112. Cystic fibrosis (adults and children): risk factors and clinical problems

P1111

Cystic fibrosis registry: A preliminary report of CF patient from NRITLD

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Introduction: Cystic fibrosis is an inherited chronic disease that affects the lungs and digestive system of about 70,000 children and adults in the worldwide.To Create more accurate picture of current state of Iranian CF patients, we started to collect and register the data of CF cases at pediatric Pulmonary ward of Masih Daneshvari Hospital.
Material and method: This cross-sectional study was performed on 66 CF patients aged 0-18 years (mean: 14.46 yrs) to describe the demographics, clinical features and outcome among patients hospitalized in Masih Daneshvari Hospital from (2000-2011). 56% of the patients, had Consanguinity marriage and 10.6% had family history of CF in one sibling. Regarding the age at the onset of symptoms, in 33.4% the symptoms was started at the newborn period but only in 9% diagnosis was made at this period. For all patients HRCT of the lung was performed and the dominant finding was Bronchiectasis in 92.70%. Bacteriological findings of cases detected P.A in 68.18% cases, Staph.a in 6.06% cases, AsP.F in 6.06% cases, Candida in 4.54% cases, B.Cepasia in 1.51%. According to the ABPA diagnostic criteria 4 patients diagnosed with ABPA. 17% of the patients are candidate for lung transplantation and 3 patients underwent lung transplantation. Out of 66 cases 19 cases expired due to respiratory failure.
Considering the result of our study and high rate of morbidity and mortality in our cases, we propose that CF registry and evaluation of the disease progression by means of a routine monitoring will increase the survival and improve quality of life of the patients.

P1112

Early deaths in cystic fibrosis: Analysis of causes and risk factors in a pediatric cohort

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Early diagnosis and proper care improved survival in Cystic Fibrosis.
Aim: Analysis of causes and risk factors associated with premature deaths in a pediatric CF cohort.
Methods: Retrospective clinical file analysis of patients followed up during 1999-2011 in a Children's Hospital correlated with level of funding and standards of care.
Results: 50 patients diagnosed and followed up, mean age at diagnosis 11 months (90% before age of 1 year). 13/50 patients had meconium ileus (MI), all underwent surgery, 80% complications rate. 20/50 patients died, 11 before 1 year of age; 50% before 2004 (national funding available as source of care). Mean age at diagnosis in deceased patients was 5 months vs.15 months in the still followed up. Overall prevalence of del F508 was 67%; 55% homozygous status. Identified risk factors for early death were: 85% low socioeconomic status and poor understanding of disease, 80% male gender, 75% homozygous del F508, 60% malnutrition at the time of diagnosis. All 6 deceased MI patients had poor outcomes (reinterventions, poor nutrition, early lung involvement). Severe ABPA, liver disease lead to death in 2 patients. Main cause of death was severe pulmonary disease associated with severe malnutrition in some patients.
Conclusions: Severity of simptoms and rate of disease progression varied widely; and early death was mainly influenced by poor socioeconomic status and presence of del F508 mutation.
With current treatment strategies, specialized care, the majority of patients should reach adulthood with good quality of life. This could be achieved in developing countries too, with involvement of more dedicated clinicians and funding directed to organising of care.

P1113

Improving identification and treatment of children & adolescents with cystic fibrosis related diabetes (CFRD)

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Introduction: Our CF center performed a quality improvement project to improve identification and treatment of CFRD. Our primary goal was to screen at least 80% of patients ≥8 years of age with oral glucose tolerance testing (OGTT) in 2011. Our secondary goal was to improve the mean HbA1c of CFRD patients.
Methods: With CF family and staff involvement, we identified barriers in our existing screening process and after multiple PDSA (Plan, Do, Study, Act) cycles implemented new processes:
Screening>
1) Changed lab protocol for patients receiving OGTT to improve screening efficacy
2) Scheduled annual labs at the first clinic visit of the year
3) Improved mesosystem relationships via weekly huddles between pulmonary and endocrine teams
4) Developed protocol for follow-up of identified patients with CFRD
Protocol Design to Improve HbA1c
1) Plan endocrine consult and transition to endocrine clinic using principles of clinical microsystems
2) Implement system to contact families who failed clinic appointment
3) Provide additional education of staff and families
Results: Screening of outpatients with OGTT improved from 2010 (66%) to 2011 (99%). The mean HbA1c in patients with CFRD in 2010 was 7.0 (n=4). Their levels improved to 6.6 in 2011, and the mean for all patients including newly identified patients with CFRD in 2011 was 6.0 (n=17).
Conclusion: The development of an outpatient screening process successfully identified patients with CFRD. We successfully screened 99% of our patients with OGTTs, and reduced the mean HbA1c in CFRD patients.

