238* Working capacity and oxygen uptake in CF

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A cross-sectional study of exercise capacity in all CF patients over 10 years of age was attempted. Exclusion criteria were previous lung transplantation (6 patients) and colonisation with certain multiresistant bacteria (5 patients). Of 81 eligible patients 23 declined participation or were deemed unsuitable for the exercise protocol. In 58 participants (53% males) with a mean age of 24±9 years, FEV_1 was 78±14% of predicted. Pancreatic insufficiency was present in 78% and CF-related diabetes in 10%. The genotype group was ΔF508/ΔF508 in 53%, ΔF508/other in 40% and other/other in 7%. A cycle ergometer exercise protocol with increments of 10–30W/min was performed as determined by previous year's test. Heart rate, blood pressure, ventilation and pulmonary gas exchange were monitored until maximum. At peak exercise heart rate was 180±12 bpm, respiratory rate 46±9/min and respiratory quotient ($V'CO2/V'O2$) 1.23±0.74 and ventilatory quotient ($VE/V'O2$) was 40±6, all indicating a physiologic maximum. The mean maximum work load (206±63 W, 3.3±0.8 W/kg body weight), and oxygen uptake 2.48±0.69 L/min (39±8 ml/kg/min) was normal. Despite higher FEV_1% predicted in children (<18 years), there was no difference between adults and children for peak performance or oxygen uptake per kg body weight.

Conclusion: In adult CF patients with relatively well-preserved lung function, normal exercise capacity is attainable up to young middle age.

239 Clinical and polysomnographic profile in miscigenated patients with cystic fibrosis

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No data exists on Obstructive Sleep Apnea Syndrome (OSAS) prevalence in children with Cystic Fibrosis (CF) and little data on the sleep architecture considering racial miscigenated group.

Objective: describe clinical and polysomnographic characteristics in clinically stable children with CF.

Study design: cross sectional study.

Material and Methods: 65 patients between 2 and 14-years-old from Hospital Octávio Mangabeira, in Salvador, Bahia, Brazil, were studied from November 2006 up to October 2007. We evaluated age, gender, z-score weight/age, Shwachman–Kulczycki (S–K) score, snoring, restless sleep, sleep efficiency (SE), REM latency, percentage of sleep stages 1, 2, 3, 4 and REM sleep, apnea index (AI), oxygen saturations.

Results: 56.9% were male; age range was from 2 to 14-years-old, mean and SD 7.6±3.1 years. The majority of population (80%/65) was composed of black and mulatto children. Median and SD z-score weight/age was –0.462±1.14. Mean and SD S-K score of patients was 85.43±9.42. Sleep complains were snoring in 50.7% and restless sleep in 76.9%. Mean SE was 81.25%±10.75%; median and SD REM latency was 170.32±109.65; median and SD stage 1 was 7.38±3.9, stage 2 was 39.6±11.39, stage 3 was 3.4±1.2 and stage 4 was 21.8±6.9; median and SD stage REM was 14.3±7.7. Median AI was 1.00±1.94 events/sleep hour, range 0–11 events/sleep hour. 53.8% had mild OSAS, 4.6% moderate OSAS and 1.5% severe OSAS. The oxygen saturation nadir values were 81.6±5.7 range 70%-92%.

Conclusion: Our patients with CF had frequent sleep complaints and significant changes in their sleep architecture. The majority of our CF children had mild obstructive sleep apnea. The results call attention because may have a significant on quality of life and clinical outcomes in this population.

240 Wisconsin–Brittany CF Newborn Screening Study: Comparison of progression of lung disease using the Wisconsin Chest X-ray scoring system in two screened cohorts

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Aim: This study compares the longitudinal progression of lung disease in CF patients identified through newborn screening in Wisconsin and Brittany.

Methods: The study included 115 patients screened in Brittany and 69 patients screened in Wisconsin, between 1989 and 2000. Lung disease was radiographically quantified using the Wisconsin Chest X-ray (WCXR) and the Brasfield Chest X-ray (BCXR) scoring systems. De-identified films were scored by a single pediatric pulmonologist. Generalized estimating equation (GEE) models were used to compare longitudinal progression of scores in the two cohorts and were adjusted for gender, age, genotype, pancreatic insufficiency, and meconium ileus.

Results: 1364 films were scored (an average of 7.4 per patient). Quality control revealed reproducible results in scoring over time. Adjusted GEE analysis showed worse WCXR scores in Brittany patients compared to Wisconsin patients (3.46±0.78; $p < 0.0001$) whereas this was not found with the BCXR scores (−0.29±0.22; $p = 0.19$). By examining subcomponents of WCXR scores, there were significant differences in bronchiectasis, peribronchial thickening, nodule, and hyperinflation but not in opacity or atelectasis.

Conclusion: The finding of milder radiographically-quantified lung disease using the WCXR scoring system in Wisconsin patients may be explained by variations in clinical practices or an unmeasured confounder. Further investigations are needed to elucidate these findings.

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241* Walking distance as an outcome of antibiotic course in patients with cystic fibrosis

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The six-minute walking test (6MWT) is widely used to evaluate exercise capacity in patients with Cystic Fibrosis (CF). We evaluated the 6MWT changes induced by an antibiotic iv course and their relationships with the change in pulmonary function in a group of CF patients.

We studied 33 CF inpatients (19 F, mean age 23.9±SD 7.3, range 14–42 yrs), who received an anti-pseudomonal antibiotic iv course for at least two weeks. In each patient, forced vital capacity (FVC), forced expiratory volume at 1 second (FEV_1), walk distance (WD), oxygen saturation (SpO_2) and pulse rate (PR) at rest and during walk, dyspnoea after walk assessed by visual analogue scale (VAS) were measured before and after the antibiotic course. Results (mean±SD) are listed in the table.

No significant relationship was found between the changes in 6MWT and the corresponding in pulmonary function test variables. Our results show that in CF patients the walking distance improved significantly after antibiotic iv course, and its change is not related to pulmonary function changes. This study suggests that the walking distance may be used as an outcome measure of therapeutic interventions in CF patients.

	Before	After		Before	After
WD (mt)	625±11*	645±11	PR during walk (bpm)	152±21*	159±19
SpO_2 rest (%)	96.7±1.3*	97.2±1.2	VAS (mm)	45±20	42±21
SpO_2 during walk (%)	93.6±6	94.4±4.1	FVC (% pred)	85±18*	95±20
PR rest (bpm)	104±15*	97±16	FEV_1 (% pred)	63±19*	72±22

p value by means of paired t test, *$p < 0.05$