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ORIGINAL ARTICLE

Distortion of body image perception in the Prader-Willi syndrome: Relationship with the perceptual reasoning index

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Self-perception;
Intelligence quotient;
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Abstract
Introduction: Self-perception of body image has been scarcely evaluated in people with Prader-Willi syndrome (PWS), who, in addition to intellectual disability, are often obese. Therefore, we explored whether people with PWS can accurately identify their true image and how this self-perception is impacted by their neuropsychological profile.
Methodology: This observational study included patients with PWS with regular attendance to transdisciplinary treatment at a center specialized in the management of rare diseases. All patients were evaluated with the Stunkard scale (including silhouettes ranging from extremely skinny to extremely obese) and the WISC-IV and WAIS-III questionnaires, specifically the perceptual reasoning index (PRI).
Results: Among the 21 participants, 62% misperceived their body image, most underestimating their body dimensions (actual BMI $28.0 \pm 8.3\,kg/m^2$ vs self-perceived BMI $23.2 \pm 4.7\,kg/m^2$, $p = 0.03$). While BMI differences between accurate and inaccurate body image perception were nonsignificant (accurate $26.6 \pm 8.8\,kg/m^2$ vs inaccurate $28.9 \pm 8.1\,kg/m^2$, $p = 0.56$), individuals with accurate perception showed both higher PRI scores (accurate 67.6 ± 8.2 vs inaccurate 60.2 ± 7.2, $p = 0.043$) and, to a lesser extent, intelligence quotients (IQ) (accurate 60.0 ± 4.9 vs inaccurate 55.0 ± 7.5, $p = 0.079$).

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Conclusion: In this study, we identified distortion of body image perception as a very common finding among PWS patients, in most cases as underestimation, and influenced by the neuropsychological profile.

PALABRAS CLAVE
Autopercepción;
Cociente intelectual;
Estado nutricional;
Obesidad;
Escala Stunkard

Distorsión de la percepción de la imagen corporal en el síndrome de Prader-Willi: relación con el índice de razonamiento perceptivo

Resumen
Introducción: La autopercepción de la imagen corporal ha sido escasamente evaluada en personas con síndrome de Prader-Willi (SPW), quienes, además de discapacidad intelectual, suelen presentar obesidad. Por este motivo, en este estudio exploramos la capacidad de las personas con SPW de identificar con precisión su verdadera imagen corporal y cómo esta autopercepción se ve influida por su perfil neuropsicológico.
Metodología: Este estudio observacional incluyó pacientes con SPW con asistencia regular a tratamiento transdisciplinario en una institución especializada en enfermedades raras. Todos los pacientes fueron evaluados con la Escala Stunkard (que comprende siluetas que van desde extremadamente delgadas hasta extremadamente obesas) y los cuestionarios WISC-IV y WAIS-III, específicamente el índice de razonamiento perceptivo (IRP).
Resultados: Entre los 21 participantes, el 62% percibió erróneamente su imagen corporal, y la mayoría subestimó sus dimensiones corporales (IMC real $28{,}0 \pm 8{,}3\,kg/m^2$ vs. IMC percibido $23{,}2 \pm 4{,}7\,kg/m^2$, $p = 0{,}03$). Si bien las diferencias en el IMC entre la percepción correcta e incorrecta de la imagen corporal no fueron significativas (correcta $26{,}6 \pm 8{,}8\,kg/m^2$ vs. incorrecta $28{,}9 \pm 8{,}1\,kg/m^2$, $p = 0{,}56$), los individuos con percepción correcta mostraron puntuaciones más altas de IRP (correcta $67{,}6 \pm 8{,}2$ vs. incorrecta $60{,}2 \pm 7{,}2$, $p = 0{,}043$) y, en menor medida, de cociente intelectual (CI) (correcta $60{,}0 \pm 4{,}9$ vs. incorrecta $55{,}0 \pm 7{,}5$, $p = 0{,}079$).
Conclusión: En este estudio, identificamos la distorsión de la percepción de la imagen corporal como un hallazgo muy común entre los pacientes con SPW, en la mayoría de los casos como subestimación, e influido por el perfil neuropsicológico.

