

Effects of the COVID-19 pandemic and lockdown on the mental and physical health of adults with Prader-Willi Syndrome

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Research

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Abstract

Background

Prader-Willi syndrome (PWS) is a neurodevelopmental disorder with hypothalamic dysfunction leading to obesity and behavioral disabilities, including eating disorders (EDs). We evaluated the effects of COVID-19 infection and lockdown on mental and physical health in PWS. At the end of April, 85 adults with PWS completed a self-administered questionnaire, including lockdown conditions, physical activity (PA), ED, and medical and behavioral outcomes. Body weight was measured at home and self-reported.

Results

Patients (53% women, 45% disomic) were assessed, with a mean age of 28.1 ± 8.7 years and body mass index (BMI) of 37 ± 11 kg/m². Seventy percent lived in the Paris region (France) and were confined with their parents. The mean weight change was 1.0 ± 3.3 kg. We compared patients showing weight loss ($n = 39$, -3.3 ± 2.9 kg) to patients showing weight gain ($n = 22$, $+2.3 \pm 1.5$ kg): BMI was lower (34.6 ± 9.2 versus 40.4 ± 9.5 kg/m², $p = 0.02$), PA increased (26% versus 5%, $p = 0.04$), and EDs improved (51% versus 14%, $p = 0.005$). Behavioral disorders increased for 13% of the cohort. Three individuals (3.5%) were diagnosed with non-severe COVID-19.

Conclusion

Lockdown during the COVID-19 pandemic was associated with positive effects in most French adults with PWS, with weight loss probably associated with a more favourable environment during this period. We observed no severe forms of COVID-19.

Background

Prader-Willi Syndrome (PWS, ORPHA739) is the most frequent etiology of syndromic obesity (ORPHA240371): the prevalence is approximately 1 in 21,000 newborns. This genetic syndrome results from the lack of expression of paternally-inherited imprinted genes in chromosomal region 15q11.2-q13, leading to neurodevelopment disorder and hypothalamic dysfunction leading to behavioral and endocrine disorders, mild cognitive impairment and multiple co-morbidities. Adults with PWS present difficulty in adapting to changes, emotional lability, hyperphagia, and food impulsivity. Outbursts may occur, caused by frustration over food or misunderstanding of social situations [1]. Uncontrolled eating behavior can lead to severe obesity, with many comorbidities (e.g. type 2 diabetes, hypertension). Early multidisciplinary care is required, with permanent supervision of eating practices and adapted physical activity (PA), with the aim of limiting weight gain and progression of obesity and development of complications [2].

During the COVID-19 pandemic, individuals with obesity [3], [4] and and/or diabetes [5] were shown to be at higher risk of severe forms of COVID. Whether syndromic obesity, such as PWS, also represents a risk for severe forms of COVID-19 is unknown. Moreover, complete lockdown was implemented in France in mid-March 2020 to contain the spread of COVID-19. There was concern that individuals with PWS may experience decreased PA and poorer food control during the lockdown period, as observed in children with common obesity [6], potentially worsening their condition. People with psychiatric disorders or/and intellectual disability were also shown to be particularly vulnerable, with increased anger and impulsivity [7], [8]. The aim of this cross-sectional study was to assess changes in weight, mental and physical health in adults with PWS during the COVID-19 lockdown.

Results

Cohort characteristics (Table 1)

Table 1
General characteristics of Prader-Willi patients

Patient characteristics	
N	85
Age (years)	28.1 ± 8.7 [18.0–51.0]
Sex N (%)	Female 45 (53) Male 40 (47)
BMI (kg/m ²)	36.8 ± 10.7 [19.6–68.0]
Diabetes N (%)	21 (25)
Hypertension N (%)	10 (12)
Conditions of lockdown	
Usual place of residence N (%)	Family home: 60 (70), Health-care institution: 25 (30)
Geographic region N (%)	Greater Paris region: 59 (70)
Number of rooms/Number of habitants (except institutions)	1.4 ± 0.6 [0.5–3.7]
Daily occupations	
Usual daily activity N (%)	Work 27 (32) School 18 (21) Leisure activities 22 (25) No activity 19 (23)
Physical activity	
Change in physical activity N (%)	Increased 16 (19), Decreased 57 (67), Stable 12 (14)
Daily number of physical activities	1.6 ± 1.0 [0.0–4.0]
Type of physical activity (%)	Walking 39% Fitness 21% Exercise bike/Treadmill 15% Gardening 11% Housekeeping 8% Outside games 5%

Results are expressed as the means ± SD [range] for continuous variables and as frequencies and percentages for categorical variables. BMI: body mass index.

