## 289* Respiratory exacerbations and muscle strength in adult patients with cystic fibrosis

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Muscle strength is decreased in adult patients with cystic fibrosis (CF). CF is characterized by periodical worsening of the respiratory symptoms or exacerbations. The aim of this study was to investigate whether the frequency of respiratory exacerbations has an influence on the long-term change in muscle strength.
A sample of 45 adult CF patients (age 18–30 yrs; $FEV_1$ 67±19%pred) who performed at least 2 maximal isometric quadriceps strength (QS) measurements during the period 2005–2008 was included. Each 2 consecutive tests were considered as one observation (n = 87) and the mean number of exacerbations per year between the 2 tests was recorded. Based on exacerbation frequency, patients were divided in 3 groups: 0 (group A), 1–2 (group B) and >2 (group C) exacerbations per year. As expected, the table shows that QS increases more between 2 measurements in group B compared with group C. Surprisingly the increase in QS in group A tends to be less than in group B. Patients in group A appear to have a better preserved health status ($FEV_1$) and better QS, partially related to a better preserved body weight (see Table). Probably according to the regression to the mean phenomenon these patients tend to have less increase in QS over time.
As frequent exacerbators show a low baseline QS and a slower increase over years, these findings suggest that muscle strength should be regularly assessed in these patients.
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| No. of exacerbations/ year | Age (yrs) | FEV1 (%pred) | Baseline QS (Nm) | Baseline QS/ body weight (Nm/kg) | Change in QS (% compared with baseline) |
|---|---|---|---|---|---|
| A. 0 (n = 43) | 24±4 | 78±14* | 166±58º | 2.6±0.7 | +3±15 |
| B. 1–2 (n = 16) | 23±4 | 65±21 | 138±40 | 2.3±0.5 | +16±21º |
| C. >2 (n = 28) | 22±2 | 58±16 | 131±43 | 2.4±0.7 | +2±14 |

*$p < 0.05$ vs B and C; º$p < 0.05$ vs C.

## 290 Electromyographic evaluation of muscular function in adults with cystic fibrosis

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Peripheral muscle dysfunction is now recognised as one of the main systemic effects of cystic fibrosis (CF) and contributes to the exercise intolerance and reduced quality of life. Therefore, evaluation of muscular function shows clinical relevance in CF. The purpose of this study was to validate surface electromyographic (sEMG) technique in adults with CF to allow a non invasive evaluation of skeletal muscle function. Fourteen adults with CF (FEV1: 55.1±8%, BMI: 21±2 kg/m$^2$, none was treated with oral corticosteroids) were test on two occasions, six weeks apart. sEMG was recorded from the rectus femoris, vastus lateralis and vastus medialis muscles during sustained submaximal isometric contractions until exhaustion. Quadriceps fatigue was described using two measures for both frequency and time domain: normalized index and slope method. Relative (ICC) and absolute (SEM%) reliabilities were applied to asses test-retest reliability. sEMG measurements at the quadriceps muscle is a reliable method to assess muscular function in CF if the following recommendations are taken: use normalized index in frequency domain for assessment of fatigue change and record signals from rectus femoris. sEMG method can be used in quality control of rehabilitation processes and muscle training in patients with CF.
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Reliability of sEMG variables

| Muscle | Method | Frequency domain | | Time domain | |
|---|---|---|---|---|---|
| | | ICC | SEM (%) | ICC | SEM (%) |
| Vastus medialis | normalized | 0.72 | 7.7% | 0.41 | 30% |
| | slope | 0.80 | 45% | 0.73 | 69% |
| Rectus femoris | normalized | 0.82 | 3.7% | 0.61 | 15% |
| | slope | 0.87 | 37% | 0.76 | 39% |
| Vastus lateralis | normalized | 0.78 | 7.2% | 0.60 | 14% |
| | slope | 0.72 | 45% | 0.74 | 51% |

## 291 Assessment of specific exercise for correction of postural disorders in CF patients

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**Introduction:** Disorders of posture are common in cystic fibrosis. Typically they are postural disorders, which not corrected in time becomes fixed (structural). Fix chest is associated with stiffness of joints between ribs and vertebra. These two factors reduce the mobility of the ribs and the ability to develop the necessary intrapleural pressure for normal lung expansion, which may lead to repercussions like: chest pain, chest rigidity, muscles contracture, muscle weakness.
**Material and Method:** The study was conducted on a number of 8 patients in the group 12–18 years: 3 postural kyphosis, 2 with structural kyphosis, 2 with scoliosis, 1 kyphoscoliosis. To quantify the disease evolution we imagine a questionnaire in which the patient, over a period of 6 months, noted on a scale of 0 to 3, the subjective perception: pain, muscle contracture, joints reduced mobility and muscle weakness. In exercise selection we pursued: postural improvement, increased flexibility in the spine, increasing muscle strength, improve breathing.
**Results:** In patients with postural disorders, subjective symptoms clearly improved for all parameters. In patients with structural disorders exercise had a specific role in the prevention and/or delay progression of postural disorders.
**Conclusions:** An improved symptom after these exercises was superior to those with postural problems, compared with those who became fixed disorder (structural). This shows the importance of applying specific physical exercises near the beginning.

## 292 Musculoskeletal problems in Cystic Fibrosis

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**Background:** With an increase in life expectancy even up to the third decade, physiotherapy has now developed to encompass the whole individual in a long-term perspective. Physical therapy involves a much wider spectrum than only airway clearance and especially focuses on preserving physical function in order to improve quality of life, as well as continuous therapeutic education about the disease and its treatment.
**Objective:** To evaluate and screen posture in adult patients with CF and derive if necessary physical therapy approaches for their treatment.
**Design:** Eighteen patients with CF (including 13 females) aged 19 to 49 years were included in the pilot study. The following tests were performed: observation of the posture(viewing from front, sides and back), measurement of the posture (flexion and extension in sagittal plane) and mobility using SpinalMouse®, and the health status questionnaire EuroQol (EQ-5D).
**Results:** Patients with CF showed three posture types i.e. physiological (n = 5), hyperkyphosis (n = 7) and loss of physiological kyphosis and lordosis (n = 6, including scoliosis). The level of health (EQ-5D) ranged from 50 to 89 points (100 = best health status).
**Conclusions:** Our pilot study showed 13 patients (72%) with incorrect posture of both types. Adequate body posture plays an important role in regards to positive body image and self-esteem. With assistance of SpinalMouse®, a non-invasive objective evaluation of the thoracic shape (graph of the posture) is used for the first time in physiotherapy assessment. Further studies employing a greater number of patients, differentiated age groups and measurement of the spinal shape in the frontal plane are needed to evaluate and confirm specific CF posture types.