who are not able to meet recommendations through food and beverage intake alone. With the inclusion of supplement intake, the only youth in the PWS group with an intake of vitamin A, vitamin B12, folate, magnesium, and zinc below EAR were those who did not take a multivitamin/mineral. However, the amount of vitamin D (≤5 μg) and calcium (≤250 mg) in a few of the multivitamin/mineral supplements used was not sufficient to meet recommendations. Fiber and potassium intake may be best increased through increased intake of vegetables/fruits and whole grains as the amount of potassium in supplements is limited and these food sources are high in fiber. As children's dietary intake patterns are likely to track into adulthood, early efforts to improve food choices may have positive health benefits longer term into adult life.

The primary limitation of this study was the use of self-reported dietary intake. Currently, no feasible objective measure of usual dietary intake is available. As with other subjective measures of behavior, misreporting of dietary intake or a change in usual dietary behavior does occur (Schoeller, Bandini, & Dietz, 1990). Specific to our study, it is unknown if accuracies of dietary intake reporting is different between the PWS and control group. The method of dietary assessment used in this study and guidance provided for the completion of the records were based on minimizing inaccuracies. Food records were obtained from both weekdays and weekends to account for variation in dietary intake that typically occurs For example, diet quality has been reported as lower on weekends compared to weekdays in youth (Rothausen et al., 2012). The average distribution of weekdays and weekends reported in our study are close to the representative distribution of 71% weekdays and 29% weekend days (Thompson, Larkinab, & Brownab, 1986). While the weekday-weekend distributions were not matched between groups, higher diet quality was found in the PWS group despite the higher percentage of weekend dietary intake.

Our sample size was sufficient to detect differences between the groups and is the second highest sample size of published studies that examine nutrient intake in individuals with PWS (Lindmark et al., 2010; Nordstrom et al., 2015; Rubin et al., 2015). However, the sample size may have limited detection of smaller differences between groups.

5 | CONCLUSION

Our study characterized the dietary intake of essential nutrients and diet quality in youth with PWS compared to normally developing youth of similar age. Overall, the PWS group had indicators of a healthier intake of food and beverages compared to the control group including better adherence to healthy eating guidelines and higher energy adjusted intake of two foods groups, fiber and seven micronutrients. Dietary intake guidance should continue to emphasize the importance of selecting nutrient-dense foods and limiting foods with low nutrient-density in order to increase nutrient intake and maintain energy balance. Youth with PWS should have their dietary intake assessed to determine if micronutrient supplementation is warranted to reduce the risk of inadequate intake. Current studies in our group will assess whether fiber supplementation in PWS can induce changes in the gut microbiome profile, leading to improvements in hyperphagia and rate of weight gain in PWS. Prospective studies are also needed to determine PWS-specific recommendations for dietary intake that promotes a healthy body weight and adequacy of nutrient intake.

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CONFLICT OF INTEREST

All authors declare no conflict of interest.

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