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# 1 Abstract

2 **Background:** Regular physical activity (PA) is increasingly recognised as important in the care of patients with Cystic Fibrosis (CF) but there is a dearth of evidence regarding physical 3 activity levels (PAL) or how these are accrued in those with CF. Methods: Physical activity 4 5 was measured by a hip-worn accelerometer for seven consecutive days by eighteen children 6 (10 boys;  $12.4 \pm 2.8$  years) with mild to moderate CF and eighteen age- and sex-matched 7 controls (10 boys;  $12.5 \pm 2.7$  years). **Results:** Both CF and healthy children demonstrated 8 similar PAL and patterns of accumulation across the intensity spectrum, with higher levels of PA during weekdays in both groups. FEV<sub>1</sub> was predicted by high-light PA in CF compared to 9 low-light PA in healthy children. Conclusion: These findings highlight weekends and light 10 PA as areas warranting further research for the development of effective intervention 11 strategies to increase PA in the youth CF population. 12

# Introduction

Cystic Fibrosis (CF) is the most prevalent lethal autosomal recessive disease in the Caucasian population.<sup>1</sup> Mutations in the cystic fibrosis transmembrane regulator (CFTR) gene lead to malfunctioning or absent CFTR proteins, impairing mucosal clearance mechanisms. As such, CF is characterised by excessive viscous secretions in almost all organs, particularly the lungs, resulting in recurring infections, inflammation, airflow obstruction, and ultimately progressive functional decline. Whilst there remains no cure, advances in the treatment for patients with CF have resulted in an increased median life expectancy from 8 years in 1974, to 31 years in 2005 and 41 years in 2012.<sup>2</sup>

Whilst physical activity (PA) has been associated with numerous physiological and psychosocial benefits for healthy children,<sup>3</sup> there are additional health benefits for patients with CF. These include slower lung function decline,<sup>4</sup> reduced hospital admissions,<sup>5</sup> improved quality of life and nutritional status,<sup>6</sup> improved bone mineral density,<sup>7</sup> and enhanced airway clearance,<sup>8</sup> and ion channel function, which could lead to improved mucus hydration and clearance,<sup>9</sup> PA could be imperative for ultimate survival in patients with severe lung deterioration, given the strong positive relationship with aerobic capacity.<sup>10,11</sup> However, recent research suggests that as little as 2.1% of children with CF<sup>12</sup> meet the government PA guidelines of at least 60 minutes of moderate-to-vigorous-intensity physical activity (MVPA) every day.<sup>13</sup> Whilst regular PA is increasingly important in the care of patients with CF,<sup>14</sup> there is a dearth of research and indeed little consensus on physical activity levels (PAL) in children and adolescents with CF. Moreover, little is known as to whether beneficial outcomes may be achieved with engagement of PA at different intensities, which would be critical information for interventions and on going care.

Variations in PAL reported in the existing literature may be due to methodological inconsistencies. Earlier research employing self-reported measures found that children aged 7-17 years with CF participated in less very strenuous (> 6 METs) PA relative to healthy controls, even when patients had well-preserved lung function. PA relative to healthy controls, even when patients had well-preserved lung function. Conversely, Selvadurai and colleagues reported no significant differences between CF patients and age- and sexmatched controls in similarly-aged children (9-17 years), using uniaxial accelerometry. Advancing previous research, which only reported total counts, Aznar et al. utilised Evenson to find that 6-17 year old children with CF engaged in significantly less MVPA and vigorous physical activity (VPA) but demonstrated higher total and light physical activity (TPA and LPA, respectively). Yet Jantzen et al. found similar PAL in CF patients across the age and intensity spectrum, but less engagement in strenuous activities for schoolaged children (6-13 years) compared to healthy controls. Interestingly, when extreme values were removed, no relationship was present between strenuous PA and percentage predicted FEV<sub>1</sub>. 16

