

Peculiar body composition in patients with Prader-Labhart-Willi syndrome^{1,2}

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See corresponding editorial on page 1540.

ABSTRACT Prader-Labhart-Willi syndrome (PWS)—characterized by severe obesity, short stature, hypogonadism, and muscle hypotonia—appears to be an interesting model for body-composition abnormalities. Twenty-seven PWS patients (15 males and 12 females) aged 6–22 y underwent total-body analysis by dual-energy X-ray photon absorptiometry (DXA). For each PWS patient two age- and sex-matched control subjects were studied: one obese subject with a relative body weight (RBW > 120%) and body mass index (BMI) similar to that of the patient and one normal-weight subject (RBW < 120%). Percentage body fat was significantly greater in PWS patients than in obese subjects ($47.4 \pm 7.2\%$ compared with $41.9 \pm 9.9\%$, $P < 0.0001$) and the same difference was evident for arms and legs but not for the trunk. Lean mass was significantly lower in PWS patients (26.4 ± 8.2 kg) than in normal-weight subjects (32.9 ± 10.2 kg) and even more so than in obese subjects (40.3 ± 13.2 kg) ($P < 0.0001$). The most affected regions were limbs; thus, the ratio of lean mass in the trunk to that in the limbs was significantly higher in PWS patients (1.19 ± 0.15) than in obese (1.07 ± 0.13) and normal-weight (1.07 ± 0.09) subjects ($P < 0.002$). The ratio of fat mass to lean mass was significantly higher in PWS patients than in obese subjects (0.90 ± 0.32 and 0.74 ± 0.27 , $P < 0.05$). Bone mineral content (BMC) was significantly lower in PWS patients (1503 ± 46 g) than in normal-weight (1876 ± 677 g) and obese (2322 ± 773 g) subjects ($P < 0.0001$); this difference was most pronounced in the limb region. Bone mineral density (BMD) in PWS patients (0.993 ± 0.116 g/cm²) did not differ significantly from that of normal-weight subjects (1.033 ± 0.147 g/cm²) but was significantly lower than that of obese subjects (1.154 ± 0.139 g/cm²). The influence of age on body composition was assessed by comparing two age subgroups (< 12 y, $n = 10$; and ≥ 12 y, $n = 17$). The older PWS patients had higher adiposity, lower BMC, and dramatically lower BMD. Also, the lean mass deficit increased with age so that the ratio of fat mass to lean mass was close to 1. In conclusion, PWS patients showed a peculiar body composition, to some extent similar to that found in subjects deficient in growth hormone or even to sedentary and elderly people. These results suggest the importance of an accurate analysis of body composition in PWS patients. *Am J Clin Nutr* 1997;65:1369–74.

KEY WORDS Prader-Labhart-Willi syndrome, body composition, dual-energy X-ray photon absorptiometry, DXA, fat mass, lean mass, bone mineral content, bone mineral density, obesity, hypogonadism

INTRODUCTION

Prader-Labhart-Willi syndrome (PWS), generally sporadic in occurrence in the population at large, is characterized by severe obesity, mental deficiency, hypogonadism, hypotonia, short stature, small hands and feet, and a characteristic facial appearance (1, 2). The cause of this syndrome is not clear, although partial deletion of chromosome 15 or uniparental disomy has been found in virtually all cases (3, 4). Obesity usually develops in PWS patients after the first year of life and often becomes massive as patients age; patients have been shown to be > 200% of ideal body weight, a weight that is poorly controlled by dietetic treatment (5–10).

Hypothalamic and pituitary abnormalities have been postulated because of low growth hormone (GH) and insulin-like growth factor concentrations in PWS patients (11–13). This hormonal pattern closely resembles GH deficiency (14). Moreover, hypogonadism with low sex steroid concentrations has been documented (15). All these factors may influence the degree of obesity in such patients as well as body-composition abnormalities. Few data about body composition in PWS patients are available (12, 16–20); these studies, which used different techniques in relatively small samples ($n = 10$ –18 patients), suggest increased adiposity and decreased lean and water components. Dual-energy X-ray photon absorptiometry (DXA) allows direct and accurate measurement of three body components [fat mass, lean mass, and bone mineral content (BMC)], subdivided into three regions (arms, trunk, and legs) (21–24). This technique seems to be the best available to evaluate body composition, even in subjects who cooperate poorly, like PWS patients.

