

Physical Activity in Children with Prader-Willi Syndrome: A Parents' Perspective

Daniela A. Rubin¹, Michele Mouttapa¹, Jie Wu Weiss¹, and Angelica Barrera-Ng²

¹California University Fullerton, Fullerton

²University of California, San Diego

Abstract

Background. Physical activity (PA) is vital for the management of weight among those with Prader Willi Syndrome (PWS). However, little is known about characteristics of PA in individuals with PWS.

Objective/Hypothesis. To assess from the parent's perspective, PA levels among individuals with PWS, their preferences for specific activities, and perceived benefits, barriers, and resources needed to participate in PA.

Methods. Participants were 90 parents and caregivers of a child with PWS, predominantly in California. Survey questions included their child's participation in different PAs, perceived benefits and barriers to enrolling their child in a PA program, and perceived needs to facilitate their child being physically active.

Results. Walking was the predominant PA, representing 66% of the activity time across all age groups. Children 10-17 years old engaged in more moderate intensity PA per week than children 5-10 years old. Children 5-10 years old engaged in more vigorous PA than those 18+ years old.

Parents reported that they would be encouraged to enroll their child in a PA program if it would improve their children's motor skills and balance (78.2%) as well as stamina and strength (74.4%). Time commitment and travel were most common barriers. Less financial constraints (70.1%) and having more time (54.7%) were listed as needs.

Conclusions. Similar to individuals without PWS, vigorous PA declines with age. It is recommended that caregivers and health care providers emphasize the role of vigorous and bone-strengthening PA as children with PWS approach adolescence.

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Keywords:

Introduction

Prader-Willi syndrome (PWS) is the most characterized form of congenital obesity and results from an alteration of the paternal chromosome 15. The prevalence of PWS ranges from 1 in 10,000 to 1 in 25,000 live-births (Butler, Phillip, & Whitman, 2006). The syndrome is characterized by infantile hypotonia with failure to thrive at birth. During childhood the onset of hyperphagia takes place and strict dietary management is needed. Other characteristics of the syndrome include growth hormone deficiency (Burman, Ritzen, & Lindgren, 2001), poor motor coordination and muscle tone, increased adiposity, and low

energy levels (Butler, Phillip, & Whitman, 2006). Individuals with PWS also present a lower proportion of muscle mass compared to people of the same body size, therefore, their energy expenditure is considerably lower when compared to controls (Butler, Theodoro, Bittel, & Donnelly, 2007). In addition, individuals with PWS present behavioral challenges and wide ranges of cognitive levels (Ho & Dimitroupulos, 2010). As a result, management of this syndrome requires a comprehensive approach including different therapies and treatments (Eiholzer & Whitman, 2004).

In individuals of all ages, with and without PWS, physical activity (PA) is a key factor for

sustaining energy balance and weight maintenance (Eiholzer & Whitman, 2004; Fulton, Garg, Galuska, Rattay, & Caspersen, 2004; Haskell et al., 2007; Mullins & Vogl-Maier, 1987). Physical activity stimulates muscle growth, motor coordination and balance (Eiholzer et al., 2003; Haskell et al., 2007). Physical activity offers psychological benefits to individuals such as improved self-esteem (Altintas & Asci, 2008) and quality of life (Schwimmer, Burwinkle, & Varni, 2003). Previous reports show that school-day levels of PA in children with PWS are lower than in children without the syndrome; however these differences are less when overall daily PA is considered (van den Berg-Emons, Festen, Hokken-Koelega, Bussmann, & Starm, 2008). Similar to children with PWS, adults with PWS present less spontaneous PA even in comparison to obese adults without the syndrome (Butler, Theodoro, Bittel & Donnelly, 2007).

Parents and caregivers play a pivotal role in children's PA, through example (e.g., the parent engages in regular PA) and through the provision of opportunities (e.g., enrolling their children in organized sports activities, walking with their child to school) (Bauer, Nelson, Boutelle, & Neumark-Sztainer, 2008). Challenges and barriers to participation in PA in children and adults are varied (Sherwood & Jeffery, 2000). Because of the wide spectrum in cognitive profile and behavior in PWS (Ho & Dimitropoulos, 2010), parents of these individuals with PWS may face other facilitators and challenges to PA participation.