Introduction

Prader-Willi syndrome (PSW) is the most frequent cause of obesity of genetic origin, which in many cases is severe and associated with multiple comorbidities.[1] PWS is also characterized by cognitive, mental and behavioural symptoms.[2] The most common pathopsychological signs include intellectual disability, obsessions, impulsivity, autistic-like behaviours, and self-harm.[3]

For people with obesity and other eating disorders, the problems associated with impaired body image and body dissatisfaction are well established.[4] However, self-perception of body image has not been widely evaluated in people with intellectual disabilities or in those with a genetic marker directly associated with body weight such as PWS.[1] It is estimated that people with intellectual disabilities are more likely to develop negative self-concepts and low self-esteem due to their perception of intellectual insufficiency, general failures in academic and social domains, and stigmatization.[5] In line with this, Napolitano et al. reported elevated levels of body dissatisfaction in individuals with PWS.[1] However, as far as we know, it is unknown whether these individuals are able to accurately self-perceive their body image, being this possibly relevant given the comorbidities associated with obesity.

Therefore, the objective of this study was to evaluate the relationship between body self-perception and indicators of cognitive ability, specifically the perceptual reasoning index.

Methods

Study population

This prospective observational study included subjects of both sexes with a confirmed genetic diagnosis of PWS and regular attendance at a center specialized in the management of rare diseases. All included patients received transdisciplinary treatment tailored to their individual needs, with sessions occurring weekly, biweekly, or monthly, as determined by the treating multidisciplinary team. The treatment involved the following disciplines: nutrition,

psychology, educational psychology, occupational therapy, speech therapy, kinesiology, clinical medicine, and psychiatry.

Instruments

Body image perception was evaluated through the Stunkard scale, and validated by Stunkard, Sørensen, and Schulsinger.[6] The Stunkard figure rating scale is validated in children and adults, and consists of 9 images (male and female silhouettes), ranging from extremely skinny (silhouette 1) to extremely obese (silhouette 9).[7]

The body mass index (kg/m^2) represents the most practical anthropometric method, keeping a close relationship with the degree of adiposity. Its classification allows establishing the nutritional status of each patient, as follows: BMI <18.5 kg/m^2, low weight; 18.5 up to 24.9 kg/m^2, normal weight; 25 up to 29.9 kg/m^2, overweight; 30 up to 34.9 kg/m^2, grade I obesity; 35 up to 39.9 kg/m^2, grade II obesity; >40 kg/m^2, grade III obesity. Regarding pediatric individuals, the World Health Organization (WHO) BMI-for-age charts and their corresponding *Z*-score were used to establish their nutritional status.[8]

The silhouettes displayed on the Stunkard scale have been previously associated with the BMI, relating figure 1 as low weight, figures 2–4 as normal weight, figure 5 as overweight, and figure 6 or higher as obesity.[9,10]

The BMI and corresponding nutritional status of each patient were recorded and adjudicated to a specific silhouette of the Stunkard scale. Thereafter, without knowledge of such adjudication, the patient was asked to identify the silhouette more closely related to their self-perception. The answers were, then, classified as concordant or discordant, according to the agreement or disagreement with their actual BMI.