P1114

Adults with cystic-fibrosis related diabetes have lower pulmonary function and more infections

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Background: Life expectancy of the CF-patient has increased over the past two decades. As a consequence, more patients develop cystic-fibrosis related diabetes (CFRD). Previous studies showed that CFRD is associated with a stronger decline in pulmonary function.
Aims: The aim of this retrospective study was to examine whether in current times CFRD-patients have more infections and a stronger decline in both pulmonary function and Body Mass Index than non-diabetic CF-patients (NDCF).
Research design and methods: A population of 138 CF-patients aged 17–46 years from the UMCUtrecht Cystic Fibrosis Center was studied. Pulmonary function is measured on a three-monthly basis. All data on pulmonary function were reviewed in the period 1991–2011 using a mixed model analysis. BMI and infection frequency were reviewed in the period 2009–2011.
Results: The FEV1 at baseline was 57% of predicted in CFRD-patients compared to 74% in NDCF-patients ($p<0.001$). Both colonisation with Pseudomonas Aeruginosa and having CFRD were independently associated with a lower FEV1 at baseline ($p=0.025$ and $p=0.036$ respectively). The decline in FEV1 was similar in CFRD-patients and NDCF-patients (0.07 liters/year). There was no decline in BMI in either group. CFRD-patients had more infections requiring i.v. antibiotics than CF-controls, most notably in the group of patients with a FEV1< 45% of predicted ($p=0.034$).
Conclusions: Pulmonary function at baseline was worse in CFRD-patients than in non-CFRD patients. The decline in FEV1 in CFRD-patients was comparable to NDCF-patients. CFRD-patients with FEV1< 45% had the highest number of infections, possibly predisposing to pleural adhesions and more complicated lung transplantation.

P1115

Cystic fibrosis: Heartburn and lung function – "Too hot to handle"

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Background: Respiratory tract injury is the major cause of morbimortality in Cystic fibrosis (CF) patients. Gastroesophageal reflux disease (GORD) might contribute to lung function decline and progression of pulmonary disease in them. Despite the increased gastroesophageal reflux (GOR) symptoms in CF patients, its prevalence and association with gastric aspiration and respiratory impact are not well characterized.
Aim: Assess prevalence of GORD in CF adults, and study possible associations with genotypes, specific colonizations, lung disease severity and GORD treatment.
Methods: Retrospective study conducted in 23 CF patients attended at Pneumology Unit of Coimbra University Hospital, through a direct questionnaire (adapted from *Mayo GER Questionnaire)* concerning GERD symptoms in previous 12 months.
Results: Of 23 studied patients, mean age 30±7,6, 70% female; DF508 homozygote was the most frequent genotype. *P. Aeruginosa* and *S.Aureus* organisms were the most prevalent pulmonary colonization. In 48% lung function was severely depressed. 21 patients (91%) experienced at least one GERD symptom, 12 (57%) referred the occurrence of 5 or more. Concerning lung function severity, we established significant statistical difference between groups under treatment *versus* untreated group, being the first the one that presented the worst CVF values (p=0,032).
Conclusions: Despite the limited evidence that GERD adversely impacts lung function,the possibility that acid suppression improves the outcome of this patients, leads to a strong recommendation for aggressive treatment of GERD. However, according to our study, acid suppression treatment might be insufficient, turning it in a contemporary and scientifically important subject.

P1116

Dynamic hyperinflation in cystic fibrosis

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Introduction: Cystic fibrosis (CF) is the most common life-limiting autosomal recessive disease. CF affects the exocrine glands of lungs, pancreas, intestines and liver. Lungs are usually affected more critically. Exercise tolerance is reduced in patients with CF. It has been shown that pulmonary function weakly correlates with peak exercise capacity. Furthermore, it has been demonstrated that oxygen desaturation during exercise is present in severe lung involvement, but not in mild to moderate disease. Peripheral muscle dysfunction has been demonstrated to be to main factor limiting factor of exercise tolerance. Few studies have looked at dynamic hyperinflation as a limiting factor of exercise tolerance.
Objectives: The aim of this study is was to evaluate the relationship between dyspnea and dynamic hyperinflation in CF during exercise.
Results: Six patients with mild to moderate CF (FEV_1 45 to 79%) were studied during incremental treadmill exercise and changes in inspiratory capacity (IC) were obtained to measure dynamic hyperinflation. During the exercise, dyspnea was assessed by Borg scale. Of the 6 CF patients, 5 patients showed a decrease in IC (dynamic hyperinflation). Furthermore, there was a direct relation between dyspnea assessed by Borg scale and change in IC (r = 0,38).
Conclusion: Our study is the first to demonstrate dynamic hyperinflation in mild to moderate CF and a direct relationship between dyspnea and IC in CF during exercise. This provides a better understanding on exercise limitations in CF.