The aim of our study was to evaluate total and regional three-component body composition by DXA in PWS patients and to compare these values with those in both obese and normal-weight control subjects. The influence of age on body composition in PWS patients was also studied.

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SUBJECTS AND METHODS

Subjects

Twenty-seven PWS patients (15 males and 12 females) aged 6–22 y were studied. Their condition was diagnosed by using the fluorescent in situ hybridization technique (25) and methylation test for chromosome 15 deletion (26). For each PWS patient two age- and sex-matched control subjects were chosen from patients followed in our department: 1) normal-weight healthy control subjects with a relative body weight (RBW) 80–120% of ideal according to Tanner and Whitehouse (27), and 2) subjects with simple obesity with an RBW > 120% of ideal. None of the PWS patients were receiving estrogen or testosterone therapy nor treatment with human GH before or during the study.

Anthropometric characteristics of the study population are shown in **Table 1**. None of the obese children had a history of endocrine, nutritional, growth, or renal problems and subjects with other causes of secondary obesity were excluded. Normal-weight children were chosen from healthy children referred to our outpatient clinic for routine growth checks.

Pubertal stage was assessed on the basis of staging by Tanner and Whitehouse (27). All subjects were subdivided into two age groups: 10 children (< 12 y) and 17 adolescents or young adults (12–22 y). The cutoff age of 12 y was chosen because puberty begins in normal-weight and obese subjects at about this age, ie, all patients aged < 12 y were prepubertal and all patients aged ≥ 12 y were pubertal. PWS patients aged ≥ 12 y showed incomplete pubertal development or true clinical hypogonadism. The mean age of the younger group was 8.8 ± 2.3 y for PWS patients, 9.2 ± 2.5 y for obese subjects, and 9.6 ± 2.7 y for normal-weight subjects, whereas the mean age of the older group was 17.0 ± 3.7 y for PWS patients, 16.0 ± 2.5 y for obese subjects, and 16.1 ± 2.4 y for normal-weight subjects.

Informed consent was obtained from parents and children and ethical approval for the study was granted by the Hospital Ethical Committee. PWS patients were recruited with the assistance of the Italian Prader-Willi Association.

Methods

Weight was recorded with an electric scale while subjects wore minimal clothing. Height was measured to the nearest millimeter with a Harpenden stadiometer (British Indicators Ltd, London). Body mass index (BMI) was calculated as

weight (kg)/height² (m). RBW was calculated according to Tanner and Whitehouse (27) as related to ideal body weight for sex and height.

DXA is a scanning technique that measures the differential attenuation of two X-rays as they pass through the body. It distinguishes total-body BMC from soft tissue and subsequently divides the latter into fat and lean tissue. Therefore, body weight is the sum of fat mass, lean mass, and BMC (28). Total-body analysis with DXA (Lunar DPX-L, version 1.5 E; Lunar Corp, Madison, WI), equipped with child and adult software, was performed with subjects in the supine position; no sedation was required. Clinical and DXA analysis were performed on the same day before any dietary restriction in the obese subjects.

The entire body of each subject was scanned, beginning at the top of the head and moving in a rectilinear pattern down the body to the feet. PWS patients and obese subjects were scanned in the “slow” scan mode (scan speed of 38.4 mm/s with a sample size of 4.8 mm × 9.6 mm, a sample interval of 0.125 s, and source collimation of 1.68 mm); normal-weight subjects were scanned in the “medium” scan mode (scan speed of 76.8 mm/s with a sample size of 4.8 mm × 9.6 mm, a sample interval of 0.062 s, and source collimation of 1.68 mm) or in the “fast” scan mode (scan speed of 153.6 mm/s with a sample size of 4.8 mm × 9.6 mm, a sample interval of 0.03 s, and source collimation of 1.68 mm) according to their size. These technical specifications were indicated by the manufacturer. Mean measurement time was 20 min, analysis time was 15 min, and radiation exposure was < 8 μ Sv.

