374. Cystic fibrosis (adults and children): new aspects of risk factors, treatments and diagnosis

P3369

Lung function decline in a modern cystic fibrosis cohort

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Introduction: Though the starting point for lung function measured by spirometry in children with cystic fibrosis (CF) has improved, the annual rate of decline has not changed significantly during the critical period of adolescence. The aim of this study was to describe factors associated with longitudinal decline in lung function in a contemporary cohort of children with CF.
Methods: Best annual lung function data from children attending the CF service of the Royal Children's Hospital Melbourne were reviewed to determine rate of decline in FEV_1 up until time of transfer to an adult centre. Mixed multi-level modeling was used to determine the influence of age, sex, genotype (homozygous F508del), CF related diabetes mellitus (CFRD), *Pseudomonas aeruginosa* (PsA) infection, and body mass index (BMI) on lung function decline.
Results: Longitudinal lung function data (range 5–20 years) were obtained for 98 patients with CF (55 male) on a median 12 (range 3–16) occasions. Overall, FEV_1 declined by a mean of -0.12 z-score each year ($p<0.001$). Homozygous F508del genotype (-0.10, $p<0.01$), CFRD (-0.15, $p<0.01$) and mucoid PsA infection (-0.09, $p<0.01$) were all independently associated with an increased rate of decline in FEV1 z-score. Taken together, these three factors resulted in a cumulative reduction of -0.25 FEV_1 z-score ($p<0.001$).
Conclusion: Genotype, CFRD and PsA infection are all associated with an increased rate of decline in lung function during adolescence. How these findings relate to underlying lung structural changes, and whether PsA eradication success can influence rate of decline in future cohorts, warrants further investigation.

P3370

Is lung function worsening during adolescence in cystic fibrosis? A retrospective study

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Adolescence is a vulnerable period for patients with cystic fibrosis (CF). It is generally recognized that during adolescence morbidity is higher, lung function declines at higher rates and adherence to treatments decreases. However, specific data on spirometry in this age have only seldom been reported.

The aim of our study was to analyze lung function throughout infancy and adolescence in cohorts of CF patients diagnosed after neonatal screening.
We rectrospectively evaluated the spirometry of patients 59 born in the years 1992–1994 who had been visited at least three times yearly and compared the decline in FEV1 and FEF 25-75 (both % predicted) in pre- adolescence (pre ado; age range 8–12 years.) and during adolescence (age range 13-17 years).
The mean decrease in FEV1% was -0.35 (SD ± 11.9) in the pre ado period and -1.41 (SD ±11,7) during adolescence with no significant difference in decline between the two periods, (p=0.45, paired t test). However, the mean variation in FEF 25-75 was 7.9% (SD ± 32) in pre ado and -6.7 (SD ±26) during adolescence (p=0.009)
Our data show that lung function decreases during adolescence in CF patients as reflected by a loss in FEF 25-75 which is consistent with an anatomical damage initially located in smaller peripheral airways. Although FEV1 is universally adopted as a surrogate outcome it seems to be poorly sensitive in evaluating lung disease progression in CF during adolescence.

P3371

Comparative study of three quality of life instruments in adolescent and adults with cystic fibrosis

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Multiple patient reported outcomes have been used in order to measure quality of life in Cystic Fibrosis in Spain, but it is important to find a consensus on which is the most reliable and valid quality of life instrument.
Objective: To compare three quality of life questionnaires widely used among CF patients in order to prove which of them is more valid and reliable when measuring quality of life and disease severity.
Methods: Two disease-specific questionnaires: Cystic Fibrosis Questionnaire-Revised (CFQ-R), and St. George Respiratory Questionnaire (SGRQ), and a generic instrument: the Short-form-36 health survey (SF-36), were simultaneously administered in to 75 adolescents and adults with CF (≥14 years old), 20 of which were awaiting lung transplantation. Internal consistency (Cronbach alpha), and convergent and construct validity were calculated, as well as external validity (correlations of each questionnaire with demographic and clinical variables: lung transplantation list, FEV1, pulmonary exacerbation and intravenous treatment, digestive disorders, etc.).
Results: Similar reliability, but different validity of the questionnaires was shown. Strong correlations (≥0.70) were found between several of the scales in the three questionnaires: physical functioning, role limitation, energy and vitality, and mental health. The CFQ-R allows a better differentiation of the varying levels of disease severity, plus it has disease-specific scales.
Discussion: An instruments' validity and applicability should be considered when choosing a questionnaire to measure HRQoL in CF patients. The CFQ-R proved to be the most suitable, although some of its subscales could be reviewed.