A potential limitation of earlier studies is the lack of age- and population-specific cut-points, although, in healthy populations, it is pertinent to note that Trost and colleagues<sup>17</sup> supported the use of Evenson cut-points. Arguably, the relative intensity for children and adolescents with CF could be greater and therefore light physical activity may be more beneficial to their health in comparison to their healthy counterparts. However, with the exception of Aznar and colleagues,<sup>12</sup> and to some extent Jantzen et al.,<sup>16</sup> the majority of studies did not consider PA across the spectrum.<sup>6,10,18</sup> As such, the identification of patients with CF participating in more LPA and TPA<sup>12</sup> may warrant further investigation. Specifically, previous research has suggested that time spent in low-light physical activity (low-LPA) and high-light physical activity (high-LPA) may have some favourable independent health benefits.<sup>19</sup> Additionally, a

sedentary lifestyle has been shown to contribute to the progression of both functional and physical impairment in CF populations,<sup>20</sup> yet little research has objectively assessed time spent being sedentary, nor the accumulation of PA or sedentary time. Indeed, the majority of physical activity research to date has focused on the total volume of PA rather than the manner in which this activity is accumulated with regards to bout frequency and duration. Gabel et al.<sup>21</sup> reported sedentary bouts of  $\geq$ 5 minutes to be detrimentally associated with C-reactive protein in healthy children, whereas PA bouts of  $\geq$ 1 minute, which have previously been used to identify sporadic bouts of PA, are reported to be associated with lower BMI.<sup>22</sup> Identifying patterns of accumulation in youth with CF, and those patterns that may be associated with functional gains, is important for advancing the design and evaluation of future interventions in this population.

In order for effective interventions aimed at improving PAL in children and adolescents to be developed, it is important to further understand current levels, intensities and accumulation of PA children and adolescents with and without CF. Therefore, the purpose of this study was to investigate PA and sedentary time patterns of children and adolescents with CF, in comparison to age- and sex-matched healthy controls. Furthermore, the study sought to ascertain whether such parameters could predict lung function. We hypothesised that PA intensity and duration would be significantly lower in patients with CF and be a significant predictor of disease severity (i.e., lung function).

#### Methods

- *Participants*
- 86 In total, 36 participants (12.6  $\pm$  2.7 years; 18 CF) were invited to take part in the study.
- Properties are shown in Table 1. Eighteen patients (10 boys) with mild-to-

moderate CF, confirmed by a sweat chloride  $> 60 \text{ mmol} \cdot 1^{-1}$  and genotyping (8  $\Delta$ F508 Homozygote, 10  $\Delta$ F508 Heterzygote; 4 CF-related liver disease) were recruited from an outpatient CF clinic in South Wales (United Kingdom). Patients were included in the study if they were aged 6 – 17 years old, had no increase in symptoms or weight loss two weeks prior to testing, and had a stable lung function (within 10% of best in the preceding six months); unstable non-pulmonary comorbidities or acute infections warranted exclusion. Eighteen age-and sex-matched non-clinical children were recruited from local schools to act as a healthy comparison group. Ethical approval was granted by the Bromley NHS research ethics committee (REC reference: 13/LO/1907) and written informed consent and assent were obtained from parents/guardians and patients, respectively. All patients were instructed to continue prescribed medications as usual throughout the duration of their study involvement.

#### Measurements

At their routine visits to the clinic, participants forced vital capacity (FVC) and forced expiratory volume in 1s (FEV1) were assessed using flow-volume loop spirometry (Vitalograph, UK)). The best of three consistent exhalations (<5% variability) was recorded. All lung function measurements were expressed as a percentage predicted normal, using appropriate reference data.<sup>23</sup> Furthermore, body mass (Seca 220; Hamburg, Germany), stature and sitting stature (Seca 220; Hamburg, Germany) were measured to the nearest 0.01 kg and 0.01 m, respectively. Waist circumference was measured to the nearest 0.01 m using a non-elastic anthropometric tape (Seca Ltd., Birmingham, UK) at the narrowest point between the bottom of the ribs and the iliac crest. Healthy age- and sex-matched counterparts were asked to attend one laboratory session at Swansea University for all measurements to be undertaken. All participants were provided with a hip-mounted ActiGraph GT3X+

accelerometer (ActiGraph LLC, Pensacola, FL) to assess habitual PAL over seven consecutive days.