Intelligence indices were assessed using the Wechsler Intelligence Scale for Children (WISC-IV) and Adults (WAIS-III), which assesses verbal and non-verbal intelligence.[11,12] The first (WISC-IV) is used in children from 6 years and 0 months up to 16 years and 11 months, while the adult version (WAIS-III) can be used in people from 16 and 0 months up to 90 years and 11 months. We evaluated the total intelligence quotient (TIQ) and the perceptual reasoning index (PRI). Such index includes fluid reasoning (manifested in tasks that require handling abstract concepts, rules, generalizations, and logical relationships, especially on new material), spatial processing, and visualmotor integration.[3] Within the Wechsler scales, raw results were transformed into scalar scores, being the mean of each administered subtest 10 points and the standard deviation, 3. Based on these scores, the composite scores of the TIQ are constructed.[13]

In the case of qualitative intelligence classifications, the classification given by the Wechsler scales was used and categorized between ''very low'' (≤69 IQ), ''inferior'' or borderline (IQ 70 up to 79), ''normal-low'' (80 up to 89 IQ), ''average'' (90 up to 109 IQ), ''normal-high'' (110 up to 119 IQ), ''high'' (120 to 129 IQ) and ''very high'' (≥IQ 130).[11,12]

If the descriptive classification obtained was ''very low'', the degree of mental impairment was determined, taking into account the Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition (DSM-5): mild mental impairment (50 up to 69 IQ), moderate mental impairment (36 up to 49 IQ), severe mental impairment (35 to 20 IQ) and deep mental impairment (<20 IQ).[14]

Procedure

Data mining was performed through individual consultation with each patient by different health professionals with experience in nutrition and mental impairment according to the administered scale.

The administration of the Stunkard scale took approximately 15 min, while considering that patient's performance could vary in relation to attention and concentration due to environmental causes, mood, and fatigue. This scale, administered to the patients within the nutrition space by the related area health professional, was presented to the patients through a digital device to improve its visualization. The Stunkard scale consisted of asking the subjects which figure they identified with, according to their perception, and was compared with their actual BMI.

The administration of the Wechsler scales required several sessions (approximately 4 sessions lasting 30 to 40 minutes each) conducted in the center's psychopedagogy ward.

This study was approved by the local ethical committee CEIC-Stamboulian (Project No.: 10223) in full compliance with the ethical standards set forth in the Declaration of Helsinki of 1964 and its additional amendments. All patients/families gave their prior written signed informed consent for the anonymous use of their data (habeas data).

Statistical data

Continuous variables are expressed as means ± standard deviation or medians (interquartile range) in the case of non-uniform distribution, and the categorical ones as numbers and percentages. Comparisons between groups were evaluated using paired and independent-samples Student tests. Analyses were performed using SPSS software, version 22.0 (IBM SPSS Statistics for Windows, Armonk, New York, NY, United States).

Results

A total of 21 patients with a confirmed diagnosis of PWS were included (Table 1), with a mean age of 24.9 ± 10.0 years. All had cognitive deficits, with a very low IQ (56.9 ± 6.9), while in the classification of mental deficiency, 19 (91%) individuals had mild mental impairment.

Regarding nutritional status, 9 (43%) had normal weight, 6 (29%) were overweight, 4 (19%) had grade 1 obesity, and 6 (10%) grade 2 obesity.

According to the BMI and the Stunkard scale, 13 (62%) patients had an inaccurate perception of their body image (Fig. 1), with a significant underestimation of their figure (actual BMI 28.0 ± 8.3 kg/m^2 vs self-perceived BMI 23.2 ± 4.7 kg/m^2, $p = 0.03$).

We did not identify any significant differences regarding the BMI between patients with accurate or inaccurate body image perception (accurate 26.6 ± 8.8 kg/m^2 vs inaccurate

Table 1 Group characteristics.

Patient	Age	Sex	Weight (kg)	Height (m)	BMI	PRI	IQ
#1	15	M	43.3	1.33	24.40	72	69
#2	20	F	47.1	1.48	21.50	58	56
#3	19	F	50.4	1.46	23.64	71	58
#4	35	M	66	1.51	29.00	54	45
#5	38	M	55.8	1.52	24.00	84	69
#6	44	M	81.2	1.57	32.94	70	64
#7	22	M	50.5	1.47	23.30	62	56
#8	31	M	58.2	1.51	25.65	69	58
#9	19	M	49.6	1.48	22.64	56	62
#10	30	M	58.2	1.59	23.02	65	57
#11	26	M	66.9	1.59	26.46	56	53
#12	14	M	36	1.44	17.36	57	57
#13	23	F	56	1.42	27.70	58	50
#14	9	F	22.9	1.16	17.10	66	63
#15	26	M	75.5	1.55	31.22	62	53
#16	21	F	51.6	1.46	24.17	67	62
#17	39	M	85.7	1.30	50.32	64	54
#18	40	M	67.3	1.50	29.90	64	53
#19	23	F	91.3	1.40	46.58	69	61
#20	17	F	68.5	1.44	33.17	47	41
#21	11	M	72.8	1.47	33.92	53	54