P3372

Comparison of the Cystic Fibrosis Questionnaire with the St George's Respiratory Questionnaire in adult patients with cystic fibrosis

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Background: The Cystic Fibrosis Questionnaire (CFQ-R) is widely use in outcome studies. Nevertheless, correlation with pulmonary function test (PFT) is weak, namely in mild to moderate impairment.
Aim: To prospective evaluated the accuracy of the CFQ-R with PFT (FEV_1, obstructive ventilation defined as ratio FEV_1 to vital capacity (FEV_1/FVC), and hyperinflation defined as ratio residual volume to total lung capacity (RV/TLC)) and to compare it with the St George's Respiratory Questionnaire (SGRQ).
Methods: Clinical data including PFT were collected. CFQ-R and SGRQ were obtained. Spearman correlation was performed.
Results: 32 patients (13 females) with a mean age of 29*m*11y, body-mass index (BMI) of 22.3±3.4kg/m^2, FEV_1 of 67±29%, FEV_1/VC of 0.67±0.14 and RV/TLC of 0.46±0.15 are evaluated. Results of CFQ-R were: physical well-being 74±25, vitality 59±23, emotion 79±19, eating 93±20, treatment burden 75±19, health perception 72±24, social role 72±18, body image 73±24, role 73±24 weight 77±32, respiration 68±19 and digestion 79±23. Total score of SGQR was 23±17, with following subdomains: symptom 44±25, activity 25±23, impact 15±14. PFT was strongly correlated ($p<0.0001$) with SGQR activity (FEV_1 rho=-0.67) and total score (-0-68), moderately correlated ($p<0.005$) with CFQ-R physical (0.63) and SGQR symptom score (-0.59) and weakly correlated ($p<0.01$) with CFQR treatment burden (0.54), health perception (0.57) and role (0.55).
Conclusions: In adult CF patients CFQ-R is only moderately correlated with pulmonary function. In contrast, the SGQR, initial developed for patients with chronic obstructive pulmonary diseases showed a better correlation with PFT.

P3373

WITHDRAWN

P3374

The use of high frequency chest wall oscillation during an acute infective pulmonary exacerbation of cystic fibrosis

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Background: Cystic fibrosis (CF) patients hospitalised for an acute infective pulmonary exacerbation require increased airway clearance. Specialist physiotherapists may be a limited resource. We investigated the use of high frequency chest wall oscillation (HFCWO), in addition to 'usual' airway clearance techniques (ACTs).
Objective: The aim was to assess the utility of HFCWO (The Vest® Airway Clearance System, Hill-Rom) as a self administered therapy compared to European ACTs in facilitating recovery from an acute infective pulmonary exacerbation in people with CF when used in addition to supervised physiotherapy.
Method: A non-blinded randomised, controlled design was used. Patients who met inclusion criteria were randomised to control or HFCWO groups. All patients received four daily sessions, two supervised by a specialist CF physiotherapist and two carried out independently. The control group carried out their usual ACTs, the study group used HFCWO with pauses to huff and cough. The primary outcome measurement was change in FEV_1.
Results: n=36 (64% male). Data was analysed using the Wilcoxon Rank Sum test.

	Control	HFCWO	P value
Age, Mean (SD) (years)	29.8 (±11.7)	25.8 (±7.3)	NS
Baseline FEV1, Mean (SD) (mls)	1490 (±900)	1570 (±540)	NS
Change in FEV1, Median (IQR) (mls)	120 (50, 260)	240 (80, 360)	0.18
Change in FVC	70 (–170, 370)	370 (90, 620)	0.05
Change in FEF25	170 (–10, 540)	500 (–50, 820)	0.35
Change in FEF75	25 (–10,150)	10 (–30, 50)	0.28

Conclusion: Change in FEV_1 was not significantly different between groups, however a significant improvement in FVC was demonstrated. HFCWO should be further explored as an adjunct in treatment of infective pulmonary exacerbations of CF.