Body fat was expressed as both kg (fat mass) and as a percentage of body weight (%Fat); lean soft tissue mass was expressed both as kg and as g/cm ht, as suggested by Barlett et al (29), to minimize the influence of height differences among subjects. The distribution of fat and lean mass (in kg) was estimated by using the ratio of fat or lean mass in the trunk to that in the limbs (T:L Fat and T:L Lean, respectively). The relation between fat mass and lean mass (in kg) was estimated by the ratio of fat mass to lean mass (Fat:Lean). BMC was expressed both as total mass (in g) as well as the percentage of lean mass only (BMC %Lean). Bone mineral density (BMD) was expressed as g/cm².

Technical details of DXA were described previously (22, 23, 30). The body was subdivided into four regions: head, arms, trunk, and legs. A three-component analysis (fat mass, lean mass, and BMC) was performed in the arm, trunk, and leg regions. Total fat mass, lean mass, and BMC were the sum of regional data. Data concerning the head are not shown. The arms were defined by the line passing through the scapular-humeral joint. The trunk region was delineated by an upper horizontal border below the chin, vertical borders lateral to the ribs, and a lower border formed by oblique lines passing through the femoral necks. The leg region was defined as the tissue below the oblique lines passing through the femoral necks (31). The CV was 0.6% on phantom (plastic and water). The precision of our instrument was calculated as 0.7% for fat mass, 0.9% for lean mass, and 1.5% for total BMC of normal-weight adult subjects. Multiple scanning of children was not considered ethical. Daily quality-assurance tests were performed according to the manufacturer's direction. All scans were performed and analyzed by the same operator.

TABLE 1
Clinical characteristics of the study population¹

	PWS	Obese	Normal weight
Age (y)	14.2 ± 5.1	13.7 ± 4.1	13.7 ± 4.1
Weight (kg)	54.1 ± 19.1	73.0 ± 22.0 ²	45.9 ± 14.9
Height (cm)	139.8 ± 13.6 ³	155.3 ± 17.3	153.8 ± 17.4
RBW (%)	156 ± 33 ⁴	158 ± 30 ⁴	97 ± 10
BMI (kg/m ²)	27.1 ± 6.6 ⁴	29.7 ± 5.4 ⁴	18.7 ± 2.7

¹ $\bar{x} \pm$ SD; n 15 M, 12 F per group. RBW, relative body weight; PWS, Prader-Labhart-Willi syndrome.

^{2,3} Significantly different from other two groups (ANOVA, post hoc Scheffé *F* test): ² $P < 0.0001$, ³ $P < 0.001$.

⁴ Significantly different from normal weight, $P < 0.0001$ (ANOVA, post hoc Scheffé *F* test).

Statistical analysis

Differences between PWS patients and obese and normal-weight subjects were analyzed by using analysis of variance (ANOVA); group comparisons were done by using the post hoc Scheffé *F* test. Multiple-linear-regression analysis was used to assess the influence of anthropometric indexes on the lean component, assuming that total lean mass was the dependent variable (32); sex was introduced in the regression model as an independent dichotomous variable, with males assigned a value of 0 and females a value of 1. The significance level was chosen at 5%. Data were expressed as means $\pm$ SDs.

RESULTS

Clinical characteristics of PWS patients and control subjects are shown in Table 1. Mean RBW and BMI were not significantly different between PWS patients and obese subjects, whereas PWS patients were significantly shorter than normal-weight and obese subjects. Total and regional body composition of fat and lean components is shown in Table 2. Total adiposity was significantly higher in PWS patients than in obese and normal-weight subjects. Total as well as regional fat distributions were significantly higher in PWS patients ($P < 0.001$) than in normal-weight subjects. Moreover, arms %Fat and legs %Fat were significantly higher in PWS patients than in obese and normal-weight subjects ($P < 0.001$), whereas trunk %Fat was not significantly different between PWS patients and obese subjects. T:L Fat values were significantly higher in PWS patients and obese subjects than in normal-weight subjects.