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ActiGraph monitors, shown to have acceptable reliability and validity in paediatric populations,<sup>24</sup> sampled raw data at 100Hz. Data were downloaded using ActiLife software (v6.10.4; ActiGraph, Pensacola, FL), processed into 15s epochs and reduced using a customised Excel macro. Sustained periods of 20 minutes of consecutive zero's were used to define non-wear time, which has been found to result in an almost identical wear time and a smaller difference between sedentary time and sitting time estimates (assessed using activPAL; PAL Technologies, Glasgow, Scotland) compared with a 60 minute definition in children.<sup>21</sup> Sedentary time was defined as <100 counts·min<sup>-1</sup>, shown to be a good estimate of free-living sitting.<sup>25</sup> Time spent in MPA (4-5.99 METs) and VPA (≥6 METs) was determined using age-specific cut-points, 26 which demonstrated comparable accuracy to Evenson cutpoints.<sup>17</sup> A threshold of 4 METs was used to define MPA, as brisk walking has been associated with this energy cost in calibration studies.<sup>27,28</sup> MPA and VPA were summed to create MVPA. The rest of the time was classified as either low light-intensity physical activity (low-LPA; 100-799 counts·min<sup>-1</sup>) or high light-intensity physical activity (high-LPA; 800-<4 METs). The 800 counts·min<sup>-1</sup> threshold was selected as this published sedentary cut-point captures both sedentary time and static light-intensity activities such as standing, 25 and has been found to have differential associations with cardiometabolic biomarkers in adolescents. 19 A valid day was defined as ≥9 hours · day 1, which has been previously used in clinical populations.<sup>29</sup> To be included in the analyses, children were required to have worn the ActiGraph for at least three days, which has been shown to have a reliability coefficient of 0.7.30 PAL are reported for overall, weekday and weekend days separately.

Patterns of sedentary time and PA accumulation were also calculated. Breaks in sedentary time were defined as the number of times that the accelerometer exceeded 25 counts per 15s epoch following a 15s epoch of <25 counts per epoch.<sup>31</sup> The frequency and duration of time spent in sedentary (≥5 min),<sup>21</sup> and low-LPA, high-LPA, MPA and VPA (≥1 minutes) were also determined.<sup>22</sup> No interruptions to these bouts were permitted.

# Data Analysis

Gaussian distribution was confirmed by the Shapiro-Wilks test. Following this, the participant groups and weekday vs. weekend day were compared using a multivariate ANCOVA with group as a fixed factor and day as a repeated measure, controlling for wear time. A stepwise linear regression was used to analyse the association between FEV<sub>1</sub> and PA intensity levels and patterns, adjusting for predefined potential confounders (age, sex, stature, mass and wear time). To explore differences between the groups in terms of those that met current government guidelines for PA (i.e., average of  $\geq$ 60 minutes of MVPA/day), a Chisquare test was used. All statistical analyses were conducted using PASW Statistics 21 (SPSS, Chicago, IL). All data are presented as means  $\pm$  SD. Statistical significance was accepted when P  $\leq$  0.05.

# Results

No significant differences were observed between boys and girls with regards to anthropometrics or lung function, with the exception of maturity offset, which was significantly greater in boys (Table 1). Consequently, all data were pooled for subsequent analyses. The healthy and CF groups did not differ in anthropometrical characteristics.

However, those with CF presented with a significantly lower percentage of predicted FEV<sub>1</sub> 161 and FEV:FVC when described in both absolute and relative to predicted terms. 162 163 A total of four (2 healthy controls; 2 patients with CF) participants did not fulfil the wear 164 time criteria for valid accelerometry data and were therefore excluded from further analyses. 165 Those excluded did not differ in anthropometrics or lung function to those retained. Overall, 166 participants achieved  $4.5 \pm 1.2$  and  $1.8 \pm 0.6$  valid weekdays and weekend days, respectively. 167 168 169 CF patients and healthy controls engaged in similar levels of PA across the intensity spectrum, irrespective of whether weekday, weekend day or overall days were considered 170 (Table 2). There was a trend for greater time spent in LPA in CF patients (222.7  $\pm$  12.8 vs. 171  $207.3 \pm 12.4$  mins; P > 0.05), although this failed to reach significance. There were 172 significant differences between weekday and weekend day PA with regards to total LPA 173  $(229.3 \pm 52.4 \text{ vs. } 203.8 \pm 50.6 \text{ mins, respectively; } P < 0.05), MPA (45.1 \pm 21.5 \text{ vs. } 36.6 \pm 1.0 \text{ mins, respectively; } P < 0.05)$ 174 27.9 mins, respectively; P < 0.05) and MVPA (62.4  $\pm$  32.1 vs. 51.2  $\pm$  39.9 mins, respectively; 175 P < 0.05), with greater levels of activity achieved during weekdays than weekend days in 176 both groups. 177 178 Overall, 44.4% (n=8) vs. 38.9% (n=7) in the healthy and CF groups, respectively, met the 179 180 current guidelines for MVPA. Fewer children met the guidelines on weekend days (44.4% vs. 30.6%; P < 0.05). The percentage meeting government guidelines did not differ between CF 181 and healthy children during week or weekend days. 182 183 Healthy controls and CF patients demonstrated similar patterns of physical activity 184 accumulation (Table 3). However, different patterns were evident during weekday and 185