P3375

Long inhalation time is associated with short treatment time when using the I-neb AAD system in target inhalation mode

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The I-neb Adaptive Aerosol Delivery (AAD) System is designed to emit aerosol only during the inspiratory phase of breathing and can be operated in tidal breathing mode (TBM), in which the user breathes in a normal manner during treatment, or target inhalation mode (TIM), in which the user is guided to longer inhalations via feedback from the I-neb AAD system. The I-neb AAD system is equipped with a patient logging system (PLS) to facilitate the analysis of patient breathing by recording data on treatment time and mean inhalation time per breath per treatment.
Forty nine patients with cystic fibrosis were enrolled in a 13 week handling study; PLS data was analyzed for 43 of these patients. Each patient's mean treatment (nebulization) time and inhalation time per breath were calculated for treatments taken in TBM or TIM in order to determine the relationship of these variables.

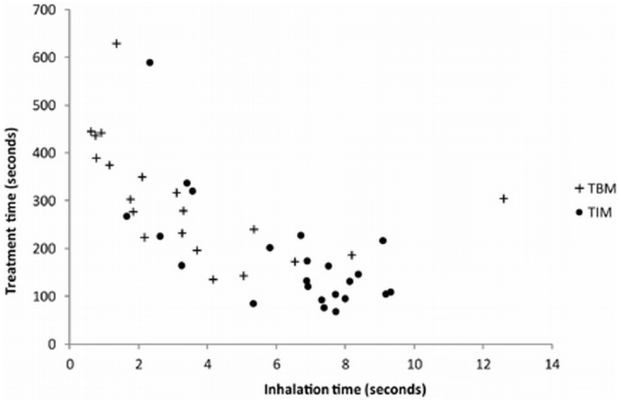

Figure 1. Mean patient treatment times versus mean patient inhalation times.

The median of patient mean treatment times for TBM and TIM were 291 and 146 s, respectively. The median of patient mean inhalation times were 2.6 and 6.9 s, respectively. Patients with longer inhalation times generally had shorter treatment times and patients using TIM had shorter treatment times than those using TBM. It might therefore be of benefit for patients using TBM to switch to using TIM.

P3376

Long-term efficacy and safety of tobramycin 300mg/4mL nebuliser solution in patients with cystic fibrosis and chronic *Pseudomonas aeruginosa* infection

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Introduction: Inhaled tobramycin is considered as standard of care for the management of chronic *Pseudomonas aeruginosa* (*Pa*) infection in patients (pts) with cystic fibrosis (CF).
Objectives: The long-term efficacy and safety of tobramycin 300mg/4mL nebuliser solution (TNS4: Bramitob®, Chiesi FarmaceuticiS.p.A.) administered over 56 weeks (seven 28-day on/off cycles) was assessed in CF pts chronically infected with *Pa.*
Methods: A total of 324 CF pts aged ≥ 6 years with baseline 1-second forced expiratory volume (FEV_1) 40-80% predicted were randomized in an initial 8-week, open-label trial (Core) to receive TNS4 or tobramycin 300mg/5mL (TNS5, Tobi®, Novartis) using PARI Turbo Boy N compressor and PARI LC Plus nebuliser. 209 pts (of which 100 received TNS4 in the Core) continued for an additional 48-week, single-arm extension phase with TNS4 only (Ext). FEV_1% predicted and *Pa* bacterial load in sputum were measured during 14 study visits. Safety was assessed through monitoring of adverse events and audiometry.
Results: Non-inferiority in terms of FEV_1 % predicted between TNS4 and TNS5 was demonstrated in the Core (mean changes from baseline 7.1% and 7.6%, respectively). After 56-week treatment with TNS4, the mean change from baseline in FEV_1% predicted was 5.7% [95% CI: 2.8;8.6]. Reduction in $\log_{10}$CFU/g *Pa* bacterial load was -1.13 [95% CI: -1.58;-0.68]. No remarkable safety findings were detected.
Conclusions: TNS4 demonstrated a sustained and significant improvement in lung function over a 56-week period and a reduction in *Pa* density in sputum. TNS4 was safe and well tolerated.
Supported by Chiesi Farmaceutici S.p.A.