Patient characteristics	
Change in time being sedentary N (%)	Increased 60 (75), Decreased, 5 (6) Stable 15 (19)
Sleep	
Change in time spent sleeping N (%)	Increased 20 (24), Decreased 5 (6), Stable 58 (70)
Weight/eating habits	
Weight variation (kg and %)	kg: -1.0 ± 3.3 [-13.0; +5.0] %: -1.1 ± 3.3 [-10.0; +6.2]
Weight change N (%)	Gain 22 (28), Loss 39 (49), Stable 19 (23)
Change in eating behavior N (%)	Worsening 17 (21), Improvement 29 (35), Stable 36 (44)
Aggressivity towards food N (%)	Yes 16 (20)
Psychological impact	
Difficulties during lockdown N (%)	Major 11 (13), Minor 38 (45), None 36 (42)
Results are expressed as the means \pm SD [range] for continuous variables and as frequencies and percentages for categorical variables. BMI: body mass index.	

Eighty-five patients with PWS were interviewed. Their mean age was 28.1 ± 8.7 years, mean BMI 36.8 ± 10.7 kg/m², and 87% were overweight or obese. Twenty-five percent of patients had diabetes and 12% had hypertension. Fifty-five percent had a deletion and 45% maternal disomy.

Seventy percent of patients were confined at their family home and 30% at their social health-care institution. Seventy percent of patients were living in the greater Paris region and 11% ($n = 9$) in French regions where the prevalence of COVID-19 was high. Lockdown was total in 51% of cases, partial in 48% (one hour maximum per day outside of the house), and not respected in 1%. Seventy-seven percent of patients normally performed a daily activity that was completely stopped during lockdown.

Weight change during lockdown (Table 2, Figure 1)

Table 2
Characteristics of Prader-Willi patients according to weight change categories

Characteristics	Weight loss	Weight gain	p-value
N	39	22	
Age (years)	28.9 ± 8.3 [18.0–51.0]	27.3 ± 9.9 [18.0–50.0]	0.20
BMI (kg/m^2)	34.6 ± 9.2 [19.6–52.0]	40.4 ± 9.5 [22.5–58.6]	0.02
Weight variation (kg and %)	kg - 3.3 ± 2.9 % -3.7 ± 2.5	kg + 2.3 ± 1.5 % $+2.4 \pm 1.6$	< 0.0001
Sex N (%)	Female 22 (56), Male 17 (44)	Female 11 (50), Male 11 (50)	0.8
Type of residence N (%)	Institution 6 (15) Family home 33 (85)	Institution 2 (9) Family home 20 (91)	0.69
Usual daily activities N (%)	Yes 34 (87), No 5 (13)	Yes 15 (68), No 7 (32)	0.10
Change in physical activity N (%)	Increased 10 (26) No increase 29 (74)	Increased 1 (5) No increase 21 (95)	0.04
Change in time being sedentary (%)	Decreased 3 (8) No decrease 35 (92)	Decreased 1 (5) No decrease 21 (95)	> 0.99
Change in eating behavior (%)	Improvement 20 (51) No improvement 19 (49)	Improvement 3 (14) No improvement 19 (86)	0.0054

Results are expressed as the Means \pm SD [range] for continuous variables and as frequencies and percentages for categorical variables. BMI: body mass index.

During lockdown, 67% of patients reported a decrease in daily PA. The mean number of daily PAs was $1.6 \pm 1.0/\text{days}$ [0–4]. Feeding behavior was reported to have improved for 35% of patients and did not change for 44%. Forty-nine percent of patients lost weight (mean \pm SD: -3.3 ± 2.9 kg), it was stable for 23%, and 28% gained weight ($+2.3 \pm 1.5$ kg). Mean weight loss was 1.0 ± 3.3 kg (-1.1 ± 3.3 % [-10.0; +6.2]).

Patients who lost weight had a lower BMI before the lockdown than those who gained weight (34.6 ± 9.2 versus 40.4 ± 9.5 kg/m^2 , $p = 0.02$). There were no differences between groups in terms of age, sex, genetic subtype, or living conditions during lockdown. Weight loss was more frequently observed for patients with increased PA (26% versus 5% $p = 0.04$) and those with improved eating behavior (51% versus 14% $p = 0.005$).

Medical and mental health during the COVID-19 pandemic and lockdown

Three patients had symptoms of non-severe COVID-19 (3.5%), although 13% ($n = 11$) were living with relatives diagnosed with COVID-19. No confirmation with PCR was performed but a serological assay performed at the end of June confirmed the seroconversion and the SARS-CoV2 infection for the three patients. No hospitalization was required. Their age was 18–25 years, their BMI 21.2–48.8 kg/m², and two had diabetes, with glycated hemoglobin between 6.6 and 8.2%.

