195 Pulmonary function in cystic fibrosis children younger than 3 years old

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Studies suggest that the decline in pulmonary function begins early in CF infants even in those patients without respiratory symptoms.
Objective: to describe pulmonary function in CF children younger than 3 years old and to determine if there is any association between pulmonary function and clinical features.
Methods: observational and retrospective, study. We collected data from CF patients who underwent an Infant Pulmonary Function Test (IPFT) before the age of three years. IPFTs were assessed by partial flow/volume curves with the rapid thoracic-abdominal compression technique to obtain the maximal flow at the functional residual capacity ($V'max_{FRC}$). Clinical issues recorded were: age, gender, genetics, pancreatic sufficiency, nutritional status and microbiological findings.
Results: 46 patients (31 males) with a median (interquartile range) age of 8 months (5 to 15) were included. Overall $V'max_{FRC}$ Z score was –0.8 (–1.4 to 0.3), –0.1 (–1.1 to 0.7) in males and –0.9 (–1.8 to 0.3) in females (p = NS). $V'max_{FRC}$ Z score was –0.3 (–1.1 to 0.2) in children younger than 6 months and –0.1 (–1.3 to 1) in older ones (p = NS). In patients homozygous p.508del $V'max_{FRC}$ Z score was –0.5 (–1.2 to 0.8) while in patients with other mutations it was –0.2 (–1.2 to 0.6) (p = NS). There was also no statistical difference in $V'max_{FRC}$ between well-nourished and bad-nourished patients, and between patients with and without *Pseudomonas aeruginosa (PA)* in sputum.
Conclusion: IPFT assessed by partial flow/volume curves was normal in our CF population less than 3 years old. We have not found any difference in pulmonary function comparing CF children with different clinical features.

196 FEV_1 decline in cystic fibrosis (CF): much attention should be devoted to children and adolescents

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The rate of decline in FEV_1 percentage predicted has been studied in CF to identify the risk of death, risk factors for decline and effect of therapeutic interventions. The aims of our study were to assess the FEV_1 decline in CF patients and to compare it in male and female population.
atients aged 6 to 11 (A), 12 to 17 (B), 18 to 23 (C), 24 to 29 (D), 30 to 35 (E) and >36 years (F), attending at our CF Centre were included. All spirometries, performed over a 4 year period (2006–2009), were considered for each patient. Individual decline in FEV_1 predicted was evaluated using linear regression analysis. 176 (94 males) patients were included. Median (IQR) value of number of spirometries was 15 (9, 22). Median values of baseline FEV_1 and decline in FEV_1 were: 105 (89, 117) and –2.02 (–8.55, 2.23) in A, 84 (74, 94) and –1.85 (–4.20, 2.16) in B, 70 (50, 84) and –0.77 (–2.04, 1.02) in C, 66 (38, 93) and –1.18 (–1.71, 0.92) in D, 49 (39, 66) and –1.12 (–2.66, –0.14) in E, 48 (35, 75) % predicted and 0.24 (–1.56, 1.78) % pred/yr in F, respectively. We found no significant differences in FEV_1 decline between males and females in all age groups. In conclusion, we observed an higher decline in lung function during the pediatric age compared to adult age.
This finding suggests to adapt therapeutic interventions in children and adolescents with the aim to slow the loss of lung function. Gender is not a risk factor in our population. Other risk factors should be assessed to identify which subjects need aggressive therapy.

197 Longitudinal assessment of *Pseudomonas aeruginosa* in children with cystic fibrosis. Differences with healthy infants

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Aims: To identify the age at which young children with CF first acquire *P. aeruginosa* (PA). To elucidate oropharyngeal PA in healthy infants same aged.
Methods: Observational, prospective study. *CF cohort patients*: infant with cystic fibrosis disease diagnosed by neonatal screening, from October 1999 to February 2005 and follow-up from diagnose to four years of life. *Healthy infants*: up to 4 year old children, without lung pathology who needs scheduled surgery. Data from CF patients was a longitudinal study, but Healthy infants data was a cross-sectional information, because ethical aspects. Specimens were taken applying the oropharyngeal suction (OPS) technique. *CF cohort patients*: 877 specimens were obtained from 18 patients; were taken monthly and should they develop lung infection. *Healthy infants*: 104 specimens were taken, from 104 infants, during anaesthesia induction. A signed consent was obtained previously.
Conclusions: PA was isolated at 15/18 of CF patients before 4 years of life, and 44.4% before12 months old. The 1st PA isolated was at 2–31 months (median 11). We detected PA in 3/104 specimens from healthy patients, at 7, 18 and 38 months old. We isolated PA at both groups (CF patients 3.76% vs Healthy infants 2.88%) and no statistically significant differences were found.
The presence of PA in upper airway culture from CF patients is early. The isolation of PA in an upper airway culture should not be patognomonic for cystic fibrosis.

198 Comparison of pulmonary function tests in cystic fibrosis patients colonized with *Staphylococcus aureus* (SA) and *Pseudomonas aeruginosa* (PA)

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Aim: The aim of the study was to compare the results of PFTs in CF pts colonized with SA and PA.
Material and Methods: The group under study comprised the selected group of CF pts able to undergo PFT. The study was carried out from 2008–2010. The pts had 3–4 appointments a year. Two standard tests, PFT and bacteriological test were performed at each appointment. The Master Lab – Jaeger's was used to carry out PFTs. The specimens for bacteriology evaluation were sputum samples and oropharingeal swabs. Identification testing was performed by use of standard methods.
The group under study consisted of 34 pts. The group colonized with SA comprised 22 pts, 12 girls and 10 boys aged 7–18. Out of total SA strains (n = 57) there were 5 MRSA isolated. The group colonized with PA comprized 12 pts (6 pts PA chronic infection, 6 pts PA intermittent infection), 7 girls, 5 boys aged 7–18. In 58 cultured samples there were 31 MSSA, 1 MRSA, 48 PA isolated (out of them 21 PA mucoid phenotypes).
Results: See the tables.

The characteristics of CF pts.

CF patients	age [years] mean±SD	height [cm] mean±SD	weight [kg] mean±SD	BMI [kg/m2] mean±SD
S.a. pts n = 22	12.74±2.76	151.45±14.95	40.18±13.17	16.93±2.64
Pa. pts n = 12	12.61±2.42	151.61±11.16	38.23±10.28	16.30±2.43
p*	0.3952	0.4724	0.1865	0.0947

PFT in CF patients

CF patients	VC %pred mean±SD	FEV1 % pred mean±SD	FEV1%VC mean±SD	FEF50 %pred mean±SD	Raw %pred mean±SD	TLC %pred mean±SD
S.a. pts n = 22	89.15±20.90	80.06±23.54	73.91±12.89	63.70±28.27	203.90±96.10	102.83±16.71
Pa. pts n = 12	83.80±21.06	73.11±28.51	69.78±15.08	56.29±35.29	233.07±169.81	99.68±7.73
p*	0.0922	0.0883	0.0669	0.1198	0.1503	0.0895

* T-Student's test

Conclusions: In the chronically colonized CF pts under study SA colonized pts comprise larger group, approximately 65%. The mean age, height, weight and BMI are very similar in both groups. PFT results in SA pts are slightly better, but the difference is not statistically significant.