weekend days, with weekdays characterised by a greater frequency and duration of LPA and MPA bouts and a lower duration of sedentary bouts compared to weekend days.

Linear regression revealed that FEV<sub>1</sub> was predicted by height and LPA when both groups were pooled for analysis ( $F_{(2,31)} = 62.93$ , P < 0.001;  $R^2 = 0.80$ ). More specifically, when LPA was split into low-LPA and high-LPA, height and low-LPA significantly predicted FEV<sub>1</sub> ( $F_{(2,31)} = 68.07$ , P < 0.001;  $R^2 = 0.82$ ). When the groups were considered independently, the intensity of LPA that predicted FEV<sub>1</sub> differed, with FEV<sub>1</sub> predicted by height and high-LPA in CF patients ( $F_{(2,14)} = 79.60$ , P < 0.001;  $R^2 = 0.92$ ) compared to height and low-LPA in healthy controls ( $F_{(2,14)} = 24.31$ , P < 0.001;  $R^2 = 0.78$ ).

# Discussion

Children with CF and age- and sex-matched healthy controls did not differ in overall PAL or the pattern in which these levels were accrued. Interestingly, despite these similarities, FEV<sub>1</sub> was dependent on LPA levels in both CF patients and their healthy counterparts, although the intensity within LPA differed across the groups. Finally, we observed significant decreases in PAL during weekends, with increased sedentary time and decreased frequency and duration of LPA and MPA bouts, irrespective of disease status.

In agreement with some, <sup>6,18,32</sup> but not all, <sup>10,12</sup> previous studies, no significant difference was observed in the PAL of children with and without CF, although a considerably higher proportion of our CF population met recommended guidelines compared to previous research. <sup>12</sup> Given the numerous additional health benefits for patients with CF, <sup>4-9</sup> over and above the physiological and psychosocial benefits of regular PA identified in healthy children, <sup>3</sup> these findings highlight the need for strategies to increase PA in this population.

Indeed, the importance of PA has been recognised by the European Cystic Fibrosis Society (ECFS) and recent Cochrane Reviews, <sup>14,33</sup> which advocate the cost-effectiveness and beneficial effects of PA for promoting quality of life in patients with CF. However, information regarding PA behaviours in CF is limited and although PA as a treatment is becoming increasingly valued by CF clinical teams, <sup>34</sup> it remains underutilized in routine CF management <sup>35</sup>. Furthermore, there is a paucity of evidence-based guidance regarding the optimal combination of intensity and duration to elicit health benefits.

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Further controversy surrounds the relationship between CF and the intensity of PA undertaken, including with regards to the direction of causality. In earlier studies, Nixon et al. 10 suggested that, even when lung function was preserved, children with CF engaged in significantly less VPA relative to healthy peers, whereas Selvadurai et al.<sup>6</sup> and Britto et al.<sup>18</sup> found no differences in the intensity undertaken, with Britto et al. 18 reporting VPA participation to decline with age irrespective of disease status or severity. In contrast to the present findings, Aznar et al.<sup>12</sup> and Jantzen et al.<sup>32</sup> have previously reported lower total daily VPA in children with CF. Moreover, Aznar et al. 12 also found a greater engagement in daily TPA and LPA, the latter in agreement with the current study. Whilst the reason(s) for this lack of consensus are likely to be multi-faceted, certain methodological differences should be noted. Specifically, whilst a similar age range has been used in the majority of studies, <sup>6,12,18,32</sup> pooling of data from boys and girls 12,32 and a failure to account for maturity 16,18,32 or disease severity<sup>12,18</sup> limits further inter-study comparisons. Indeed, Selvadurai et al.<sup>6</sup> reported significant influences of maturity and sex on PAL in those with CF and their healthy counterparts. Caution is required when interpreting the PAL reported in previous studies that have used long measurement epochs<sup>12,16</sup> or questionnaires, <sup>6,10</sup> with concerns raised regarding the validity of questionnaire-derived PA estimates in chronic conditions such as CF, 16,36

