

Physical activity in patients with Prader-Willi syndrome - A systematic review of observational and interventional studies

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SUPPLEMENTARY MATERIAL

Table S1. Study quality of intervention studies

References	Criteria														Number of "fatal flaws"	Quality rating
	1	2	3	4	5	6	7	8	9	10	11	12	13	14		
<i>Randomized or non-randomized controlled trials</i>																
Vismara 2010 [1]	No	NA	NR	No	No	NR	Yes	Yes	Yes	Yes	Yes	No	Yes	Yes	1	Fair
Eiholzer 2003 [2]	No	NA	NR	No	No	Yes	Yes	NA	NR	Yes	Yes	No	Yes	NR	2	Fair
Rubin, 2019 [3]	No	NA	NR	No	No	Yes	No	Yes	Yes	Yes	Yes	Yes	Yes	Yes	0	Good
Schlumpf 2006 [4]	No	NA	NR	No	No	Yes	NR	NR	NR	No	Yes	No	Yes	NR	2	Poor
Shields 2020 [5]	Yes	Yes	Yes	No	Yes	Yes	NR	NR	Yes	Yes	Yes	No	Yes	NR	0	Good
<i>Single-group intervention studies</i>																
Bellicha 2020 [6]	Yes	Yes	No	NR	No	Yes	Yes	No	Yes	Yes	No	NA	-	-	1	Fair
Grolla 2011 [7]	Yes	No	No	NR	Yes	No	Yes	NR	NR	No	No	NA	-	-	4	Poor
Hsu 2018 [8]	Yes	Yes	No	NR	No	Yes	Yes	NR	Yes	Yes	No	NA	-	-	1	Fair

Criteria for controlled trials: #1- Randomized study, #2- Adequate randomization method, #3- Treatment allocation concealment, #4- Blinding treatment assignment, #5- Blinding outcome assessors, #6- **Similar baseline characteristics**, #7- Drop-out rate <20%, #8- Differential drop-out rate between groups <15%, #9- **High adherence**, #10- **Similar background treatments**, #11- Valid and reliable outcome measures, #12- Sample size justification, #13- Pre-specified outcomes/subgroups, #14- All randomized participants analyzed (ITT analysis).

Criteria for single-group interventions: #1- Question/objective clearly stated, #2- **Eligibility criteria pre-defined**, #3- **Representativeness**, #4- Enrolment rates, #5- Sufficient sample size, #6- Intervention clearly described, #7- Valid and reliable outcome measures, #8- Outcome assessors blinded, #9- **Drop-out rate <20% or intent to treat analysis**, #10- **Statistical analyses examined changes in outcomes**, #11- Multiple time points for outcome measurement, #12- Account for individual changes.

Criteria defined as fatal flaws are grey shaded.