BMI: body mass index; PRI: perceptual reasoning index; IQ: intelligence quotient; F: female; M: male.

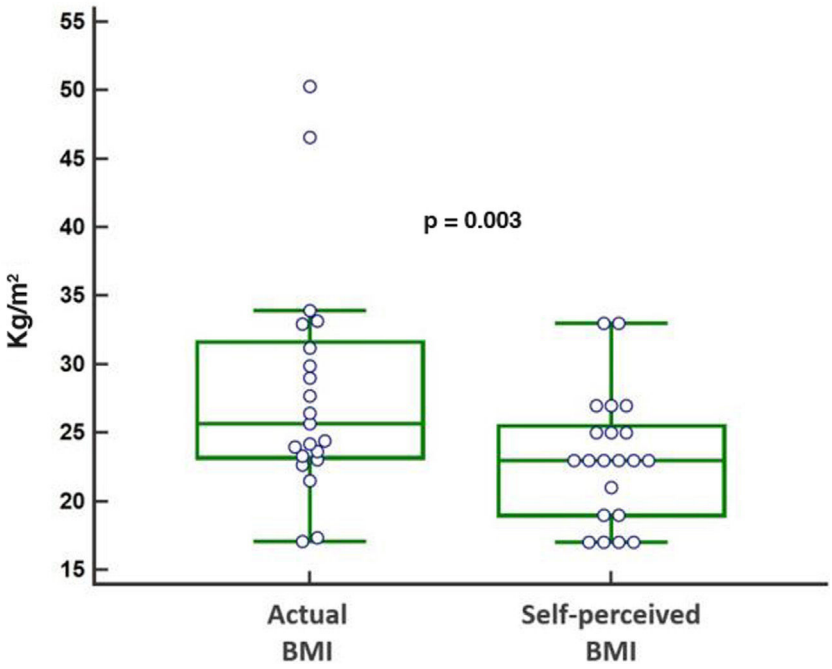

Figure 1 Differences between the actual and self-perceived body mass index (BMI).

Table 2 Relationship between body image perception and age, body mass index (BMI), intelligence quotient (IQ), and perceptual reasoning index (PRI).

Body image perception	Age	BMI	PRI	IQ
Accurate ($n = 8$)	25.1 ± 10.5	26.6 ± 8.8	67.6 ± 8.2	60.0 ± 4.9
Inaccurate ($n = 13$)	24.7 ± 10.0	28.9 ± 8.1	60.2 ± 7.2	55.0 ± 7.4
p value	0.93	0.56	0.043	0.079

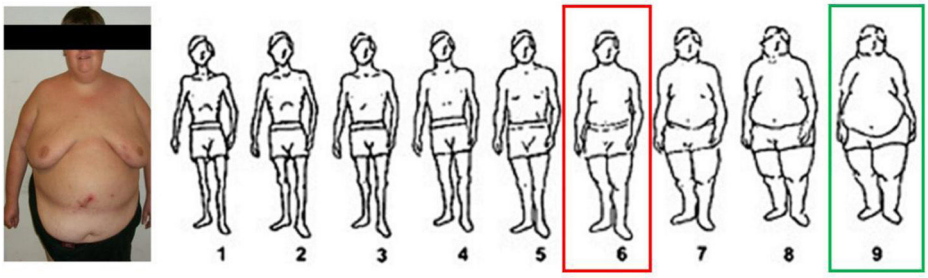

Figure 2 Distortion of body image. Patient with severe obesity and a body mass index of 32.9 kg/m^2, corresponding to figure 9 of the Stunkard scale (green) who perceived himself as figure 6 (red) corresponding to a body mass index of 28.5 kg/m^2. Stunkard scale obtained from Stunkard AJ, Sørensen T, Schulsinger F. Use of the Danish Adoption Register for the study of obesity and thinness. Res Publ Assoc Res Nerv Ment Dis 1983;60:115–20.