which are susceptible to several forms of bias. In light of the highly sporadic nature of children's PA,<sup>37,38</sup> with the median duration of high-intensity bouts suggested to be only 3s and 95% lasting less than 15s,<sup>38,39</sup> the use of 15s epochs in this and previous studies may have influenced the findings, with VPA potentially miscategorised as MPA. Whilst the present study utilised this method to increase inter-study comparability of the results, future studies are suggested to use 1s epochs in accord with recommendations for the accurate assessment of PA intensity.<sup>40</sup>

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Alternatively, or additionally, discrepancies between accelerometry studies may be related to the cut-points used to delineate activity intensities. As there is a lack of age- and populationspecific cut-points developed and validated for CF populations, each study has utilised different cut-points, which has implications in the estimation of the time spent in different activities. 41 The impact of cut-point selection may be especially relevant in clinical populations in whom it could be argued that the relative intensity of a given count rate is higher than in their healthy counterparts. Whilst emphasising the need for disease-specific cut-points to be developed, this notion also highlights that the higher LPA reported here and elsewhere in CF children may be clinically meaningful. Indeed, it has previously been reported that time spent in low-LPA and high-LPA may have some favourable independent health benefits<sup>19</sup> but the minimum PA intensity and volume required to confer health benefits remains to be elucidated. The present study further supports the potential importance of low-LPA and high-LPA by demonstrating these factors to significantly predict lung function (FEV<sub>1</sub>) in healthy and CF children, respectively. Further work is warranted to investigate whether targeting increases in low-LPA and high-LPA rather than increases in MVPA per se, may have beneficial health outcomes in this population, particularly given the high correlation between LPA and sedentary time <sup>19</sup>. Increasing LPA through interventions may be a more feasible and constructive first step for the large proportion of patients not meeting current PA guidelines.<sup>19</sup>

Despite the increasing attention on sedentary behaviour as an independent risk factor for cardiometabolic disease in children and youth, 42 there is a lack of data regarding sedentary behaviours in the CF population. In accord with Aznar et al., 12 we found no difference in the time spent sedentary by children with CF and their healthy counterparts. Whilst not the focus of the present study, no relationships were found between sedentary behaviour and disease severity, although the limited sample size should be considered when interpreting these findings. Future studies should explore the potential relationship and interactions between sedentary behaviour, PA and health in CF patients using objective measures and novel statistical approaches to allow the optimal combination of these independent factors to be identified. Indeed, a growing body of evidence in healthy children suggests that the specific type of sedentary behaviour (e.g., television viewing, computer use), rather than being sedentary per se, may be an important determinant of health. 43,44

Emerging evidence suggests that the pattern in which PAL and sedentary time are accrued may be an important determinant with regards to health. In healthy children, sedentary bouts have been associated with C-reactive protein<sup>21</sup> and HDL cholesterol.<sup>45</sup> However, in contrast, Carson and Janssen<sup>46</sup> found that patterns of sedentary behavior were not related to cardiometabolic risk factors in 6-19 year olds. Therefore, whether differences in the pattern of sedentary time and PA have implications for health, particularly when TPA is similar, remains to be resolved. The present study revealed no significant differences between the groups with regards to the frequency or duration of sedentary or PA bouts, although there was a trend for longer high-LPA bouts in the CF children. We did, however, observe significant

differences in the pattern of PA and sedentary behaviours during weekdays and weekend days, which were similar across the groups. Specifically, weekend days were characterized by greater time spent sedentary with a lower frequency and duration of LPA and MPA bouts. Since children potentially have more control over weekend free-time, it could be postulated that intra-individual differences may be most evident on weekend days.<sup>47</sup> Indeed, the greater PAL during weekdays may, at least in part, be attributable to participation in Physical Education lessons and/or extra mural sports teams, although the effect of such isolated events is likely to be minimal across seven days of objective PA assessment. Nonetheless, these findings highlight the importance of considering different strategies to target week and weekend day PA promotion.