The purpose of this study is to present, from the parents'/caregivers' perspective, descriptive PA data on individuals with PWS, as well as the perceived barriers and facilitators for PA in this population. We believe that identification of barriers and facilitators, such as parents' perceived benefits of their child's engagement in PA, can help define future intervention strategies for children with PWS.

Methods

Participants

Participants were recruited mostly from the Prader-Willi California Foundation registry.

Four hundred surveys were mailed out to parents in the registry or distributed at PWS related functions. The response rate was approximately 25 % which provided an initial n=104. As the results from surveys containing children younger than five years old (n=14) were excluded, the analytic sample consisted of 90 participants (n=90). All participants had one child who had been medically diagnosed with PWS (diagnosis letter from physician not provided). Only one parent/caregiver from each household was considered for the survey.

Procedures

The survey was mailed to all 375 members of the Prader-Willi California Foundation (PWCF) registry with an information sheet about the purpose of the project. An advertisement about the study was also placed in the Prader-Willi Syndrome USA Association website. PWCF members were invited to complete the self-report survey and mail it back to the researchers using a pre-addressed stamped envelope. Parents not belonging to PWCF but were interested in participating contacted the principal investigators via email and requested survey materials. Participants who returned the completed survey received a \$10 gift card by mail. Survey responses were entered into a password-protected computer, with identifying information removed. Once the data was entered, it was compared to the original surveys to ensure accuracy of data entry by a different researcher. The study protocol was approved by the Institutional Review Board of the investigators' affiliation as well as sponsoring agency.

Measures

Demographic Characteristics.

Parent participants reported their age, sex, ethnicity, language spoken at home, employment status, and educational level.

Levels of PA among Parents.

By answering two separate multiple choice questions, parents reported the number of days in the past week that they engaged in PA, as well as the usual duration of each PA session (in minutes).

Levels of PA among individuals with PWS and other activities.

First, parents reported the number of times per week and the number of minutes per day that each of their children (including their child with PWS) engaged in structured and recreational PA. Then, parents were asked to report how many minutes and times per week their child with PWS engaged in a list of activities. The list of activities was developed from activities previously used on other children PA questionnaires (Gilmer, Speck, Bradley, Harrell, & Belyea, 1996) as well as activities reported in four group interviews with parents of youth with PWS from Southern California. Activities were then classified using metabolic equivalents (MET) into moderate intensity (≥ 3.8 -5.9 METS), vigorous intensity (≥ 6 METS), and bone-strengthening based on the compendium of PA (Ainsworth et al., 2000; Harrell et al., 2005). METs were used to categorize activities, but not to estimate energy expenditure. Because individuals with PWS have less lean mass, it is likely their energy expenditure for physical activities is lower than normal. Therefore, actual MET expenditure may not be reliably estimated based upon the survey measures we utilized (M. G. Butler et al., 2007). However, although the absolute MET value for activities in individuals with PWS may be different, the relative difference in MET values among the activities may remain relatively constant, allowing for their use to classify activities into moderate or vigorous intensity. Parents also reported whether or not their child with PWS has ever engaged in the following therapies: physical, aquatic, horse, speech, and occupational. In addition, parents reported if their child attended learning disability or psychological services, or was under growth hormone replacement therapy, dietary management, and structured PA.

Perceived benefits and barriers. Participants were asked the extent to which the following reasons would *encourage* them to enroll their child in a PA program: [their child] benefitting from the program, improving motor skills and balance, improving stamina/strength, and socially interacting with other children, [the parent participant] talking to doctors and other experts, receiving increased opportunities for

participation in other programs, and meeting other parents of children with PWS. Participants were also asked the extent to which the following reasons would discourage them to enroll their child in a PA program: the time commitment, traveling to the site, and not getting along/relating to the program staff. Responses to all items ranged from 1 (Strongly Disagree) to 5 (Strongly Agree). The reasons were derived from feedback obtained in four group interviews with 20 parents of youth with PWS.

Data analysis

Participants were stratified into the following age groups: children 5 to 10 years ($n=28$), adolescents 11 to 17 years old ($n=27$) and adults 18 years old and over ($n=35$). These age – groups were chosen because they represent physical and developmental milestones. Descriptive statistics (frequencies and percentages, means and standard deviations where appropriate) were calculated for parent and sibling characteristics (parent’s age, gender, employment, education level, marital status, PA frequency and duration, and siblings’ participation in structured and recreational PA in minutes/week), and characteristics of the child with PWS (gender, age group, and lifetime participation in treatments and therapies).