Lean mass (in kg and in g/cm ht) was significantly lower in PWS patients than in normal-weight and obese subjects (Table 2). Lean mass in arms and legs of PWS patients was significantly lower than that in normal-weight and obese subjects, but not at the trunk level. Obese subjects had a significantly higher lean mass at trunk and legs than did normal-weight subjects.

TABLE 2

Total body and regional fat and lean compartments in the study population¹

	PWS	Obese	Normal weight
%Fat (%)	47.4 $\pm$ 7.2 ²	41.9 $\pm$ 9.9 ³	20.6 $\pm$ 9.4
Arms %Fat (%)	43.9 $\pm$ 8.7 ²	38.4 $\pm$ 11.9 ³	17.2 $\pm$ 8.6
Legs %Fat (%)	50.3 $\pm$ 7.8 ²	43.7 $\pm$ 11.3 ³	25.5 $\pm$ 10.6
Trunk %Fat (%)	44.7 $\pm$ 9.9 ³	44.0 $\pm$ 16.0 ³	17.2 $\pm$ 9.2
T:L Fat	0.99 $\pm$ 0.36 ³	0.96 $\pm$ 0.24 ³	0.68 $\pm$ 0.19
Lean mass (kg)	26.4 $\pm$ 8.2 ²	40.3 $\pm$ 13.2	32.9 $\pm$ 10.2
Lean mass (g/cm ht)	184.9 $\pm$ 43.5 ²	251.0 $\pm$ 58.4 ³	210.5 $\pm$ 45.0
Arms Lean mass (kg)	2.1 $\pm$ 0.7 ²	3.7 $\pm$ 1.5	3.1 $\pm$ 1.2
Legs Lean mass (kg)	9.0 $\pm$ 3.0 ²	14.5 $\pm$ 4.9 ³	11.5 $\pm$ 3.9
Trunk Lean mass (kg)	13.2 $\pm$ 4.4	19.2 $\pm$ 6.9 ²	15.4 $\pm$ 4.8
T:L Lean	1.19 $\pm$ 0.15 ²	1.06 $\pm$ 0.13	1.07 $\pm$ 0.09

¹ $\bar{x} \pm$ SD; $n = 15$ M, 12 F per group. T:L, trunk–limbs ratio; PWS, Prader-Labhart-Willi syndrome; % Fat, body fat as a percentage of body weight.

² Significantly different from other two groups, $P < 0.001$ (ANOVA, post hoc Scheffé *F* test).

³ Significantly different from normal weight, $P < 0.0001$ (ANOVA, post hoc Scheffé *F* test).

Therefore, T:L Lean was significantly higher in PWS patients than in obese and normal-weight subjects.

Multiple-regression analysis with total lean mass as the dependent variable confirmed a significant difference between PWS patients and normal-weight subjects; the influencing variables were weight, height, and sex, as shown in Table 3.

When lean mass was related to fat mass (Fat:Lean), PWS patients had a significantly higher ratio (0.90 ± 0.32) than obese (0.74 ± 0.27) and normal-weight (0.26 ± 0.16) subjects (Figure 1). Moreover, eight PWS patients had a Fat:Lean value > 1 , ie, the fat component was greater than the lean mass component.

PWS patients had a significantly lower total BMC than normal-weight and obese subjects (Table 4). PWS patients also had a significantly lower arms BMC and legs BMC than the other two groups. On the contrary, obese subjects had a significantly greater total BMC value than the normal-weight group and a significantly greater legs and trunk BMC than the PWS and normal-weight groups. There were no significant differences in BMC %Lean among the three groups. BMD was similar in PWS patients and normal-weight subjects; however, BMD in obese subjects was significantly higher than that in PWS patients and normal-weight subjects.

Age influence on anthropometric and body-composition indexes are shown in Tables 5 and 6. Weight and height were not significantly different between PWS children and their normal-weight peers although BMI and %Fat were significantly higher in PWS patients than in normal-weight subjects, but were similar to values for the obese subjects. PWS adolescents and young adults had higher weights and lower heights than normal-weight and obese subjects ($P < 0.01$). BMI was similar between PWS patients and obese subjects, whereas %Fat was significantly higher in PWS patients than in normal-weight subjects ($P < 0.01$). PWS children had less lean mass than older PWS patients: 86% compared with 78% of normal values for total lean mass, 81% compared with 63% for arms, 84% compared with 77% for legs, and 93% compared with 83% for trunk. The distribution of lean mass, however, did not change significantly with age.