28.9 ± 8.1 kg/m^2, $p = 0.56$) (Table 2). On the contrary, the PRI (accurate 67.6 ± 8.2 vs inaccurate 60.2 ± 7.2, $p = 0.043$) and, to a lesser extent, the IQ (accurate 60.0 ± 4.9 vs inaccurate 55.0 ± 7.5, $p = 0.079$) were higher in the patients who correctly identified their body image (Table 2).

Of those who were wrong, 11 underestimated and 2 overestimated their body perception.

Discussion

The main finding of our study was that over half of the individuals with PWS had a distorted perception of their own body image, with most underestimating their BMI. Interestingly, patients who were unable to accurately assess their body image had a lower mean IQ and PRI compared to those who correctly identified their body image.

It is particularly noteworthy that some of our patients were significantly inaccurate in their self-perception of body image (Fig. 2), often underestimating it and perceiving themselves as thinner than they actually are. Considering the complexity of PWS and the high prevalence of obesity in this population, with obesity rates up to 40% in children and adolescents and between 82% and 98% in adults,[15] these findings could have important clinical implications. Underestimating BMI may diminish patients' awareness of the need for weight-control measures, potentially affecting their adherence to nutritional treatment.

In patients with PWS, hypothalamic dysregulation disrupts food inhibition. Moreover, unlike other patients with obesity in whom social and environmental conditioning, as well as a greater understanding and awareness, might attenuate food carving, patients with PWS are generally devoid of aesthetic and social influences that could limit their eating behaviour.[16–18] For this reason, it is important to work on the perception of body image from an early stage. Our findings provide an indication of the low level of awareness of their overweight or obesity and the risks associated with this.

Former studies that evaluated perception in other conditions with intellectual disabilities also showed a relationship with image distortion.[15] It might be hypothesized that the above-mentioned relationship between a low PRI and body image distortion could be the justification. However, this might not account for all those who failed to perceive their actual body image despite a normal PRI. Future studies might explore whether weight underestimation influences adherence to treatment and promotes the development of preventive measures and assistance that could be useful in the management of PWS, particularly regarding weight control in these patients.

Our findings might also encourage working from early instances on the perception of body image, from the different areas of approach. It would also be interesting to assess the effect of body image distortion on the effectiveness of weight management.

Our study has some limitations, including a small sample size and the absence of a control group. The applied tool, despite being a validated and widely used scale, is not specific for this syndrome, was not developed for people with intellectual disabilities and does not include within its figures a silhouette representing morbid obesity.

Despite these limitations, this is the first study to objectively investigate the relationship between self-perception and PRI in PWS, and provides a preliminary understanding of this topic to encourage future research.

Conclusions

In this study, we identified a distortion of the body image perception among patients with Prader-Willi syndrome, underestimating their body mass index in most cases. We also identified a significant relationship between body image perception and the perceptual reasoning index.

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None declared.

Authors' contributions

NRI, AG, and JS conceived and coordinated the study and acquired the data. NRI and AG verified the underlying data. NRI, AG, and JS designed the study. NRI and AG prepared the figures and tables. NRI and AG drafted the first manuscript. NRI, RC, MPJ and AG contributed to the analysis. NRI, RC, MPJ, AG, DM and JS contributed to its interpretation. NRI, AG and DM edited the manuscript. All authors critically reviewed the paper and approved its final version.

Conflicts of interest

None declared.

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