For the child with PWS, frequencies and percentages were calculated for weekly participation (yes or no) for the various PAs that were categorized as moderate, vigorous, or bone-strengthening. Chi-square tests were conducted to determine whether percentages for these PA variables differed according to the age of the child with PWS. Means and standard deviations were calculated for the average number of minutes/week that the child with PWS engaged in each of the moderate, vigorous and bone strengthening PAs, as well as the total number of minutes for each of these three PA categories. ANOVAs were performed to determine age group differences on these PA variables. Last, ANOVA tests were performed to determine whether responses for each of the perceived benefit, perceived barrier, and perceived need variables differed among the three age groups.

Results

Demographic Characteristics

Parent and sibling characteristics. Mean age of parent participants was 49.2 ± 10.5 years. The majority of the parent participants was female (86.7%), Caucasian (70.8%), and spoke English at home (92.2%). Sixty-three percent of the sample was employed; of them 26.8% worked over 40 hours per week. The education level of participants was high, as 26.3% had a college degree, and 35.1% had at least some graduate school or a graduate/professional degree. Most participants (82.2%) were married. Only six parents (6.7%) reported that they did not engage in any past week PA; 31.4% engaged in PA 1 to 2 times in the past week, 35.6% 3 to 4 times in the past week, 17.8% 5 to 6 times in the past week, and five of them (5.6%) every day. Among those who did engage in PA (n=84), 60.7% reported an average duration of at least 30 minutes per PA session. Only 18 parents (20%) of the entire sample engaged in PA at least 5 times per week for 30 minutes or more each time. Parents' PA did not vary by age of their child with PWS (5 to 10 years, 11 to 17 years, and 18+ years).

Table 1

Lifetime participation in treatments and therapies to manage PWS Symptoms (n = 90)		
Treatment	N	%
Learning disability services	46	51.7
Psychologist	35	39.3
Growth hormone replacement	51	57.3
Dietary management	70	79.5
Structured PA	54	61.4
Therapy	N	%
Physical	55	61.1
Aquatic	27	30.3
Horse	27	30.3
Speech	76	85.4
Occupational	56	62.9

The children with PWS had an average of 1 ± 1 sibling. Siblings participated in 379 ± 614

minutes per week of recreational PA and 255 ± 370 minutes per week of structured PA. Siblings' PA did not vary by age group that the child with PWS belonged to.

Characteristics of the individuals with PWS. Forty-eight of the individuals with PWS (53.3%) were male. There were twenty-eight children (31.1%) ages 5 to 10 years old, 27 adolescents (30.0%) ages 11 to 17 years old, and 35 adults (38.9%) ages 18 to 49 years old (mean 28 ± 9 years). Frequencies and percentages for the types of treatments and therapies to help manage PWS symptoms that the child had participated in are presented in Table 1. The majority (60%) of individuals with PWS engaged in both PA and physical therapy.

Moderate, Vigorous, and Bone Strengthening PA among individuals with PWS

Moderate PA (MPA).

There were no age group differences in past week participation (yes or no) in any of the MPAs, all of which are listed in Table 2. However, there was an age group difference in the total number of minutes per week spent in MPA: adolescents engaged in more MPA than children (262 ± 222 vs. 100 ± 76 minutes per week; $p < 0.05$).

Vigorous PA (VPA).

There were age group differences for past week participation (yes or no) for some activities. Running was more common in adolescents than in children or adults (37.0% vs. 17.9% and 8.6%, respectively; $p < 0.05$). Swimming ($p < 0.05$) and jumping on a trampoline ($p < 0.01$) were significantly more common in children (64.3% and 25.0%, respectively) compared to the other groups. In addition, children engaged in more VPA than adults 18 years and above (239 ± 361 vs. 70 ± 161 minutes per week, $p < 0.05$). See Table 3. Relatively few individuals with PWS engaged in competitive sports (8 out of 90 children, 8.9%) and martial arts (3 out of 90 children, 3.3%).

Muscle/bone strengthening PA.