Fat:Lean values in PWS children and obese subjects were not significantly different, whereas older PWS patients had a Fat:Lean value close to 1, significantly higher than that of their obese peers (Figure 1). BMC and BMD were not significantly different between PWS children and normal-weight subjects; however, BMC and BMD were significantly lower in PWS

TABLE 3

Results of multiple-regression analysis of the influence of several anthropometric variables on lean mass in patients with Prader-Labhart-Willi syndrome (PWS)

Variable	β coefficient ¹	SE	<i>P</i>
Sex	3126.6	889.4	0.001
Age	42.9	183.4	0.81
Height	333.2	47.6	0.0001
Weight	246.1	50.1	0.0001
PWS patients compared with normal-weight subjects	-4303.9	1221.0	0.001

¹ Represents the difference expressed in grams between patients with PWS and normal-weight subjects.

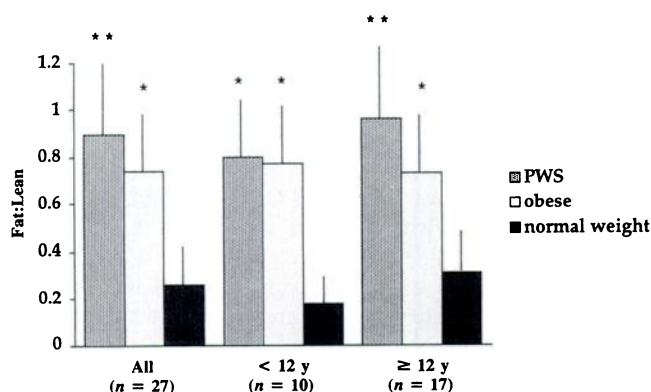


FIGURE 1. The ratio of fat mass to lean mass (in kg) in the study population. $\bar{x} \pm SD$. *Significantly different from normal weight, $P < 0.05$ (ANOVA); **significantly different from obese, $P < 0.05$ (ANOVA).

adolescents than in normal-weight and obese subjects (Table 6).

DISCUSSION

Most PWS patients are obese but many of their clinical features suggest that the pathogenesis of their excess weight as well as their body composition could be different from what is observed in simple obesity. To verify this we carefully selected our control population: each PWS patient was matched for age and sex with a normal-weight control subject and also with an obese control subject with a comparable degree of obesity (similar RBWs and BMIs). BMI was chosen instead of body weight to adjust for height differences between PWS patients and obese subjects.

Few data are available about body composition in PWS patients (12, 16–20) and the sample sizes in these previous studies were small relative to the sample size of our PWS population, which was large considering the low prevalence of the disease; moreover, the wide age range of the patients in our study was sufficient to permit the evaluation of its influence on body components. Our data show that PWS patients were fatter than obese subjects even when the groups had similar ponderal indexes.

The comparison between young and older PWS patients showed that differences in excessive weight and short stature became significant only among teenagers, as described elsewhere (8, 9). However, a longitudinal survey of such patients is necessary to verify this trend.

Fat distribution was similar in obese subjects and PWS patients and both groups also had similar fat depots. T:L Fat was higher in PWS patients and obese subjects than in normal-weight control children, in whom leg adiposity is prevalent; however, PWS patients had significantly higher adiposity in the limbs than did obese subjects.

Lean mass was significantly lower in PWS patients than in normal-weight control subjects and even lower than that in obese subjects. The muscles in the limbs were the most compromised region as shown by the higher T:L Lean value in PWS patients than in normal-weight subjects. DXA is not able to discriminate between muscles and viscera when used to measure lean mass. Nevertheless, our findings suggest that muscle mass was probably more impaired than viscera. This hypothesis is consistent with the moderate clinical hypotonia described previously in PWS patients (2). Multiple-regression analysis with lean mass as the dependent variable showed that lean mass was strongly influenced by weight, height, and sex, but also by the syndrome itself.

