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Health-Related and Motor Performance-Related Fitness and Physical Activity Among Youth With Cystic Fibrosis

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Abstract

Little is known about motor competence and the longitudinal development of motor performance among youth with cystic fibrosis (CF). In this study, we assessed aspects of motor performance in different age groups of young patients with CF and compared them with a healthy reference group of same aged children. We also examined

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the development of motor performance among different age groups of these children with CF, using The Deutscher Motorik Test (DMT) to assess attributes of health-related and motor performance-related fitness. We used an incremental ergometer cycle test to determine maximal exercise capacity (expressed as peak workload). We evaluated and recorded habitual physical activity (PA) as measured by the number of steps per day and the time spent in different PA intensities (expressed in metabolic equivalents). In total, 31 children and adolescents with CF agreed to participate (13 girls,18 boys) aged 6–17 years ($M = 11.3$, $SD = 3.3$ years); they had a mean one second forced expiratory volume (expressed as a percentage of predicted value [% pred]) of 87.2% ($SD = 22.3\%$). We found their values of health-related and motor performance-related fitness to be significantly lower ($p < 0.05$) than those of their healthy peer participants. In contrast to the reference group, participants with CF up to 14 years of age showed a linear improvement in these values and in their PA, followed by a plateau or even a nonsignificant decrease after age 14. These findings have important implications for the development and prescription of exercise programs for children with CF. Besides aerobic and strength exercises, we recommend that neuromuscular training be integrated into exercise programs to improve the coordinative abilities of youth with CF. More attention should be paid to vulnerable older adolescents to ensure their long-term motivation to maintain exercise participation.

Keywords
CF, adolescence, middle childhood, Deutscher Motorik Test, expiratory volume, aerobic exercise

Introduction

Knowledge of the effects of physical activity (PA) and exercise as components of chronic disease treatment has improved over recent years (Pedersen & Saltin, 2015). Regular PA and exercise are now recommended aspects of routine care for cystic fibrosis (CF) (Swisher et al., 2015). PA and exercise have been shown to increase physical fitness, lung function, and quality of life and to decrease hospital stays, bone disease, and mortality (Gruber et al., 2014; Hebestreit et al., 2010; Radtke et al., 2017; Schneiderman-Walker et al., 2000; Swisher et al., 2015). Physical fitness can be described as a set of attributes related to the ability to perform PA. It includes two aspects: health-related fitness (cardiorespiratory endurance, muscular endurance, muscular strength, flexibility, and body composition) and motor performance-related fitness (power, speed, agility, coordination, reaction time, and balance) (Caspersen et al., 1985; Haga, 2008; Howley, 2001).

The level of motor performance is positively related to PA; thus, motor skill proficiency in childhood has seemed to predict PA habits in adolescence and adulthood (Barnett et al., 2009; Loprinzi et al., 2015; Utesch et al., 2019). Children and adolescents with lower levels of motor skills have preferred a more sedentary lifestyle and have avoided situations that demand motor skills (Barnett et al., 2016; Field & Temple, 2017). In addition, there has been a higher risk of injury among children with poor motor performance (Larsen et al., 2016). Appropriate motor skills and abilities are important for normal psychological, social and physical development in children and adolescents (Bremer & Cairney, 2018; Lima et al., 2017).

Among youth with CF, higher levels of exercise participation have led to better cardiorespiratory fitness, expressed as peak oxygen uptake (VO2peak), which is a strong predictor of mortality in this population (Hebestreit et al., 2019; Nixon et al., 1992; Vendrusculo et al., 2019). For this reason, training studies of individuals with CF have mostly focused on cardiorespiratory endurance and the improvement of VO2peak. In contrast, little is known about other aspects of health-related fitness among individuals with CF, particularly with respect to motor performance-related fitness as an important prerequisite for participation in sports and leisure-time PA (exercise, sport, dance, play and games) (Arikan et al., 2015; Corten & Morrow, 2020; Gruber et al., 2008, 2010).