Table S2. Description of habitual physical activity and sedentary behavior

References	Patients with PWS	Control group	P-value
Outcomes			
Bellicha 2020 [6]	Adults with PWS	Adults with NSO	
Activity counts, counts/min	Median (P25; P75): 211 (141; 333)	334 (292; 384)	< 0.05
Sedentary time, % wear time	72 (69; 75)	58 (53; 64)	< 0.001
Prolonged sedentary bouts (\geq 30 min), % total sedentary time	41%	18%	< 0.001
Light - intensity PA, % wear time	24 (23; 28)	38 (32; 45)	< 0.001
MVPA, % wear time	1.7 (0.8; 5.0)	3.2 (2.6; 4.5)	0.11
MVPA in \geq 10 - min bouts, min/week	5 (0; 101)	20 (0; 68)	0.38
Meet MVPA guidelines, n (%) ¹	2 (20%)	1 (5%)	0.52
Borland 2020 [9]	Adults with PWS	Adults with ID: 132 (42%)	< 0.05
Reported participation in sports/PA over the past 3 months, %	19 (63%)	Down Syndrome: 39 (61%)	NR
		Fragile X Syndrome: 29 (56%)	NR
		Williams Syndrome: 24 (53%)	NR
		Autism: 51 (54%)	NR
Butler 2007 [10]	Children/adults with PWS	Children/adults with NSO	
PA-induced EE, kcal/min	Mean (SD): 0.86 (0.37)	1.27 (0.64)	< 0.001
Exercise-induced EE, kcal/min	1.92 (0.61)	3.08 (0.99)	< 0.001
Total mechanical work ² , W/8h	30 (12)	46 (34)	< 0.001
Castner 2014 [11]	Children with PWS	Children with NSO	
Sedentary time, min/d	Mean (SD): 660.5 (15.1)	638.7 (11.7)	0.26
Light-intensity PA, min/d	130.3 (9.4)	161.7 (7.0)	< 0.01
Moderate-intensity PA, min/d	26.2 (2.4)	28.9 (1.9)	0.36
Vigorous-intensity PA, min/d	9.9 (1.2)	14.1 (1.3)	< 0.05
MVPA, min/d	36.1 (3.2)	43.0 (2.9)	0.14
Meet MVPA guidelines ³ , n (%)	2 (8%)	7 (18%)	NR
Duran 2016 [12]	Children with PWS	--	--
Light-intensity PA, min/d	Mean (SD): 129.0 (46.8)	--	--
Moderate-intensity PA, min/d	25.3 (11.2)	--	--
Vigorous-intensity PA, min/d	9.3 (5.0)	--	--
MVPA, min/d	34.6 (14.1)	--	--
Eiholzer 2003 [2]	Children with PWS	Children with normal weight	
3-days walking distance, km	Mean: 11.1	24.6	< 0.05
McAlister 2018 [13]	Children with PWS	Children with NSO	
MVPA, min/d	Mean (SD): 33.9 (14.7)	44.0 (17.7)	< 0.05
Meet MVPA guidelines ³ , n (%)	1 (5%)	3 (8.8%)	ns
Nordstrom 2013 [14]	Adults with PWS	Adults with Down-syndrome	
Total PA, counts/min	Mean (SD): 249 (114.1)	306 (91.4)	0.06
Steps/d	5781 (3053)	6949 (2673)	ns
Sedentary time, min/d	564 (66;9)	491 (74.4)	0.01
Light-intensity PA, min/d	174 (47.5)	240 (60.3)	< 0.001
MVPA, min/d	26.2 (20.8)	25.5 (17.7)	ns
MVPA in \geq 10-min bouts, min/d	14.2 (15.1)	9.4 (11.6)	ns
Meet MVPA guidelines ⁴ , %	Females: 15%, males: 25%	Females: 8%, males: 7%	NR
Rubin, 2019 [15]	Children with PWS	Children with NSO	
Total PA, min/d	Mean (SE): 165 (11)	225 (10)	< 0.01
MVPA, min/d	34 (4)	45 (3)	0.07
Vigorous-intensity PA, min/d	11 (2)	14 (1)	0.22
Schlumpf 2006 [4]	Children with PWS	--	--
3-days walking distance, km	Mean (SD): 12.5 (6.2)	--	--
Sellinger 2006 [16]	Children/adults with PWS	Down syndrome: 1.65 (0.08)	ns
Reported frequency of PA participation ⁵	Mean (SD): 1.41 (0.15)	Williams syndrome: 1.86 (0.09)	< 0.05
van den Berg-Emons 2008 [17]	Children with PWS	Children without obesity	
Duration of dynamic activities ⁶ , % of a 24-hour period	Mean (SD): 8.7 (2.5)	12.0 (3.1)	0.01

Mean mobility, g	0.023 (0.004)	0.028 (0.008)	0.09
Walking speed, g	0.178 (0.018)	0.170 (0.025)	0.32
Walking periods > 10 s, n/d	137 (42)	171 (45)	0.09
Breaks in sedentary time, n/d	116 (31)	143 (42)	0.13
Van Mil 2000 [18]	Children with PWS	Children with NSO	
PA-induced EE, MJ/d	Mean (SD): 1.07 (0.7)	2.56 (1.03)	< 0.001
PA-induced EE relative to body weight, kJ/kg/d	23.1 (17.1)	46.09 (17.8)	< 0.001
PA level ⁷	1.33 (0.15)	1.55 (0.12)	< 0.001
Woods 2018 [19]	Adults with PWS	--	--
Steps/d	Mean (SEM): 7632 (1171)	--	--

¹ MVPA guidelines in adults at the time of the study: 150 min/week

² Total mechanical work = assessed by force platforms during 8h on a calorimetric chamber

³ MVPA guidelines in children at the time of the study: 60 min/d

⁴ MVPA guidelines in adults at the time of the study: 30 min/d of MVPA in 8- to 10-min bouts.

⁵ Frequency assessed with a 5-point scale: 1= rarely, never to 5= daily

⁶ Walking, cycling, general movement

⁷ PA level = average daily metabolic rate divided by basal metabolic rate

Abbreviations: EE, energy expenditure; ID, intellectual disabilities; MVPA, moderate-to-vigorous PA; NSO, non-syndromic obesity

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