The index that best describes the body composition in PWS patients seems to be Fat:Lean; in fact, it represents adequately both the increase of fat mass as well as the reduction of lean mass. In our normal-weight control subjects lean mass was four times the amount of fat, whereas in obese subjects the Fat:Lean value was 3:4. On the contrary, PWS patients had a similar proportion of lean and fat mass, with a mean ratio close to 1. In some PWS patients we observed an inversion of this ratio up to 1.9, which is very unusual for obese subjects.

In the comparison between young and older PWS patients we found a more marked deficit in the lean component in the older subjects (from 81% to 93% of normal values in the younger, from 63% to 83% of normal values in the older PWS patients, depending on the region analyzed). A deficit in lean mass was most evident in the limbs of PWS patients, even in older patients, as shown by a T:L Lean value similar to that of PWS children. The progressive increase of adiposity to-

TABLE 4

Total and regional bone mineral content (BMC) and bone mineral density (BMD) in the study population¹

	PWS	Obese	Normal weight
BMC (g)	1503 $\pm$ 46 ²	2322 $\pm$ 773 ³	1876 $\pm$ 677
BMD (g/cm ²)	0.993 $\pm$ 0.116	1.154 $\pm$ 0.139 ²	1.033 $\pm$ 0.147
Arms BMC (g)	125 $\pm$ 44 ²	231 $\pm$ 112	205 $\pm$ 87
Legs BMC (g)	544 $\pm$ 213 ²	963 $\pm$ 373 ²	715 $\pm$ 294
Trunk BMC (g)	440 $\pm$ 157	692 $\pm$ 268 ²	560 $\pm$ 241
BMC %Lean (%) ⁴	5.8 $\pm$ 0.9	5.8 $\pm$ 0.7	5.6 $\pm$ 0.7
Arms BMC %Lean (%)	6.0 $\pm$ 1.0	6.1 $\pm$ 1.1	6.6 $\pm$ 1.2
Legs BMC %Lean (%)	5.9 $\pm$ 0.9	6.5 $\pm$ 0.8	6.1 $\pm$ 1.1
Trunk BMC %Lean (%)	3.4 $\pm$ 0.8	3.6 $\pm$ 0.9	3.5 $\pm$ 0.6

¹ $\bar{x} \pm SD$; n = 15 M, 12 F per group. PWS, Prader-Labhart-Willi syndrome.

² Significantly different from other two groups, $P < 0.001$ (ANOVA, post hoc Scheffé F test).

³ Significantly different from normal weight, $P < 0.0001$ (ANOVA, post hoc Scheffé F test).

⁴ BMC %Lean = BMC (g)/Lean mass (g) $\times$ 100.

TABLE 5Differences in body composition in the study population aged <12 y¹

	PWS	Obese	Normal weight
Height (cm)	127.4 ± 14.9	137.1 ± 12.7	137.4 ± 12.9
Weight (kg)	40.7 ± 15.1	52.4 ± 15.7 ²	31.3 ± 8.1
BMI (kg/m ²)	24.2 ± 4.9 ³	27.4 ± 4.5 ³	16.2 ± 1.8
%Fat (%)	46.7 ± 7.2 ³	42.6 ± 10.9 ³	16.0 ± 6.2
Lean mass (kg)	20.1 ± 6.7 ⁴	27.8 ± 5.2	24.2 ± 5.4
T:L Lean	1.21 ± 0.16 ⁵	1.04 ± 0.08	1.07 ± 0.09
BMC (g)	1115 ± 392	1514 ± 380	1235 ± 320
BMD (g/cm ²)	0.92 ± 0.07	1.00 ± 0.07	0.90 ± 0.11

¹ $\bar{x} \pm SD$. $n = 10$ per group. PWS, Prader-Labhart-Willi syndrome; T:L, trunk-limbs ratio; BMC, bone mineral content; BMD, bone mineral density.

²⁻⁴ Significantly different from normal weight (ANOVA, post hoc Scheffé F test): ² $P < 0.01$, ³ $P < 0.0001$, ⁴ $P < 0.05$.