Concerning behavioral issues, 11 patients (13%) displayed a recrudescence of behavioral disorders without a major effect on mean weight (+ 0.4 kg), and 10/11 presented more frequent outbursts, which required medical contact in two cases. Principal difficulties reported by caregivers were linked to exacerbations of anxiety and the ban on going outside.

Discussion

The lockdown had a positive impact for most French adults with PWS. Nearly 50% lost weight, in parallel with improved lifestyle behaviors (eating habits, PA). No severe form of COVID-19 was noted, despite a high prevalence of obesity and diabetes. However, a small subgroup experienced major difficulties, with the recrudescence of behavioral disorders and anxiety. These interviews also allowed us to receive news from the patients and support them.

In a recent Italian study, 20% adult patients gained weight and reported an increase in the consumption of “comfort food” (chocolate, ice cream, desserts, salty snacks, etc.) [9]. Rundle et al. hypothesized that people, especially children or adolescents, would fare worse concerning weight control at home than when engaged in their usual school curriculum [10]. School or working environments provide structure and routine for mealtimes, PA, and sleep schedules, the predominant lifestyle factors involved in obesity risk. Unlike the general population, PWS patients require permanent supervision to avoid food seeking and prevent uncontrolled eating behavior. Living conditions during the lockdown likely led to robust caregiver supervision, as parents or family were generally at home. Caregivers are particularly aware of the risk of weight gain during increased sedentarism. Indeed, medical-expert teams insist on the need of regular PA from childhood. In many homes, PA programs were set up during the lockdown, although PA levels were lower than usual daily life.

However, one recent study showed the negative psychological impact of strict lockdown measures on psychiatric patients during the COVID-19 epidemic [7]. Exacerbations of anger and impulsivity were significantly higher in psychiatric patients than in healthy controls. Certain patients with PWS suffer from psychiatric disorders. Aggressive behavior arises from the misunderstanding of social situations or occurrence of unpredictable situations. Only 13% of patients showed behavioral disorders. It is possible that patients with PWS benefited from more favorable conditions during lockdown, in a ritualized environment with adapted social interactions and a less stressful context, as everything was under control.

Only three cases of non-severe COVID-19 were reported (3.5%), although 80% were living in regions where the estimated prevalence of COVID-19 was high (> 9%) [11]. The respect for total lockdown in PWS was likely a contributing factor.

Our study had several limitations, including that our data were acquired in a sample from caregivers' reports and that no quantitative measures of activity levels or food consumption were available. It is possible that parents' sense of their children's behavior was heightened during the lockdown, potentially inadvertently biasing their responses.

Conclusions

In conclusion, the COVID-19 pandemic and lockdown did not have negative effects on most French adult patients with PWS, despite their well-known psychological vulnerability and susceptibility to gaining weight. Favorable changes in lifestyle behaviors during lockdown, strictly supervised by caregivers in a quiet and reassuring environment, were observed, in parallel with modest weight loss for half of the patients. In addition, there were no severe forms of COVID-19 in our PWS French cohort.

Methods

Study rationale and design

Eighty-five adults with PWS followed at the Reference Center for Rare Diseases in Pitié-Salpêtrière Hospital, Paris, France, were included in this monocentric study. Telephone interviews were conducted between mid-April and mid-May 2020 by a physician specialized in the care of PWS patients. Patients completed a detailed clinical questionnaire with the help of their caregivers (parents or social workers). The questionnaire included 32 questions related to living conditions during lockdown, physical and daily activity, sedentary time (time spent seated or lying down when awake and time spent in front of a screen), time sleeping, eating habits (food access, frequency of eating), medical and behavioral issues (need of medical contact, COVID-19 symptoms in patients and their relatives, frequency of outbursts, anxiety). Weight data before the lockdown were collected from the medical report and weight at the end of lockdown was measured by the patient with the help of the caregiver. Patients and their caregivers provided informed consent.

Statistical methods

Descriptive statistics of participant characteristics are expressed as the means and standard deviation (SD) for continuous variables and frequencies and percentages for categorical variables. The mean values of patients who lost weight were compared to those who gained weight using two-sample Fisher tests. All statistical analyses were performed using GraphPad Prism version 8.0 and statistical significance was considered for two-sided p-values < 0.05.

Declarations

Ethics approval and consent to participate:

Patients and their caregivers provided informed consent.

Consent for publication:

All authors reviewed and approved the final article.