Although the present study had numerous strengths, such as the objective measurement of PA, precisely matched healthy counterparts, and the novel consideration of the pattern in which PA is accrued in those with CF, it is important to note certain limitations. Firstly, the sample size was limited and, consequently, as was the range of disease severities included, although relative to the overall CF population, we believe that the present results provide relevant and generalizable conclusions. Given the small sample size, the results of the present linear regression should be considered exploratory; larger studies looking at the patterning of PA across the disease spectrum would be invaluable in the future. It is pertinent to note that whilst three or more days of valid PA data were required for inclusion in the analyses, no stipulations were made regarding the breakdown of these days between week and weekend days. Given that PA is suggested to differ between weekdays and weekends in healthy<sup>48,49</sup> and CF youth<sup>12</sup> this may have influenced the current findings. The integration of postural assessment may have provided greater insights into specific sedentary behaviours, such as sitting. Furthermore, the lack of consistency in how bouts are defined (i.e., bout and

interruption length) limits cross-study comparisons;<sup>50</sup> the durations utilised in the present study were informed by research in healthy populations regarding bout and interruption durations.<sup>21,22</sup> Finally, the cross-sectional design of the present study also limits the ability to make casual inferences regarding the relationships and their directionality.

#### **Conclusions**

In conclusion, the present study has demonstrated that there are no differences between CF children and age- and sex-matched healthy controls with regards to overall PAL or the manner in which these intensities are accrued, with significantly lower PA and greater sedentary behaviours during the weekend. Furthermore, the present study found LPA to be a significant predictor of lung function in both healthy children and those with CF, although the relevant intensity of LPA differed with high-LPA most important in those with CF. These findings therefore highlight weekends and LPA as areas warranting further research for the development of effective intervention strategies to increase PA in the youth CF population.

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460 Tables

**Table 1.** Participant characteristics

Total	Cystic Fibrosis	Controls
36	18	18
$12.6 \pm 2.7$	$12.4 \pm 2.8$	$12.5 \pm 2.7$
$1.48 \pm 0.14$	$1.46 \pm 0.14$	$1.51 \pm 0.13$
44.24 ± 12.99	$41.16 \pm 12.51$	$47.52 \pm 13.04$
$0.67 \pm 0.08$	$0.66 \pm 0.07$	$0.67 \pm 0.09$
$19.6 \pm 3.4$	$18.8 \pm 2.8$	$20.5 \pm 3.8$
$-1.28 \pm 3.00$	$-1.04 \pm 2.42$	$-1.54 \pm 3.57$
$84 \pm 15$	$83 \pm 12$	$85 \pm 18$
$85 \pm 14$	$80 \pm 9$	89 ± 17*
	$36$ $12.6 \pm 2.7$ $1.48 \pm 0.14$ $44.24 \pm 12.99$ $0.67 \pm 0.08$ $19.6 \pm 3.4$ $-1.28 \pm 3.00$ $84 \pm 15$	$36$ $18$ $12.6 \pm 2.7$ $12.4 \pm 2.8$ $1.48 \pm 0.14$ $1.46 \pm 0.14$ $44.24 \pm 12.99$ $41.16 \pm 12.51$ $0.67 \pm 0.08$ $0.66 \pm 0.07$ $19.6 \pm 3.4$ $18.8 \pm 2.8$ $-1.28 \pm 3.00$ $-1.04 \pm 2.42$ $84 \pm 15$ $83 \pm 12$

Mean ± S.D. PHV, peak height velocity; FVC, forced vital capacity; FEV<sub>1</sub>, forced expiratory volume