With the exception of playing games (e.g., tug of war), there were no age group differences for past week engagement (yes or no) in

Table 2

Frequency and average number of minutes per week of moderate physical activity (MPA) in individuals with PWS by age group (n= 90)

	5-10 years (n = 28)	11-17 years (n = 27)	18+ years (n = 35)	
Moderate PA	f (% yes)	f (% yes)	f (% yes)	p
Hiking	2 (7.1)	3 (11.1)	1 (3.3)	0.52
Walking	21 (75.0)	22 (81.5)	22 (62.9)	0.25
Bicycle/Tricycle riding	8 (28.6)	7 (25.9)	4 (11.4)	0.19
Exercise video games	3 (10.7)	6 (22.2)	5 (14.3)	0.48
Housework/Yardwork	9 (32.1)	14 (51.9)	16 (45.7)	0.32
Other Moderate PA ¹	4 (14.8)	4 (15.4)	5 (19.2)	0.90
	Mean (SD)	Mean (SD)	Mean (SD)	p
Total Moderate PA (min).	100.0 (76) ^a	262 (222) ^b	192 (225) ^{ab}	0.01**

**p < .01.

Note. Groups containing the same superscript letters have homogenous values for the given variable.

¹ Responses from individual participants included climbing stairs (n = 3), using cardio equipment at a moderate pace (n = 3), bowling (n = 1), horseback riding (n = 2), *Wii Sports* (n = 1), slow walking (n = 1), hula-hoop (n = 1), and moderate-level outdoor play (n = 1).

Table 3

Frequency and average number of minutes per week of vigorous physical activity (VPA) in individuals with PWS by age-group (n= 90)

	5-10 years (n = 28)	11-17 years (n = 27)	18+ years (n = 35)	
Vigorous PA	f (% yes)	f (% yes)	f (% yes)	p
Jump rope	2 (7.1)	1 (3.7)	0 (0.0)	0.29
Running	5 (17.9)	10 (37.0)	3 (8.6)	0.02*
Swimming	18 (64.3)	12 (44.4)	11 (31.4)	0.03*
Martial arts	1 (3.6)	2 (7.4)	0 (0.0)	0.27
Competitive sports	2 (7.1)	3 (11.1)	3 (8.6)	0.87
Trampoline	7 (25.0)	3 (11.1)	0 (0.0)	0.01**
Dancing	5 (17.9)	6 (22.2)	2 (5.7)	0.15
Other Vigorous PA ¹	5 (17.9)	2 (7.7)	2 (6.9)	0.34
	Mean (SD)	Mean (SD)	Mean (SD)	p
Total Vigorous PA	239 (361) ^a	146 (130) ^{ab}	70 (161) ^b	0.02*

*p < .05; **p < .01.

Note. Groups containing the same superscript letters have homogenous values for the given variable.

¹ Responses from individual participants included vigorous outdoor play (n = 3), using cardio equipment vigorously (n = 2), gymnastics (n = 2), playing basketball (n = 1), and playing tennis (n = 10).

muscle/bone strengthening PAs. Playing games was significantly more common in children than

in adolescents or adults (25.0% vs. 14.8% and 2.9% respectively, p<0.05). There was no age

group difference in the total number of minutes per week individuals engaged in muscle/bone strengthening PA. See Table 4.

Parental Perceptions of Enrolling their Child with PWS in a PA Program

Perceived benefits. The majority of parent participants “agreed” to all of the perceived benefits listed. The percentage of “strongly agree” responses were higher for their child: (1) benefitting from the program (77.0%), (2) improving motor skills and balance (78.2%), and

(3) improving stamina and strength (74.4%). The percentage of “strongly agree” responses were lower for meeting other parents of children with PWS (37.9%). ANOVA tests showed that improving motor skills and balance as a benefit from PA varied by age group ($F_{(2, 84)} = 3.91, p=0.02$). On a scale of 1 to 5, mean scores were the following: 4.9 ± 0.3 for children, 4.8 ± 0.5 for adolescents, and 4.5 ± 0.8 for adults. Parents of adults had significantly lower scores than parents with children and adolescents.