Availability of data and materials:

H.Mosbah, C. Poitou, store all data which are available.

Competing interests:

None of the authors declare a conflict of interest.

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No

Authors' contributions:

HM and CP designed the study. HM, CP, and MC contributed to patient recruitment and performed the clinical survey. FJ contributed to data management. HM and CP analyzed the data and wrote the manuscript. HM, CP, MC, MT, JMO, and KC contributed to the data presentation and editing of the manuscript. All authors reviewed the manuscript. CP is the guarantor of this work and, as such, had full access to all the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis.

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Figures

Weight variation during lockdown

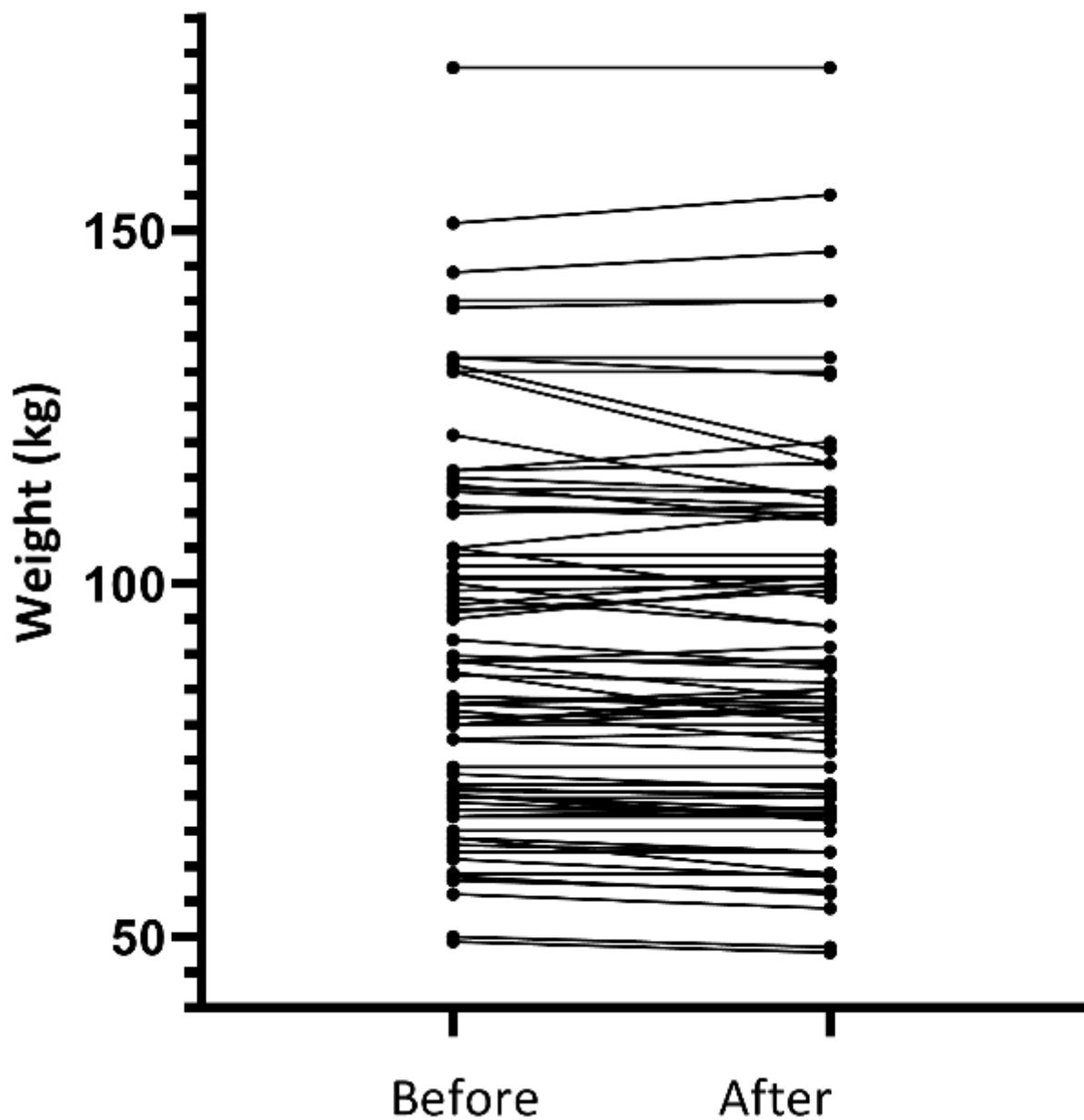


Figure 1

Weight change (individual values) in the total cohort of PWS (weight before lockdown- weight after lockdown).