in 1 second. \* significant difference between control and Cystic Fibrosis

Table 2. Physical activity data by group

	Total	CF	Control
	n = 32	n = 16	$n = \frac{16}{1}$
Overall			
Sedentary time (min·day <sup>-1</sup> )	$545.4 \pm 76.0$	$539.2 \pm 64.6$	$551.3 \pm 87.0$
Low-LPA (min·day-1)	$141.4 \pm 34.9$	$144.3 \pm 30.9$	$138.7 \pm 39.1$
High-LPA (min·day¹)	$72.5 \pm 23.9$	$77.5 \pm 20.6$	$67.7 \pm 26.3$
MPA (min·day <sup>-1</sup> )	$40.5 \pm 22.6$	$39.5 \pm 23.3$	$41.3 \pm 22.6$
VPA (min·day-1)	$12.6 \pm 10.6$	$13.1 \pm 12.4$	$12.1 \pm 9.0$
VVPA (min·day <sup>-1</sup> )	$2.7 \pm 3.4$	$2.7 \pm 3.9$	$2.7 \pm 2.9$
MVPA (min·day <sup>-1</sup> )	$55.7 \pm 32.7$	$55.3 \pm 38.0$	$56.1 \pm 27.9$
Week days			
Sedentary time (min·day <sup>-1</sup> )	$542.8 \pm 84.1$	$532.1 \pm 69.2$	$552.3 \pm 96.5$
Low-LPA (min·day-1)	$146.2 \pm 36.1$	$149.7 \pm 27.0$	$143.2 \pm 43.2$
High-LPA (min·day <sup>-1</sup> )	$76.9 \pm 26.3$	$83.8 \pm 22.0$	$70.8 \pm 29.0$
MPA (min·day <sup>-1</sup> )	$43.8 \pm 22.0$	$43.7 \pm 22.8$	$43.8 \pm 21.9$
VPA (min·day <sup>-1</sup> )	$13.7 \pm 11.2$	$14.7 \pm 12.7$	$12.8 \pm 10.0$
VVPA (min·day <sup>-1</sup> )	$2.7 \pm 3.3$	$3.0 \pm 4.0$	$2.4 \pm 2.5$
MVPA (min·day <sup>-1</sup> )	$60.2 \pm 32.4$	$61.5 \pm 38.0$	$59.0 \pm 27.6$
Weekend days			
Sedentary time (min·day <sup>-1</sup> )	$555.7 \pm 91.3$	$554.9 \pm 94.5$	$556.5 \pm 90.1$
Low-LPA (min·day-1)	$135.7 \pm 35.7$	$140.3 \pm 38.6$	$130.9 \pm 32.9$
High-LPA (min·day <sup>-1</sup> )	$66.0 \pm 22.7$	$69.2 \pm 22.4$	$62.6 \pm 23.1$
MPA (min·day <sup>-1</sup> )	$36.0 \pm 27.7$	$34.5 \pm 27.1^*$	$37.6 \pm 29.1^*$
VPA (min·day <sup>-1</sup> )	$11.1 \pm 12.5$	$11.3 \pm 14.0$	$11.0 \pm 11.1$

# Physical Activity Levels in Cystic Fibrosis Youth

	VVPA (min·day <sup>-1</sup> )	$3.1 \pm 5.7$	$2.4 \pm 3.8$	$3.8 \pm 7.2$
	MVPA (min·day-1)	$50.3 \pm 39.6$	$48.2 \pm 41.4^*$	$52.5 \pm 38.9^*$
467	Means ± SD. Low-LPA, low ligh	nt physical activity; Hig	h-LPA, high light physi	cal activity; MPA,
468	moderate physical activity; VPA,	vigorous physical activi	ty; VVPA, very vigorou	s physical activity;
469	MVPA, moderate-to-vigorous phy	ysical activity. * Significa	ant difference between w	veek- and weekend
470	day within group			
471				
472				
473				
474				
475				

Table 3. Patterns of PA accumulation on week days, weekend days and overall (average day)