In preschool children with CF, motor performance has been age appropriate or even superior to that of healthy preschool children (Gruber et al., 2010). As in healthy individuals, it can be assumed that better motor performance during childhood leads to an active lifestyle and exercise participation in older children and adolescents with CF. However, reduced levels of motor performance have been observed in school children and adolescents with CF. In addition, a significantly shorter walking distance than predicted in the 6-minute walk test has been reported in youth with CF (Arikan et al., 2015; Gruber et al., 2008). In a further study, motor delay or a risk of motor delay was observed in children with CF from 4 – 8 years of age (Corten & Morrow, 2020). Lung function has been positively correlated with some aspects of motor performance-related and health-related fitness and seems to be related to participation in sports and exercise (Arikan et al., 2015; Corten & Morrow, 2020; Gruber et al., 2008). No information is available from past research about exercise participation in youth with CF (Arikan et al., 2015; Corten & Morrow, 2020; Gruber et al., 2008, 2020). However, a recently published study showed similar moderate-to-vigorous PA and sedentary time between people with CF and healthy controls (Puppo et al., 2020). Thus, it is not clear whether disease-related causes and/or a lack of PA and exercise in daily life are the reasons for the lower health-related fitness and motor-performance related fitness values among people with CF.

Only one study has been published on the development of motor performance with age in the CF population (Gruber et al., 2008). Linear increases in cardiorespiratory endurance, flexibility, coordination, and power were seen only up to the age of 14 years, followed by only minor changes in adolescents aged 15–18 years. In contrast, flexibility as an attribute of health-related physical fitness was observed to worsen with increasing age among people with CF, in contrast with healthy individuals (Bös et al., 2008; Gruber et al., 2008).

On the basis of this prior literature on motor performance in CF (Arikan et al., 2015; Corten & Morrow, 2020; Gruber et al., 2008, 2010, 2020), we hypothesized that children and adolescents (aged 6–17 years) with CF would generally show lower levels of health-related and motor performance-related fitness. Moreover, we assumed that health-related and motor performance-related fitness and PA habits would not show a continuous increase among youth with CF during the period from middle childhood to late adolescence.

In the present study, we aimed to examine attributes of health-related and motor performance-related fitness in children and adolescents with CF aged 6–17 years and to compare them with these attributes in a healthy reference group of the same age. Our second aim in this study was to examine the progressive course of health-related and motor performance-related fitness in CF from middle childhood to late adolescence with consideration of the specific variables of peak workload (Wpeak, or W for watts), PA (in steps/day), and PA intensity (in metabolic equivalents [METs]).

Method

Participants

Data for this study were collected as part of the partially monitored exercise program CF*mobil*, conducted at the Christiane Herzog Centrum Ruhr (CHCR). The CHCR is a cooperative of three CF centers located in the Ruhr area of western Germany (University Children's Hospital Essen, University Children's Hospital Bochum, and Ruhrlandklinik Essen). The study was approved by the ethics committee of the University Duisburg-Essen (14-6117-BO) and is registered in ClinicalTrials.gov (NCT03518697).

In the present study, we only analyzed data from children and adolescents aged 6–17 years. The children with CF were in a stable condition, and we obtained written informed consent from their parents or guardians before they participated in the study. All tests were performed at baseline before the start of the 12-month partially monitored exercise program CFmobil. To compare motor competence development with respect to age, the participants were divided into three age groups (AG 1: 6–10 years, AG 2: 11–14 years, and AG 3: 15–17 years). These age groups were chosen to allow the comparison of these data with data from a previous study (Gruber et al., 2008).

Measures

Anthropometry and lung function. We measured body weight with an electronic flat scale (Seca 861; Seca, Hamburg, Germany) and determined height with a telescopic measuring rod (Seca 202; Seca, Hamburg, Germany), to the nearest 0.1 cm. We calculated the body mass index (BMI) as the child's weight divided by the square of their height (kg/m^2). Forced expiratory volume in 1 s (FEV1) and forced vital capacity were assessed according to the American Thoracic Society guidelines (Miller et al.,2005) using standard spirometric techniques (JAEGER MasterScreen Body; CareFusion, Höchberg, Germany) and expressed as the percentage of predicted value (% pred).