Table 4

Frequency and average number of minutes per week of muscle/bone strengthening physical activity in individuals with PWS by age group (n=90)

	5-10 years (n = 28)	11-17 years (n = 27)	18+ years (n = 35)	p
Muscle/Bone Strengthening PA				
Games (e.g., tug-of-war)	7 (25.0)	4 (14.8)	1 (2.9)	0.04*
Push-ups/Pull-ups	3 (10.7)	2 (7.4)	0 (0.0)	0.16
Rope climbing	1 (3.6)	0 (0.0)	0 (0.0)	0.33
Sit-ups	2 (7.1)	2 (7.4)	2 (5.7)	0.96
Resistance exercises	2 (7.1)	6 (22.2)	5 (14.3)	0.28
Other	7 (25.0)	3 (11.1)	2 (6.9)	0.13
	Mean (SD)	Mean (SD)	Mean (SD)	p
Total Muscle/Bone Strengthening	41 (65)	59 (129)	30 (94)	0.52

*p < .05; **p < .01.

Note. Groups containing the same superscript letters have homogenous values for the given variable.

¹ Responses from individual participants included physical therapy (n = 2), climbing (e.g., indoor rock climbing, n = 2), yoga (n = 1), circuit training (n = 1), core strengthening (n = 1), stretching (n = 1), and crawling (n = 1).

Perceived barriers.

Less than 40% of parents either “agreed” or “strongly agreed” that time commitment and travel to the program site would be a barrier to participating in a program. Only 15.1% of respondents “agreed” or “strongly agreed” that not getting along with program facilitators would be a barrier. ANOVA tests indicated that parents’ perceived barriers did not vary by the age of their child ($p > .05$).

Perceived needs.

Over 70.1% of parents either “agreed” or “strongly agreed” that they needed more

financial support, and over one half answered similarly for needing more time out of their busy schedule (54.7%) and more equipment (51.2%). ANOVA tests indicated that parents’ perceived need for encouragement (1 to 5 scale) varied significantly by age group. Parents of 5-10 year old children had a higher need for encouragement compared to those of adolescents or adults (3.7 ± 1.3 vs. 2.9 ± 1.3 and 2.8 ± 1.5 , respectively; ($F_{(2, 84)} = 3.47, p=0.04$). Non-significant trends were found for parents’ perceived need of financial support ($p=0.07$) and time out of their busy schedule ($p=0.06$). Specifically, parents of children who were 18

years and older reported a higher need of financial support compared to parents of children or adolescents (2.6 ± 1.3 vs. 2.0 ± 1.2 and 2.0 ± 1.1 , respectively; ($F_{(2, 84)} = 2.81$, $p=0.10$). Furthermore, to a near significant level, parents of children reported less need for time out of their busy schedule compared to parents of adolescents or adults (2.2 ± 1.2 vs. 3.0 ± 1.3 and 2.9 ± 1.4 , respectively; ($F_{(2, 81)} = 2.87$, $p=0.07$).

Discussion

This study is unique because (1) we obtained a relatively large sample of parents with children who have PWS, (2) the individuals of PWS being studied included young children, adolescents, as well as adults, and (3) we gathered data, from the parents' perspective, on the PA patterns of their child with PWS, as well as the parents' perceived benefits, barriers, and facilitators for engaging their child with PWS in PA. The findings of this study suggest that adolescents with PWS engage in more moderate intensity PA than younger children. Conversely, children ages 5-10 years appear to be engaged in more vigorous PA during the week compared to adults. The trend we observed is similar to PA patterns in children and adolescents who do not have PWS (Bradley, McMurray, Harrell, & Deng, 2000; Trost et al., 2002). Trost and colleagues found that as children progressed from elementary to high school there was a consistent decrease in time spent in vigorous PA during the week (Trost et al., 2002). Similarly, Bradley et al. (Bradley et al., 2000) showed that as children progressed from elementary to middle school their choices for sedentary and more moderate intensity activities increased.

We found that common activities among individuals with PWS were walking and yard work. Walking is also the most common activity in adults in the United States (Centers for Disease Control and Prevention, 2012), as well as with youth with PWS (van den Berg-Emons et al., 2008). European children ages 6-12 with PWS have been reported to cover about 12.6 km in three days (Schlumpf et al., 2006). We also found that very few individuals with PWS participated in competitive sports as

indicated in Table 3 (11.1% at most), which is in sharp contrast to the general population of children in California, where approximately 55.5% of children ages 10-11, 59.2% of children ages 12-14, and 47.4% of children ages 15-17 years old participate in sports team or taken sport lessons. It is possible that this lack of participation in organized sports relates to physical, developmental and cognitive characteristics of individuals with PWS. However, participation in organized sports should not be discouraged, as sports provide a social atmosphere which may be good for development of relationships with peers, contribute to character building, and improved self-esteem (Patel & Greydanus, 2010). Sports participation is highly encouraged in other people with intellectual or physical disabilities; it is a matter of finding the right activity and setting for a person with PWS (Patel & Greydanus, 2010).