	Total	CF	Control
	n=32	n = 16	n = 16
Overall			
Frequency SED	$28 \pm 7$	27 ± 7	$28 \pm 7$
<b>Duration SED (mins)</b>	$271.2 \pm 83.5$	$263.7 \pm 86.2$	$278.3 \pm 82.6$
Number SED Breaks	$301.3 \pm 57.2$	$303.6 \pm 56.2$	$299.0 \pm 59.7$
Frequency LPA	$62 \pm 18$	$66 \pm 16$	$59 \pm 20$
<b>Duration LPA (mins)</b>	$98.4 \pm 33.7$	$105.5 \pm 28.7$	$91.6 \pm 37.3$
Frequency Low-LPA	$22 \pm 8$	23 ± 6	21 ± 9
<b>Duration Low-LPA (mins)</b>	$27.0 \pm 9.8$	$28.2 \pm 7.3$	$26.0 \pm 11.8$
Frequency High-LPA	9 ± 7	11 ± 7	$8 \pm 8$
<b>Duration High-LPA (mins)</b>	$13.9 \pm 14.1$	$16.6 \pm 14.7$	$11.3 \pm 13.3$
Frequency MPA	$7 \pm 4$	7 ± 4	$7 \pm 5$
<b>Duration MPA (mins)</b>	$11.8 \pm 12.1$	$10.1 \pm 6.3$	$13.3 \pm 15.8$
Frequency VPA	$3\pm3$	$3 \pm 4$	$3\pm3$
<b>Duration VPA (mins)</b>	$5.2 \pm 5.9$	$5.3 \pm 7.4$	$5.1 \pm 4.1$
Weekdays			
Frequency SED	$27 \pm 8$	$26 \pm 7$	$28 \pm 9$
<b>Duration SED (mins)</b>	$262.4 \pm 88.8$	$251.6 \pm 88.3$	$272.1 \pm 90.6$
Number SED Breaks (mins)	$309.0 \pm 54.8$	$311.8 \pm 45.4$	$306.6 \pm 63.2$
Frequency LPA	$65 \pm 21$	69 ± 17	$61 \pm 24$
<b>Duration LPA (mins)</b>	$103.5 \pm 39.6$	$112.1 \pm 32.1$	$95.8 \pm 44.7$
Frequency Low-LPA	$23 \pm 9$	23 ± 6	22 ± 11
Duration Low-LPA (mins)	$27.9 \pm 11.3$	$28.8 \pm 7.7$	$27.1 \pm 14.0$
Frequency High-LPA	$10 \pm 8$	12 ± 8	9 ± 9

Duration High-LPA (mins)	$15.7 \pm 17.7$	$18.8 \pm 19.6$	$12.9 \pm 15.8$
Frequency MPA	$8 \pm 4$	$8 \pm 4$	8 ± 5
Duration MPA (mins)	$13.0 \pm 12.0$	$11.4 \pm 6.3$	$14.4 \pm 15.5$
Frequency VPA	$3\pm3$	$3\pm4$	$3\pm3$
Duration VPA (mins)	$5.3 \pm 6.1$	$5.8 \pm 7.5$	$4.9 \pm 4.6$
Weekend days			
Frequency SED	$29 \pm 8$	$29 \pm 9$	$29 \pm 8$
<b>Duration SED (mins)</b>	$286.8 \pm 97.1$	$278.4 \pm 108.6~^*$	$295.7 \pm 85.8$ *
Number SED Breaks	$293.0 \pm 66.1$	$299.6 \pm 78.8$	$286.0 \pm 51.1$
Frequency LPA	59 ± 16	$62\pm17$ *	$56 \pm 15$ *
<b>Duration LPA (mins)</b>	$90.8 \pm 26.2$	$97.0\pm27.8$ $^{\#}$	$84.2 \pm 23.4$ #
Frequency Low-LPA	$21 \pm 8$	22 ± 8	$19 \pm 7$
Duration Low-LPA (mins)	$25.6 \pm 10.3$	$27.5 \pm 10.7$	$23.6 \pm 9.9$
Frequency High-LPA	8 ± 6	$9\pm6$ #	$6\pm6$ #
Duration High-LPA (mins)	$10.2 \pm 8.8$	12.7 $\pm$ 9.1 $^{\#}$	$6.1\pm6.0$ #
Frequency MPA	$6 \pm 6$	5 $\pm$ 5 $^{\#}$	$6\pm7$ $^{\#}$
Duration MPA (mins)	$9.8 \pm 14.7$	$8.3\pm9.8$ *	11.4 $\pm$ 18.8 $^{\ast}$
Frequency VPA	$3 \pm 4$	$3\pm4$	$3 \pm 4$
Duration VPA	$5.3 \pm 7.6$	$4.9 \pm 8.3$	$5.7 \pm 7.1$

Mean  $\pm$  SD. SED, sedentary behaviour; Low-LPA, low light physical activity; High-LPA, high light physical activity; LPA, light physical activity; MPA, moderate physical activity; VPA, vigorous physical activity. \* Significant difference within condition between weekday and weekend P < 0.05; \* Significant difference within condition between weekday and weekend P < 0.01