Exercise testing and motor performance. All participants performed an incremental cycling test on an electromagnetically braked cycle ergometer (Ergoselect 100 P; Ergoline GmbH, Bitz, Germany) in an upright position according to the Godfrey protocol (Godfrey et al., 1971). The exercise test was monitored by an experienced physician. Heart rate was continuously monitored with a heart rate monitor and a chest strap (Polar A300; Polar, Kempele, Finland). Peak exercise capacity was expressed as Wpeak and peak heart rate, which were taken as the highest values in the last 30 seconds before stopping the test. The participants were encouraged to exert maximal effort. The test was continued until the participant could no longer maintain a pedaling cadence of 60 rpm, when a rating of 9–10 on a 0–10 Borg scale for perceived exertion was reached, or when oxygen saturation (SpO2) dropped to <85%. The tests were accepted as truly maximal if any of the following criteria were met: the participant achieved the predicted maximal work rate or reached a maximal heart rate at or above the predicted peak heart rate (Hebestreit et al., 2015).

The % pred values were determined from Godfrey's reference equations for workload (Godfrey & Mearns, 1971). The Deutscher Motorik Test (DMT) was used to assess motor performance. This test has been used in large cross-sectional and longitudinal studies in Germany, and reference values are available for children and adolescents aged 6–18 years (Bös et al., 2008, 2016; Krug et al., 2019). The DMT has been shown to be valid in children and adolescents. The test–retest reliability of all test items has been shown to range from .73 to .96 (average .82). The coefficient of objectivity has been shown to range from .87 to .99, with a mean of .95 for all test tasks (Bös et al., 2016).

The DMT was conducted by a specialized and experienced sport student. All patients received the same instructions according to the specifications in the test manual (Bös et al., 2016). The test items were as follows: strength endurance (push-ups for 40 seconds, sit-ups for 40 seconds), explosive power (standing long jump), coordination under time pressure (jumping side to side for 15 seconds), coordination for the precision aspect (balancing backward on beams with a length of three meters and different widths), and trunk flexibility (forward bend). A cohort of German and Austrian school children and adolescents from the nationwide Motorik–Modul Baseline Study (n = 4376, age 6–17 years) served as the reference group (Bös et al., 2019). Care was taken to ensure that all tests were performed at the same time of day.

Physical activity. PA was recorded using an accelerometer (wActiSleep-BT Monitor; ActiGraph Corp., Pensacola, FL, USA) over a period of four weeks before testing motor performance and maximal exercise capacity. The participants decided on where they would wear the accelerometer (wrist of the non-dominant hand, hip, or ankle). All participants decided to wear the accelerometer on the wrist. The place where the device was worn was specified during initialization of the accelerometer and was considered in the data analysis.

PA was assessed as the number of steps per day. PA intensity was expressed in METs, and PA was categorized as sedentary (<1.5 METs), light (1.5–2.9 METs), moderate to vigorous (3–5.9 METs), and vigorous (>6 METs), according to the national recommendations for PA and PA promotion in Germany (Pfeifer & Rutten, 2017).

Statistical Analysis

Descriptive statistics (means and standard deviations) were calculated for all variables, and the data were assessed for normal distribution. The data of youth with CF and the healthy peer group were analyzed using analysis of variance (ANOVA). For non-normally distributed data, we performed the Mann–Whitney U-test and the Kruskal–Wallis test.

Raw data from healthy controls (reference data) were downloaded from the MO|RE Data website (https://www.sport.kit.edu/more/index.php) (Bös et al., 2019). MO|RE Data is a project supported by the Deutsche Forschungsgemeinschaft/German Research Foundation, with the aim of building a database for motoric data and generating high-quality standard data for research (Albrecht et al., 2016). Correlations were calculated using the Spearman rank test to analyze the relationship among age, lung function, workload (Wpeak), PA (steps/day), and PA intensity in the DMT test items. All statistical analyses were performed using SPSS (version 22.0; SPSS Inc., Chicago, IL, USA). Differences were considered statistically significant at $p \leq 0.05$.

Results

A total of 31 children and adolescents with CF (18 boys and 13 girls) aged 6–17 years (M age = 11.5 years, $SD = 3.3$) participated. The anthropometric data of the youth with CF and the healthy reference group are summarized in Table 1. Lung function and Wpeak were determined only for youth with CF.

The ANOVA test showed nonsignificant differences in anthropometric data between children with CF and healthy controls ($p > .05$). However, healthy

Table 1. Anthropometric Characteristics and Lung Function of Children and Adolescents With CF and the Healthy Reference Group.

