Physical activity, lung function and lung clearance index in children with cystic fibrosis

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Objectives: Physical exercise is part of the cystic fibrosis (CF) treatment world-wide. Physical activity in steps and expressed as energy expenditure in Metabolic equivalents (METs) can be measured with a Body monitoring system; SenseWear® armband (SWa). Multiple Breath Washout (MBW), expressed as Lung Clearance Index (LCI) gives information about the involvement of the peripheral airways of the lungs which adds important information to the results from spirometry. The study is part of a cross sectional multicentre study performed at four CF centres in Europe.

We aimed to evaluate physical activity in relation to FEV1 and LCI in children and adolescents, 6–18 yrs old.

Methods: Twenty-six children (8 F) 6–18 yrs, participated at Gothenburg CF-centre. Seven declined participation. Spirometry was performed and LCI was measured by MBW with SF6 using mass spectrometry. The SWa was used for 6 × 24 hours.

Results: Mean (SD) of FEV1 (% pred), MEF25 (% pred), LCI and steps/day were: 88 (12), 70 (26), 7.6 (1.4), 14,531 (3,633) in the group 6–12 yrs (n = 13 [4 F]) and 87 (15), 59 (38), 9.3 (2.5), 12,907 (3,844) respectively in the group ≥12–18 yrs (n = 13 [4 F]). No significant difference was found between the two age groups. However, younger spent significantly longer time per day on a moderate to high activity level (6–9 METs, as jumping on a trampoline or running 10 km/h) than older (1 h 20 min compared to 30 min, p = 0.0004). Neither FEV1 nor LCI did correlate to time spent on a moderate to high activity level.

Conclusion: Activity on a moderate to high level per day was seen in both age groups, although less in the older group, and was not significantly related to LCI or FEV1.

Quantifying weight bearing activity in children and adolescents with cystic fibrosis

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Background: People with cystic fibrosis (CF) are at increased risk of developing low bone mineral density (BMD) which may lead to pathological fractures and kyphosis. Prevention and treatment are the most effective strategies for sustaining bone health to help maintaining the quality of life in CF. Weight bearing activity (WBA) has been proven as an effective way to enhance bone turnover in healthy population.

Objective: To quantify the amount of WBA of CF children and adolescents.

Methods: WBA was evaluated through a self reported questionnaire. A total peak strain intensity score (TPSS) was constructed taking into account the peak strain scores (PSS) reported by Groothausen, the frequency (mean sessions/week) and duration (mean minutes/session) of individual physical activity over the previous year, thus providing a more detailed representation of peak strain associated with physical activity on an individual basis. The minimum recommended TPSS of 120 was obtained, that is 20–30 minutes high impact weight bearing activities three times a week.

Results: 131 CF subjects (51% female) aged between 6 and 18 years (median age 11.81 yrs) with mean FEV1%pred. (SD) 93.9 (19.8), were collected. Median (IQR) TPSS was 145 (360). 36.6% (n = 48) of patients did not meet the recommendations. Further analysis did not reveal any association between the TPSS and BMD z-score, gender, mutation class and FEV1% pred.

Conclusions: Only a minor part of our sample was below the recommended WBAs. Prevention and/or treatment of bone disease might take into consideration the WBAs done by CF patients.

Assessing for ambulatory hypoxia in adult CF patients

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Objectives: Oxygen supplementation can help CF patients with ambulatory hypoxia to increase exercise capacity, reduce breathlessness and reduce the risk of chronic hypoxemia. Identification of such patients under normal clinical practice can be difficult without direct observation of exercise saturations. Using a simple treadmill assessment, we intended to find out what proportion of patients with reasonably good saturations at rest would be shown to be hypoxic under ambulatory exercise.

Method: 219 stable adult CF patients (mean age 27 [SD ±9.2], 98 male), with a resting SpO2 of 93% or above, underwent ambulatory assessment using a treadmill at an intensity which represented their ‘normal walking pace’. Oxygen saturations were measured during the activity and steady state SpO2 was recorded.

Results: 44 of the 219 (20%) patients assessed were hypoxic on exercise, dropping below 90% and ≥4% from resting saturations. Mean resting SpO2 was 95% (SD ±1.4) in the hypoxic group and 96% (SD ±1.4) in the non-hypoxic group. Mean exercise SpO2 of the hypoxic group was 87% (SD ±2.1) with a mean drop of 7.9% (SD ±2.5). Mean FEV1 of the hypoxic group was 41% (SD ±16.2) compared to 61% (SD ±20) in the non-hypoxic group.

Conclusion: This study shows regular ambulatory exercise assessment may help identify those patients with reasonably good lung function and resting oxygen saturations, who will nevertheless de-saturate on normal daily activity. Such assessment as a part of normal clinical practice can give clinicians an early warning of patients who present as well oxygenated at rest but are at risk of ambulatory hypoxia.

Development of a musculoskeletal screening tool for adults with CF identifies common causes of pain and problems that potentially decrease physical activity: a tool for use in annual reviews

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The need for a musculoskeletal screening tool (MST) was identified to diagnose and treat patients with musculoskeletal (MSK) pain, joint and muscle problems.

Objectives: To develop a MST to identify/treat MSK problems early and prevent dysfunction.

Methods: A MST was constructed and trialled in adults with CF 2008–2011. The refined MST consists of pain chart, postural alignment measures, scapulae positioning +/- winging, shoulder protraction, thoracic/lumbar mobility (0–5 score for none, mild, moderate or severe changes), pelvic tilt, hamstring/calf length.

Results: Between 2011–2013 we screened 99 patients (44 female) mean age 30 (SD 9.0), range 18–56 years; FEV1% 51 (21.3), 21–94. Percentage of patients with MSK problems: back pain 42%; neck problems 28%; thoracic kyphosis 46% (54% mild, 44% moderate), shoulder forward protraction 62% (75% mild); scapula abduction 33%; winging 20%; pelvic tilt anterior 66%/posterior 12%. Patients with movement limitation: thoracic ext./shoulder flex 19%; thoracic rotation 28%, lateral flexion 32%; hamstring tightness 70%. Mean distance sit and reach 13.0 (10.7) cm. Calf flexibility 8.2 (3.7) left, 9.1 (8.0) right. Patients with FEV1 less than 40% were more likely to have a thoracic kyphosis, r = 0.93. Hamstrings were tight across the spectrum of lung disease. The MST was well accepted, took 30 minutes to administer.

Conclusion: In order to prevent a decrease in physical activity from MSK problems the MST has become an essential screening and treatment assessment tool to optimise physical function and promote exercise participation. This objective assessment is an established component of the annual physiotherapy review in adults with CF.