⁵ Significantly different from obese and normal weight, $P < 0.01$ (ANOVA, post hoc Scheffé F test).

gether with a deficit in lean mass with age in PWS patients yielded a Fat:Lean value close to 1 in the older PWS patients.

BMC in PWS patients was significantly lower than that in normal-weight subjects, whereas BMD was lower but not significantly so, probably because of the limited number of subjects in our study; BMC %Lean also was lower at the limbs level. This finding confirms our data about a direct influence of lean mass on BMC in children (33).

In the comparison of age groups, a deficit in BMC was evident only in the older PWS group, and it was associated with a significantly lower BMD than in normal-weight subjects. This finding suggests the importance of sex hormones on lean mass, bone mass, and bone metabolism (34).

PWS patients showed a pattern of body composition similar to that in GH-deficient subjects (increased fat mass and reduced lean and bone mass, especially in the limbs). This finding is very intriguing because studies of GH secretion in PWS patients suggest an impairment of hypothalamic regulation of GH secretion (11–13). We speculate that GH deficiency, as well as pubertal delay, could be responsible for the abnormal body composition. Some reports on body composition changes in PWS patients during biosynthetic GH treatment support this hypothesis (12, 16).

A reduced level of physical activity has been reported in PWS patients, even in children (20), associated with muscle deficiency and weakness, mental retardation, severe obesity, and cardiorespiratory impairment. It has been documented that regular exercise reduces fat mass and improves lean and bone components (35, 36); the body composition of PWS patients, on the contrary, resembles that of sedentary individuals.

A similar pattern of body composition (37, 38) has been described in elderly people, in whom many of the factors described previously were indicated (ie, GH deficiency, reduced physical activity, and reduced sex hormone concentrations).

In conclusion, our data on PWS children and adolescents document a peculiar body composition, characterized by 1) increased adiposity, even compared with simply obese subjects with the same BMI; fat deposition is prevalent at the limbs and increases with age; 2) reduced lean mass, more marked at the limbs and more evident in older subjects; 3) a Fat:Lean value

TABLE 6Differences in body composition in the study population aged ≥12 y¹

	PWS	Obese	Normal weight
Height (cm)	145.9 ± 6.7 ²	164.4 ± 10.9	162.6 ± 10.5
Weight (kg)	60.7 ± 17.6 ²	83.3 ± 16.9 ⁴	52.9 ± 9.9
BMI (kg/m ²)	28.5 ± 7.0 ⁴	30.8 ± 5.5 ⁴	19.9 ± 2.2
%Fat (%)	47.7 ± 7.4 ⁴	41.6 ± 9.7 ⁴	23.4 ± 10.1
Lean mass (kg) ⁵	29.5 ± 7.0	46.5 ± 11.5	38.0 ± 8.8
T:L Lean	1.18 ± 0.14 ²	1.07 ± 0.15	1.07 ± 0.10
BMC (g) ⁵	1698 ± 366	2726 ± 573	2253 ± 527
BMD (g/cm ²) ⁵	1.03 ± 0.12	1.23 ± 0.09	1.11 ± 0.11

¹ $\bar{x} \pm SD$; $n = 17$ per group. PWS, Prader-Labhart-Willi syndrome; T:L = trunk-limbs ratio; BMC, bone mineral content; BMD, bone mineral density.

^{2,3} Significantly different from other two groups (ANOVA, post hoc Scheffé F test): ² $P < 0.001$, ³ $P < 0.0001$.

⁴ Significantly different from normal weight, $P < 0.0001$ (ANOVA, post hoc Scheffé F test).

⁵ Significant difference among all groups, $P < 0.0001$ (ANOVA, post hoc Scheffé F test).

close to 1 in most of the older patients; and 4) reduced BMC especially at the limbs and in the older PWS patients.

The typical pattern of body composition found in PWS patients is probably due to the syndrome itself but the similarity with other conditions suggests a multifactorial pathogenesis. Moreover, our data suggest that anthropometric indexes alone may fail to define fat excess, especially in younger PWS patients, if not supported by an accurate body-composition analysis. 

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