Anthropometric characteristics	CF			Healthy		
	Middle childhood (6–10 years)	Late childhood/ early adolescence (11–14 years)	Late adolescence (15–17 years)	Middle childhood (6–10 years)	Late childhood/ early adolescence (11–14 years)	Late adolescence (15–17 years)
	Mean (SD)	Mean (SD)	Mean (SD)	Mean (SD)	Mean (SD)	Mean (SD)
Age (years)	8.2 (1.7) (12)	12.4 (1.4) (12)	16.3 (0.8) (6)	8.3 (1.3) (2646)	12.3 (01.1) (1522)	15.7 (0.9) (527)
Height (cm)	126.3 (10.0) (12)	150.1 (12.4) (12)	172.4 (9.1) (6)	136.4 (10.0) (2646)	158.1 (10.4) (1522)	173.3 (10.2) (527)
Weight (kg)	25.4 (5.2) (12)	40.2 (9.1) (12)	61.3 (15.1) (6)	32.4 (8.7) (2646)	47.7 (11.6) (1522)	64.2 (12.3) (527)
BMI	15.7 (1.3) (12)	17.7 (2.0) (12)	20.3 (3.1) (6)	17.2 (3.0) (2646)	18.9 (3.2) (1522)	21.3 (3.0) (527)
FEV1 (% pred)	88.2 (17.3) (12)	84.6 (24.2) (12)	77.6 (30.7) (6)			
Wpeak (% pred)	89.3 (29.2) (12)	94.4 (22.0) (12)	91.2 (21.8) (12)			

Note. SD=standard deviation. Between groups: ANOVA, $^{*}p < .05$, $^{**}p < .01$, $^{***}p < .001$. Values in brackets represent the number of children and adolescents tested. Lung function and peak workload data are available only for youth with CF.

controls were taller and heavier and had a higher BMI. The differences between the parameters decreased with increasing age and were almost identical among the adolescent groups. FEV1 and Wpeak did not significantly differ ($p > .05$) among the CF age groups.

Table 2 shows the results of the test items of the DMT. Youth with CF had lower values in all test items, which showed significant differences ($p < .05$), except for the forward bend. In healthy controls, the parameters of motor performance increased with age and showed significant differences among age groups ($p < .05$). In youth with CF, test items based on power or strength (push-ups and sit-ups) improved up to age 14 years, without statistical significance ($p > 0.05$). A further increase up to age 17 years was seen only in standing long jump. All other parameters (balancing backward, sideway jumping, forward bend, push-ups, and sit-ups) remained stable or decreased. According to performance classes, the DMT subtest scores ranged from below average to above average. In AG 3, the subtest results were below average.

Table 3 summarizes the PA (steps/day) and PA intensity of children with CF. A nonsignificant ($p > .05$) decrease was seen among age groups in steps/day and time spent in moderate-to-vigorous and vigorous PA. Sedentary time increased with age, and the highest amount of time spent was seen in the age group 11–14 years, without reaching significance ($p > .05$). The time spent in PA with light intensity decreased from AG 1 to AG 2 and increased in AG 3. The time spent in vigorous PA was higher in AG 2 than in AG 1 and AG 3, and the values in the latter two groups were almost identical. The results for the steps/day and PA intensities did not significantly differ among the groups ($p > .05$).

The results of the correlation analyses are presented in Table 4. Anthropometric data and maximal workload were influenced by age. Age showed significant positive effects on DMT test items based on strength and power ($p < .05$) but not on balance and forward bend ($p > .05$). Interestingly, age adversely affected PA. Younger children with CF were more engaged in moderate-to-vigorous PA and achieved more steps per day than adolescents ($p < .05$). In contrast, sedentary time was significantly higher in adolescents than in younger children with CF. FEV1 showed a significantly positive correlation ($p < .05$) with Wpeak but not with the DMT test items, PA, and steps/day or PA intensity ($p > .05$). A positive correlation ($p < .05$) with Wpeak was found for strength or power but not for forward bend and balance or for PA (steps and intensity).

Discussion

The major findings of the present study were that the values of parameters of health-related fitness and motor performance-related fitness of children and adolescents with CF were lower than those of healthy controls of the same age. The present results confirm and extend the findings of previous studies

Table 2. Comparison of DMT Results Between Children and Adolescents With CF and the Healthy Reference Group.