P3377

Pharmacokinetics of tobramycin nebulizer solution (300mg/4ml) administered by Pari e-Flow rapid vs Pari LC plus nebulizer in patients with cystic fibrosis and *Pseudomonas aeruginosa* infection

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Background: New generation nebulizers are increasingly used to reduce the time required for inhalation, potentially improving patient compliance.
Objectives: To compare the pharmacokinetic (PK) profile of tobramycin nebulizer solution (TNS) 300mg/4mL (Bramitob®) administered with the Pari e-Flow rapid versus the Pari LC plus nebulizer.
Methods: Randomized, crossover trial, enrolling 27 cystic fibrosis (CF) patients with chronic *Pseudomonas aeruginosa* infection. Patients received two twice-daily, 28-day treatment periods with TNS 300mg/4mL delivered by either nebulizer, sep-

arated by a 4-week wash-out. Blood and sputum samples were collected on days 1 and 28 over the first 12 and 8 hours, respectively. Primary endpoints were plasma tobramycin maximum concentration (C_{max}) and area under the curve (AUC_{0-t}) on day 28.
Results: 27 patients were randomized and 25 completed the study. Patients were 18-47 (mean 25.5) years old. On Day 28, the geometric mean ratios (e-Flow rapid:LC Plus) for plasma tobramycin C_{max} and AUC_{0-t} were 0.85 (90% CI 0.61-1.19) and 0.87 (90% CI 0.65-1.18), respectively, while the geometric mean ratios for sputum tobramycin C_{max} and AUC_{0-t} were 1.40 (90% CI 0.86-2.27) and 1.37 (90% CI 0.80-2.33), respectively. Nebulization time was significantly shorter for the eFlow rapid as compared to the LC plus: 5.7±2.0 versus 12.1±2.2 min (mean ± SD on day 28).
Conclusion: Plasma and sputum PK data support comparable pulmonary delivery of TNS 300 mg/4mL in CF patients using different nebulizers with a shorter nebulization time for the Pari e-Flow device.
Supported by: Chiesi Farmaceutici.

P3378

WITHDRAWN

P3379
Long-term linezolid in cystic fibrosis patients chronically colonized with Staphylococcus aureus (SA)
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Treatment of chronic colonization with methicillin-resistant Staphylococcus aureus (MRSA) and non-MR in cystic fibrosis (CF) patients shows a wide variability among CF units. While some caregivers prescribe continuous treatment with inhaled/oral antibiotics, others only treat infectious exacerbations.
Methods: 52 CF patients were included. Mean age was 25, 1 years. All patients were chronically colonized by SA (5% MR) and showed a poor clinical progress and respiratory function after receiving several conventional antibiotic cycles to treat infectious exacerbation.
Every patient was prescribed continuous treatment with oral Linezolid: 600mg/12h for 15-21 days every 45 days, for a minimum of 1 year. Serial spirometry, sputum analysis and blood analysis were determined to rule out toxicity derived from a chronic treatment with Linezolid. Health related quality of life was measured every three months with the Cystic Fibrosis Questionnaire-Revised (CFQ-R) to detect any change.
Results: An inflection in the pulmonary function drop in FEV1 and FVC was observed, with a recovery and slowing down of this drop after treatment. After a treatment period of at least one-year, no resistances against Linezolid nor serious adverse events were observed. Patient reported outcomes showed improvement in functionality and in clinical symptoms, with a significant decrease of cough and expectoration during the treatment.
Conclusion: Treatment with Linezolid cycles is effective and safe in those patients colonized by SA who present both a clinical and functional torpid progress with conventional treatments. Linezolid allows the stabilization of the symptoms and lung function.