P1117

Factors predicting mortality in cystic fibrosis where lung transplantation can not be performed

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Introduction: Cystic fibrosis is the most common lethal genetic disorder affecting the Caucasian population. Severe pulmonary disease is responsible for over 80% of deaths associated with cystic fibrosis. Patients with less than 2 years life expectancy and with reduced quality of life are candidates for lung transplantation.
Aim: To evaluate the risk factors of mortality in moderate to severe cystic fibrosis lung disease that doesn't go to lung transplantation.
Material and methods: We follow up 200 cystic fibrosis patients in our clinic and 35 of them with FEV1 ≤%60 were enrolled to the study. Demographic data, pulmonary function test, high resolution computed tomography modified Bhalla score, blood gas analysis and nutritional status of patients were evaluated.
Results: Respiratory support requirement was frequent in moderate to severe lung disease (31.4%). Low pulmonary function, non invasive ventilation requirement, having diabetes mellitus, 0_2 saturation <95, pCO_2 >50 mm Hg and frequent IV antibiotic treatment were significantly related with increased mortality (p<0.05).
Conclusion: This is a single centre study of cystic fibrosis patients with moderate to severe lung disease that did not go to lung transplantation. We evaluated the clinical going and factors that were associated with mortality. We think that it is important to do similar multicentre studies to discover Turkish CF patients' information that have different genetics and phenotype.

P1118

Genotypic diversity of *Pseudomonas aeruginosa* in cystic fibrosis patients with the CFTR I1234V mutation in a large kindred family

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Background: *Pseudomonas aeruginosa* is one of the primary pathogens within the cystic fibrosis (CF) lung and a cause of morbidity and mortality. Reports of cross infection and epidemics with this pathogen within and across CF centers raised the possibility of clonal spread among siblings with CF.
Aim: To genotype *P. aeruginosa* by amplified fragment-length polymorphism (AFLP) analysis in CF patients with the CFTR I1234V mutation belonging to a large kindred tribe, and to determine whether the genotypes are identical among CF siblings and among different families with the same mutation.
Methods: Sixty seven *P aeruginosa* isolates from sputa of 27 CF patients were studied at Hamad General Hospital, Qatar. Genotypic relatedness was assessed using AFLP, and compared with 33 *P. aeruginosa* isolates of a known genotype from the Netherlands.
Results: Twenty seven CF patients were from 17 families, 16 males with a median age of 15 years (range 4 years-34 years). Twenty-two CF patients (12 families) with the CFTR I1234V mutation arising from a single large kindred tribe harboured chronic *P. aeruginosa* colonization. Twenty three unique genotype of *P. aeruginosa* were identified and different from *P. aeruginosa* strains isolated from Dutch CF patients. Ten families each had one unique genotype which were different from each other, four families harboured 2 different genotypes, one family 3 genotypes and two families had 4 genotypes. CF siblings within one family harboured the same genotypes of *P.aeruginosa*.
Conclusions: This study demonstrated genotypic diversity of *P. aeruginosa,*isolates between families and established cross-infection among CF siblings.

P1119

Nebulised antibiotics with the I-neb adaptive aerosol delivery (AAD) system: Impact on adherence in cystic fibrosis (CF) patients

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Purpose: To assess the adherence and compliance to prescribed treatment of CF patients to nebulised antibiotic utilising the AAD system.
Methods: 17 cystic fibrosis patients (12 female, age range 8-18 years) infected with *Pseudomonas aeruginosa* and treated with aerosolised colistin twice a day using the AAD were included. Data were downloaded during a clinic visit 3 months later and in 7 patients, a second downloaded after year. None of the patients were aware that the data were recorded.
Results: Mean overall adherence over the 3 months of treatment (number of recorded doses/number of prescribed doses x100) was 90.73% (SD: 26.37%) with 2 out of 17 patients having an overall adherence below 75%.The mean percentage of days each patient fully adhered was 79.48% (SD: 17.57%) and the mean of fully compliance was 84.90% (SD: 28.27%). Overall mean values adherence was maintained over the year. Mean treatment time was 8.08 minutes. Evening adherence (94.76%, SD:33.34%) was better, (but statistically not significant) than in the morning (86.20%, SD:22.04%). Also, no significant correlations were observed between better adherence and FEV1, $FEF_{25-75\%}$, age, BMI, chronic PA.
Conclusions: The high level of adherence observed in our series contrast with the poor compliance reported in previous studies. The AAD system recording offers very useful information to examine patterns of adherence during aerosolised therapy and allows to monitor aspects of the treatment that can be improved.