	CF			Healthy				
	Middle childhood (6–10 years)	Late childhood/ early adolescence (11–14 years)	Late adolescence (15–17 years)	Middle childhood (6–10 years)	Late childhood/ early adolescence (11–14 years)	Late adolescence (15–17 years)		
DMT items	Mean (SD)	Mean (SD)	Mean (SD)	Mean (SD)	Mean (SD)	Mean (SD)	Mann–Whitney U-test	Kruskal–Wallis test
Total backward balancing (steps)*	26.2 (8.8) [12]	24.5 (6.9) [13]	24.0 (10.3) [6]	31.8 (10.5)* [2646]	33.3(10.1)***‡‡ [1522]	38.7 (9.0)**‡‡‡ [527]	U = −4.795 $p < .001$	CF: $p = 0.727$, healthy: $p < .001$
Sideway jumping (# in 15 s)	17.4 (6.4) [12]	31.5 (9.0)‡‡‡ [13]	29.0 (4.2) [6]	29.0 (8.4)*** [2646]	37.5 (8.3)*‡‡‡ [1522]	42.7 (7.9)***‡‡‡ [527]	U = −3.832 $p < .001$	CF: $p < .001$, healthy: $p < .001$
Forward bend (cm)	−0.9 (10.3) [12]	−1.2 (11.4) [13]	−1.2 (7.7) [6]	1.0 (6.9) [2646]	1.6 (9.2)‡‡‡ [1522]	5.6 (9.4)**‡‡‡ [527]	U = −1.877 $p = .061$	CF: $p = .110$, healthy: $p < .001$
Push-ups (# in 40 s)	8.4 (2.9) [12]	13.4 (4.5)‡‡ [13]	12.3 (1.9) [6]	14.2 (5.4)*** [2646]	15.7 (4.6)*‡‡‡ [1522]	16.9 (4.3)**‡‡‡ [527]	U = −4.364 $p < .001$	CF: $p = .008$, healthy: $p < .001$
Sit-ups (# in 40 s)	12.4 (6.9) [12]	21.8 (6.9)‡‡ [13]	19.7 (2.1) [6]	18.8 (6.3)** [2646]	23.7 (6.6)‡‡‡ [1522]	25.8 (6.5)**‡‡‡ [527]	U=2.359 $p < .001$	CF: $p = .004$, healthy: $p < .001$
Standing long jump (cm)	113.5 (20.2) [12]	137.7 (37.2)‡ [13]	154.0 (43.7) [6]	130.0 (23.3)* [2646]	164.3 (29.5)**‡‡‡ [1522]	186.3 (37.3)*‡‡‡ [527]	U = −2.698 $p = .007$	CF: $p = .049$, healthy: $p < .001$

Note. SD = standard deviation, CF = Cystic Fibrosis, DMT= Deutscher Motorik test. Between groups: Mann–Whitney U-test, *$p < .05$, **$p < .01$, ***$p < .001$. Within CF and healthy: Kruskal–Wallis test, #$p < .05$, ##$p < .01$, ###$p < .001$. Between CF and healthy age groups: Mann–Whitney U-test, ‡$p < .05$,‡‡$p < .01$, ‡‡‡$p < .001$. Values in brackets represent the number of children and adolescents tested. *Sum of all steps on the different beams (with widths of 6.0 cm, 4.5 cm, and 3.0 cm) divided by 3.

Table 3. Steps and Intensity of Physical Activity Per Day in Different Age Groups.

Variables	Middle childhood (6–10 years) Mean (SD)	Late childhood/ early adolescence (11–14 years) Mean (SD)	Late adolescence (15–17 years) Mean (SD)	Kruskal–Wallis test *p*
Steps/day	12,576.8 (2776.8) [7502–16,629]	11,110.8 (1963.8) [8005–15,951]	9565.8 (2153.4) [7880–13,202]	.078
Sedentary time (<1.5 METs) in min	599.3 (186.2) [288–945]	770.3 (250.3) [256–1057]	748.9 (121) [542–849]	.304
Light intensity (1.5–2.9 METs) in min	419.3 (61.1) [330–517]	394.8 (224.5) [118–1073]	422.6 (171.2) [204–567]	.119
Moderate-to-vigorous intensity (3–5.9 METs) in min	187.7 (102.4) [76–363]	131.0 (99.2) [46–367]	98.6 (47.4) [43–166]	.647
Vigorous intensity (>6 METs) in min	14.7 (18.5) [0–57]	21.9 (20.5) [0–62]	15.4 (24.5) [0–56]	.752