A noteworthy finding of this study is that adults with PWS spent half of the time on bone or muscle strengthening activities (about 30 minutes per week) compared to adolescents or children. Strengthening activities are recommended for the general population at least twice a week and for youth 2-3 times per week (Faigenbaum et al., 2009). In people with PWS strengthening activities are particularly important for increasing and sustaining muscle mass. Increased muscle mass allows for more stamina to carry out everyday activities and improving overall quality of life (Eiholzer et al., 2003; Whitman, Myers, Carrel, & Allen, 2002). In addition, lean mass is more metabolically active than adipose tissue, thus a larger muscle mass contributes to sustaining energy balance (McArdle, Katch, & Katch, 2007). It appears that PA interventions, particularly those that involve muscle and bone strengthening, are particularly important for individuals with PWS, because independent of their body size, they have less muscle mass compared to the general population (Brambilla et al., 1997; Rubin, Wright, Haqq, Castner, & Judelson, 2012).

A large proportion of responding parents indicated that their child benefitted from participating in a PA program and improving

motor skills and balance was important to them. These results support the fact that 61% of the sample had been enrolled in physical therapy sessions during some period of their lifetime. Furthermore, 74.4% of parents in this study believed that a major benefit of engaging their child with PWS in PA is to improve his/her stamina and strength. The parents' beliefs are consistent with the findings of three exercise interventions that demonstrated increased spontaneous PA, muscle endurance, and aerobic capacity in individuals with PWS (Eiholzer et al., 2003; Schlumpf et al., 2006; Silverthorn & Hornak, 1993). Parents also indicated that engaging in PA had a social component that was beneficial to their child. Similar to other populations (Sherwood & Jeffery, 2000), time and financial support appear to be main barriers to participating in PA in PWS.

Limitations

A limitation of the study is the survey method, in that PA data being self-reported by parents are less reliable and valid than observation or direct monitoring. As previously indicated, we used MET values from individuals without PWS and it is likely that MET values for different activities are lower in those with PWS because of their lower amount of lean mass. In addition, the sample may not necessarily represent the entire population of parents of individuals with PWS, as our sample of parents reported a relatively high level of formal education, and the majority of them lived in the state of California.

Implications of the Results

The results of this study show that most individuals with PWS are engaged in PA of moderate intensity. However, participation in vigorous PA decreases with age. Vigorous PA

provides health-related (D. A. Rubin, McMurray, & Harrell, 2008) and fitness-related benefits above and beyond the ones of moderate intensity PA (Haskell et al., 2007). Similarly, with increased age there is less participation in bone and strengthening activities. It is recommended that caregivers and health care providers continue to emphasize the role of vigorous and strengthening PA as individual with PWS grows into adulthood.

Similar to parents in the general population, parents of individuals with PWS are encouraged to enroll their child in a PA program if they perceive that specific beneficial outcomes (e.g., improved motor skills and balance) may occur. However, parents of younger children may need additional encouragement to have their child participate in all types of PA, perhaps because they are still learning to cope with the challenges that a diagnosis of PWS involves. Parents' perceptions of their child with PWS engaging in PA are particularly important to consider, because parents are most often the lifetime caregivers and decision makers for the PWS child. Hence, parents' perceptions regarding the benefits and barrier of their child engaging in PA can significantly impact the PA levels of their children.

Acknowledgements

Authors would like to thank the Prader-Willi Syndrome California Foundation, the Prader-Willi Syndrome USA Association, and the Foundation for Prader-Willi Syndrome Research for their help with advertising the study. Authors also thank the parents and caregivers for their time. Special thanks to Lisa Graziano from the PWCF for distributing the surveys

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Author Information

*Daniela A. Rubin
California State University, Fullerton
Department of Kinesiology
800 N. State College Blvd
Fullerton, CA 92834
E-mail: drubin@fullerton.edu

Michele Mouttapa
California State University, Fullerton
Department of Health Science

Jie Wu Weiss
California State University, Fullerton
Department of Health Science

Angelica Barrera-Ng
University of California, San Diego
Center for Wireless & Population Health Systems

* corresponding author