P3380
Allergic bronchopulmonary aspergillosis (ABPA) prevalence in adult cystic fibrosis (CF) patients. Usefulness of recombinant Aspergillus fumigatus IgE (rAsp f) in diagnosis and monitoring treatment
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The prevalence of ABPA in CF is variable (6-15%). Currently, the role of rAsp f in ABPA diagnosis and treatment monitoring is unknown.
Patients: Serial determinations of serum specific IgE against recombinant AF antigens were analysed yearly during 5 years in 157 adult CF patients. 19 patients fulfilled serologic criteria for ABPA, but only 9 suffered from respiratory symptoms, lung infiltrates or lung function deterioration.
Results: 9 of 157 CF patients (3 women), prevalence 6%, mean age 22 (16-33) years, BMI mean 21 (20-22). In 8/9 Delta F508 mutation was found. At time of diagnosis, 4 patients showed AF colonization, 8 had Pseudomonas aeruginosa (PA). In all patients, total IgE was higher than 500 UI/ml, and AF specific IgE was positive. Titrations for rAsp f varied, with highest titrations for rAsp f 1 and rAsp f 2. Two cases with the most torpid progress showed high titers for rAsp f 3. Lung function's distribution (FEV1) was similar in all cases.
All patients were treated with systemic corticosteroids (3 weeks and gradually reducing the dose after 3-6 months), and itraconazole/voriconazole. There were two relapses after two years, which started with respiratory symptoms.
Conclusion: In CF patients, compatible symptoms as wheezing dyspnea and functional-radiological deterioration suggest ABPA diagnosis. During treatment, only total and AF specific IgE titers, as well as lung function, served for monitoring. In our daily practice, specific AF component IgE titration does not provide diagnostic nor prognostic advantages, and neither does conventional sputum culture.

P3381
Prevalence of tracheobronchomalacia in young children with cystic fibrosis
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Tracheobronchomalacia (TBM) is a commonly diagnosed condition with incidence in general population ranging between 1 in 1400 to 1in 2100 children.In a recent study of adults with cystic fibrosis (CF) by using multi detector CT scans a high prevalence of 29% was reported.
Aim: To study the prevalence of tracheobronchomalacia in children with CF.
Methods: A prospective observational study was done in Princess Margaret Hospital Perth, Australia from Jan 2011 to Dec 2011.Bronchoscopy findings recorded by clinician were collected in children undergoing CT chest and bronchoalveolar lavage as part of a surveillance programme (ARESTCF) in children with CF 6 years and below. A subjective visual estimation of 50% reduction in cross sectional area was used for defining TBM during breathing efforts.
Results: A total of 72 children were studied over 1 year. A prevalence rate of 13.9% was found in the age group studied. Highest prevalence was found during first 2 years (22.7%).

	No. of Children (%)			
Age group	0–2 years	2–4 years	4–7 years	Total
TBM present	5 (22.7)	4 (15.4)	1 (4.2)	10 (13.9)
TBM absent	14 (63.7)	22 (84.6)	22 (91.6)	58 (80.5)
Not observed	3 (13.6)	0 (0)	1 (4.2)	4 (5.6)
Total	22 (100)	26 (100)	24 (100)	72 (100)

Limitations: Prevalence could have been underestimated as spontaneous breathing was not observed in some children due to anaesthetic factors. As technique used was visual estimation there is a potential for observer bias.
Conclusions: A high prevalence of tracheo broncho malacia was found in young children with CF. There is a need for further studies to understand the significance of tracheomalacia in CF.
Acknowledgements: Perth division of *ARESTCF* programme.