Note. SD=standard deviation. Between groups: Kruskal–Wallis test, *$p < .05$, **$p < .01$, ***$p < .001$. Minimum and maximum values are provided in brackets.

that reported lower levels of health-related and motor performance-related fitness in youth with CF (Arikan et al., 2015; Corten & Morrow, 2020; Gruber et al., 2008, 2020). Additionally, this study highlighted that aspects of motor performance-related and health-related fitness linearly increased from middle to late childhood among youth with CF, whereas only minor changes or even a decline were observed among older adolescents. This finding was also observed in a previous study (Gruber et al., 2008). In contrast, a linear increase in motor performance up to age 17 years was found in the same-aged healthy group.

Several reasons could explain the poorer motor performance of youth with CF and the differences we observed between them and their healthy peers. In CF, diminished physical fitness can be partially explained by reduced lung function (specifically reduced FEV1 in % pred). There is strong evidence that exercise outcome measures, especially VO2peak and FEV1, are positively correlated (Radtke et al., 2018; Smith et al., 2017). However, the participants in the present study were only mildly affected or had a nearly normal FEV1 (% pred) and a normal cardiorespiratory endurance (Wpeak). FEV1 (% pred) showed a positive correlation only with Wpeak but not with the DMT test items, steps, and PA intensity. This result suggests that balance, sideway jumping, push-ups, sit-ups, and standing long jump are independent of lung function, which is in contrast to Corten and Morrow's (2020) findings.

To date, no information is available on the relationship between lung function and motor performance and on possible limiting factors. Further studies on these issues are warranted. Nutritional status may also play an important role in

Table 4. Correlation Analysis of DMT Test Items, Lung Function, and Intensity of Physical Activity in Children and Adolescents With CF.

Variables	Age	Height (cm)	Weight (kg)	BMI	FEV1 (% pred)	Wpeak (W/kg/Bw)	Total BB (steps)*	JS (counts in 15 s)	FB (cm)	PU (counts in 40 s)	SU (counts in 40 s)	SLJ (cm)	Steps/ day	Sed. time (<1.5 METs)	Light intensity (1.5–2.9 METs)	Mod. to vig. intensity (3–5.9 METs)	Vig. intensity (>6 METs)
Age (years)		**.923****	**.920****	**.710****	−.130	**.704****	−.003	**.641****		**.486****	**.593****	**.539****	**−.429***	**.409***	−.070	**−.406***	−.056
Height (cm)			**.958****	**.707****	−.197	**.739****	−.069	**.586****	−.300	**.450***	**.592****	**.560****	−.245	**.444***	.022	−.245	−.095
Weight (kg)				**.865****	−.076	**.791****	−.085	**.655****	−.297	**.448***	**.607****	**.570****	_.298	**.424***	.004	−.289	−.091
BMI					.130	**.723****	−.050	**.635****	.237	.353	**.500****	**.513****	−.270	.225	−.004	−.227	−.059
FEV1 (% pred)						**.400***	−.110	.214	−.055	.213	.169	.206	.141	−.038	−.176	−.150	.198
Wpeak (W)							.273	**.636****	−.015	**.514****	**.732****	**.682****	−.138	.263	−.299	−.034	.389
Total BB								.153	.028	−−.049	.056	−.019	.169	−.194	.327	.034	−.213
JS									−.248	**.696****	**.819****	**.534****	−.053	.240	−−.130	−.127	.101
FB										.007	−.024	−.273	−.144	−.227	−.065	−.204	.337
PU											**.737****	.552	−.003	.258	−.201	−.278	.281
SU												**.611****	−.026	.250	−.230	−.153	.217
SLJ													−.063	.271	−.348	−.236	.060
Steps/day												.		.269	.192	**.678****	−.020
Sed. time															−.073	**.601****	.152
Light intensity																.413*	**−.474***
Mod. to vig. intensity																	**−.516****
Vig. intensity																	

Note. BW = Bodyweight, BB = backward, JS = jumping side-to-side, FB = forward bend; PU = push-ups, SU = sit-ups, SLJ = standing long jump, sed = sedentary, mod. = moderate, vig.=vigorous, METs = metabolic equivalents. Spearman rank test, $^{*}p < .05$, $^{**}p < .01$, $^{***}p < .001$.
The boldfont represent a significant result.

determining exercise limitations in CF. Malnutrition among individuals with CF is associated with loss of muscle mass and body fat (Lands et al., 1992; Shei et al., 2019). Malnutrition may also contribute to muscular atrophy and can adversely affect lean muscle mass, in addition to causing mitochondrial dysfunction and altered muscle metabolism (Gruet et al., 2017; Lands et al., 1992; Troosters et al., 2009).