P3382
Oral health and some risk factors in children with cystic fibrosis
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Cystic fibrosis (CF) is one of the most common genetic diseases worldwide. Previous studies on patients with CF have reported variable dental caries prevalence and physicochemical properties of saliva. The aim of this study was to investigate prevalence of dental caries and the relation with treatment, salivary flow rate, buffering capacity in children with CF compared with healthy peers.
A cross-sectional observational survey was undertaken in children attending outpatient clinic. Study population included 30 children with CF (mean age 10.2±4.8

years), compared with a control group of 30 healthy children (mean age 9.9±1.4 years). Mean saliva pH was 7.25±0.45 for CF children and 7.45±0.34 for control (p=0.57). Mean salivary flow rate of the CF children was 1.30±0.7ml/min. and 1.54±0.9 ml/min for the control group (p=0.09). And the mean buffering capacity was 8.47±2.7 for CF children and 8.20±1.58 for control (p=0.64). Among the group of children with CF, with 23% of children being caries free. DMFT (Decay Missing Filled Teeth) was 4.6±4.0 in CF children and 7.7±2.7 in control children (p=0.001). Of all CF children, 60% CF brush their teeth once a day. Most of CF children 26(86%) start to brush teeth after 2 years of age.
Although CF is a serious systemic chronic disease of childhood, buffering capacity of saliva seems to be higher than control group. Related to this characteristic of saliva, DMFT score of CF children were found to be lower than healthy group.

P3383

Active and passive smoking by CF patients in Belgium: A national survey

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Background: In Belgium in 2011, 35% of the 15-29 years age group were regular smokers. No information was available about epidemiology of smoking by CF patients in our country.
Objectives: a) primary aim: to identify active (AS) and passive (PS) smokers in the Belgian CF patients b) secondary aim: to investigate physical, psychological and behavioural dependence of AS CF patients.
Methods: AS and PS were identified by means of a urinary cotinine dosage. Patients with positive results were asked to meet with a tobacologist, in order to evaluate their expired CO, their physical, psychological and behavioral dependance to nicotine, and their HAD score.
Results: (Interim analysis) 29 out 706 patients (4.5%, 78.8% of target) had positive urinary cotinine (> 100 mcg/ml); 17 positive patients met the tabacologist (12 AS, 5 PS). Among AS (mean age 28y ± 10, 84%M), mean urinary cotinine level was 0.863mcg/ml ± 0.464 (0.165 to1.5); mean exp CO was 13.3 ppm ± 7.6 (5-25); Fagerström score was 4.5±3 (1-9). 84% had a previous quit attempt. 41% AS smoked cannabis. Anxiety was associated with urinary cotinine level (R=0.825, p=0.009). All AS wished to receive specific help for smoking cessation at their CF reference center.
Conclusions: Active smoking is less prevalent among CF patients in Belgium than in healthy adolescents and young adults. CF AS had developed a moderate physical dependance to nicotine; their urinary cotinine was associated with their anxiety level. Smoking cessation help should be available at CF reference centers.

P3384

Comparison of nanoduct versus macroduct sweat test for the diagnosis of cystic fibrosis in the newborn screening programme in Switzerland

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Background: Newborn screening (NBS) for cystic fibrosis (CF), based on immunoreactive trypsinogen (IRT) and 7 CFTR mutations, was introduced in Switzerland on January 1st 2011. In the pilot phase, we compared the performance of two sweat test methods for diagnosing CF in the NBS.
Methods: All children with a positive screening result were referred to a CF center for confirmatory (diagnostic) testing with: a) the Nanoduct sweat test (conductivity); and b) the Macroduct test (chloride). If sweat test results were positive, borderline or inconclusive, an extensive DNA analysis was performed.
Results: Within one year, 84 children were screened positive. In 30 children the diagnosis of CF could be confirmed, 53 had normal investigations, and 1 child was not yet fully investigated. All details of the investigations were available for 76 children. The children were seen in a CF center at a median age of 24 days. The Macroduct was attempted in 64 children, the Nanoduct in 71 children. A reliable test result was available in 66% (42/64) for the Macroduct and 79% (56/71) for the Nanoduct. In 37 children both sweat tests could be performed; in 19 only the Nanoduct and in 5 only the Macroduct was feasible. In 8 children none of the two sweat tests could be performed, and confirmation or exclusion of CF was based on extensive DNA analysis alone.
Conclusions: In this pilot study, the Nanoduct sweat test showed a better feasibility for use in newborns compared to the Macroduct test, mainly because it needs a lower sweat volume. Analysis of a larger dataset will allow to compare sensitivity and specificity of the two tests for the final CF diagnosis.