Those in middle childhood and late childhood/early adolescence with a BMI of $<18\ kg/m^2$ can be classified as underweight. Malnutrition may be one of the reasons for the diminished muscular strength and power among youth with CF, especially on DMT items based on power and strength (sit-ups, push-ups, standing long jump, and sideway jumping) (Gruet et al., 2017). In contrast, the healthy reference group showed nearly normal or normal BMI, along with higher levels of strength and power. No information on body composition or muscle mass was available for the youth with CF in this study or for the healthy group. The relationship between body composition and health-related and motor performance-related fitness has been seldomly investigated (Gruber et al., 2014; Hebestreit et al., 2010; Kriemler et al., 2013; Prévotat et al., 2019) and should now address the effects of body composition and especially motor performance-related fitness in CF.

In addition to malnutrition and low PA levels, low peripheral muscular strength has been identified as another disease-specific factor in patients with CF. Chronic inflammation and an intrinsic defect in the skeletal muscle are caused by the lack of the CF transmembrane conductance regulator (CFTR) (Divangahi et al., 2009; Gruet et al., 2017). The lack of CFTR causes mitochondrial abnormalities, which may interfere with the production and use of energy. Impaired CFTR function and abnormalities lead to lower muscle strength and endurance and higher fatigue in people with CF, thus limiting physical fitness (Troosters et al., 2009). The lower values of the test items sideway jumping, push-ups, sit-ups, and standing long jump in youth with CF than in their healthy peers can therefore be attributed to disease factors.

In general, the focus of exercise programs in CF is to improve cardiorespiratory endurance. Cardiorespiratory endurance is usually measured as VO2peak, which is determined using a full cardiopulmonary exercise test (CPET) (Hebestreit et al., 2015). This parameter is a predictor of survival in CF in addition to FEV1 (% pred). A higher VO2 is associated with better prognosis (Nixon et al., 2001; Vendrusculo et al., 2019). Thus, training studies in CF have mostly focused on the improvement of VO2peak as a primary outcome and a cardiorespiratory parameter determined with a CPET (Gruber et al., 2014; Hebestreit et al., 2010; Kriemler et al., 2013; Radtke et al., 2017). A sports and exercise program aiming to maintain or improve physical fitness in CF should consider recommendations on frequency, intensity, time, and type of exercise (Pfeifer & Rutten, 2017; Swisher et al., 2015).

Only two studies have focused on parameters of motor performance-related fitness and the beneficial effects of exercise programs (Gruber et al., 2008, 2020). In these studies, motor performance tests were performed in addition to CPET or cycle ergometry without measuring gas exchange parameters. The exercise programs consisted of different sports activities with a great variety of demanding tasks. At the end of the exercise program, significant improvements in motor performance parameters were observed (Gruber et al., 2008, 2020). The authors concluded that activities highly demanding of motor skills had beneficial effects on neuromotor fitness, leading to corresponding adaptations in children and adolescents with CF (Stodden et al., 2008). Interestingly, beneficial effects on cardiorespiratory endurance (health-related fitness) were reported in only one study (Gruber et al., 2008).

Physical fitness is a set of different attributes related to performing daily tasks, locomotor movements (e.g., jumping), and fundamental movements (e.g., throwing, catching, and balance), which are the basis for developing and executing demanding motor tasks in sports, exercise, and daily life (Barnett et al., 2016; Malina et al., 2004; Stodden et al., 2008). Numerous studies have confirmed the existence of a positive relationship between motor performance and PA in healthy children and adolescents (Barnett et al., 2009, 2016; Loprinzi et al., 2015; Lubans et al., 2010; Utesch et al., 2019). Lower levels of motor performance-related fitness are associated with a more sedentary lifestyle and avoidance of situations that demand higher motor skills (Barnett et al., 2016; Field & Temple, 2017).

To date, the relationship of habitual PA to motor performance has not been addressed in previous CF studies (Arikan et al., 2015; Corten & Morrow, 2020; Gruber et al., 2008, 2020). In the present study, PA data were recorded using accelerometry and were correlated with health-related and motor performance-related fitness data for the first time. The age groups of children and adolescents with CF met the recommendation for moderate-to-vigorous PA. In contrast, the recommendation for time spent in vigorous-intensity activities was not met. A decrease in moderate-to-vigorous and vigorous PA was observed, especially in late adolescence. In addition, the time spent in sedentary and light-intensity activities increased. This result could be partially attributed to school attendance and to extensive chest and medical therapies (Castellani et al., 2018).

The intensity of PA and sedentary time have been commonly associated with motor performance in healthy individuals (Barnett et al., 2016). The present findings suggest a positive association of the time spent in PA and PA intensity with motor performance among children with CF, just with healthy individuals (Barnett et al., 2016; Haga, 2008; Hands et al., 2009). Lower vigorous PA and lower time spent in PA might explain the decline in motor performance, especially among late adolescents with CF.

Only a few studies have investigated the psychological factors that might contribute to lower motor performance in children and adolescents with CF.

Poor motor competence and unpleasant sensations (e.g., discomfort, muscle soreness, shame, coughing, breathlessness) during PA and exercise may decrease the motivation to participate in PA. As a result, children and adolescents with CF try to avoid sports and exercise. Negative sensations and perceived inadequacy at physical tasks seem to be more pronounced in the group of late adolescents with CF (Denford et al., 2020). This can be seen as a barrier to sports and exercise participation and might explain the differences between children and adolescents with CF and their healthy peers. Another psychological aspect that is positively associated with PA in healthy children and adolescents is the physical self-concept (Babic et al., 2014; Jekauc et al., 2017; Lemoyne et al., 2015). The physical self-concept can be defined as the self-evaluation of one's physical attributes (Lemoyne et al., 2015). The physical self-concept is a mediator or moderator of PA and is associated with PA in children and adolescents (Jekauc et al., 2017; Lemoyne et al., 2015). In particular, the subdomains of sport competence and perceived fitness are the moderators with the strongest association with PA in healthy children and adolescents (Babic et al., 2014; Stodden et al., 2008).

No data are available on the physical self-concept of children and adolescents with CF. It can be suggested that for persons with CF, the physical self-concept is a mediator and moderator between PA and motor performance. Individuals who are more engaged in PA and organized sports have a more positive physical self-concept and higher levels of motor performance (Denford et al., 2020; Jekauc et al., 2017). Further studies should be conducted to better understand the association among physical self-concept, motor competence, and PA participation in individuals with CF.

Limitations and Directions for Future Research

This study examined the attributes of motor performance in children and adolescents with CF. Poorer motor competence levels were found in this group than in their healthy peers. Another strength of this study was that the accelerometry data were correlated with parameters of motor performance-related and health-related physical fitness. However, despite these study strengths, this study had some limitations. First, testing of physical fitness was part of the CFmobil project, and the children and adolescents voluntarily participated in the program. Therefore, a self-selection bias cannot be ruled out for these participants. However, only those with poor physical fitness or motor performance were encouraged to participate in the project. Individuals with higher fitness and motor performance levels and/or those who engaged in regular exercise and organized sports activities may not have been motivated to enroll in this exercise program. Second, the relatively small sample size recruited from only three centers could limit the generalizability of our findings. Third, boys and girls were not separately analyzed, despite possible gender differences on these variables of interest. Cultural, psychological, and social factors that were not recorded in the present study should be addressed in future studies.

Conclusions

This results of this study revealed a lower health-related and motor performance-related fitness among children and adolescents with CF relative to their healthy peers. In addition, this study highlighted that, in late adolescence, only small increases or even a decrease in individual fitness parameters were noticed. Thus, vulnerable late adolescents warrant increased attention to ensure their long-term motivation to maintain exercise participation. The assessment of a wide range of parameters of physical fitness should be part of of routine exercise testing and monitoring. This may help professionals create individualized exercise programs and openly recommend physical exercise participation. Further studies with a greater number of participants from diverse data pools are needed to determine the underlying factors responsible for poorer health-related and motor performance-related fitness in children and adolescents with CF.

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