P1120

The prevalence and characteristics of intravenous (IV) antibiotics allergy in adult patient with cystic fibrosis (CF)

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Background: CF patients have increased risk of antibiotics allergy. There is little information on the prevalence and characteristics of IV antibiotics in adult CF patients.
Objectives: To determine the prevalence and the role of CFTR genotype in IV antibiotics allergy in adult CF patients, and to describe the characteristics of allergy.
Methods: A retrospective study on all CF patients attending the regional adult CF centre at University Hospital of North Staffordshire between January 2009 and December 2010. Age, sex, CFTR genotype, BMI, spirometry, sputum microbiology/mycology, *Aspergillus* serology, allergic reactions, the type and the number of courses of IV antibiotics administered were recorded.
Results: 39 of 54 patients received at least 1 course of IV antibiotic were studied. 16 patients with allergy (mean (±SE) age 28.1±2.0, 10 female) and 23 without (mean (±SE) age 28.5±2.6, 10 female). The 2 groups had similar spirometry

(FEV_1 % predicted 42.9±4.7 and 51.5±4.6) and BMI. No differences in sex, *ABPA* and *Aspergillus* sensitisation occurrence. 50% of the allergic patients were DF508/DF508.
The risk of allergy was 22% on *Ceftazidime*, 18% on *Tazobactam,* 17% on *Meropenem*, 15% on *Colistin*, 12% on *Aztreonam* and 5% on *Tobramycin.* The common clinical manifestations were nausea/vomiting (27%), arthralgia (19%) and diarrhoea (16%). The overall prevalence of IV antibiotics allergy was 41% (16/39). 10 antibiotics desensitisations were performed with a success rate of 70%.
Conclusions: IV antibiotics allergy is common, *Ceftazidime* has the highest risk and no correlation with CFTR genotype. Further study is required to determine the risk factors for IV antibiotics allergy.

P1121
Tuberculosis among children with cystic fibrosis
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Background: Cystic fibrosis patients are predisposed to pulmonary infections. Conditions associated with CF like underweight, diabetes mellitus (CFRD), liver disease (CFLD) are favoring factors for tuberculosis (TB). The hypothesis of a potential comorbidity of TB in CF children occurred. The aim of the paper was to evaluate the prevalence of TB in children with complicated CF.
Methods: Thirty-two patients(pts) with typical CF, associating complication like CFLD(27 pts), diabetes(3 pts) and 2 with both complication were considered for a prospective five years study. Biannual bacteriologic exam (TB specific also) were included, in addition to clinical examination and annual CT, to the patients evaluation.
Results: Tuberculosis occurred in 6.25%(2 patients), both F508 del homozygous, with CFLD and poor nutritional status; one patient had also CFRD. His evolution was unfavorable; he developed portal hypertension and died from respiratory failure. The other patient was diagnosed with active TB, *Pseudomonas* positive and poor nutritional status, but good evolution after treatment. The rest of CFLD patients had a stationary evolution, except 4 of them (15.38%) developed diabetes. Tuberculin skin test was positive in 4 patients (12.5%), 3 of them received TB vaccine. Despite the mandatory vaccination for TB in our country, only 84% pts were vaccinated. 18.75% of patients (6 pts) were considered and treated as TB cases, without bacteriological confirmation, before being diagnosed with CF.
Conclusion: Although TB is a frequent condition in our area, the prevalence among CF children is not as high as expected. It is possible that other factors, unknown so far, are implicated.

P1122
Acrodermatitis enteropathica-like skin eruption in cystic fibrosis patients
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Dermatitis is an uncommon initial presentation of cystic fibrosis(CF).Here we report 3CF patients diagnosed with severe dermatitis.
Case1: Four-month-old male patient was admitted with diffuse skin eruptions, failure to thrive, edema, hypoalbuminemia and anemia.His sweat test was not diagnostic; but he was suspected to be CF with clinical findings; treated with pancreatic enzyme replacement(PERT), and supportive therapy.His skin lesions resolved within 2 weeks. CF was confirmed with elevated sweat test(96 mEq/L) retested after the resolution of the edema, and mutation analysis that revealed 2183AA-G/-.
Case2: Two-month-old male patient was admitted with diffuse erythemateous and pathcy, exfoliated skin eruptions, anemia, hypoalbuminemia, edema, diarrhea, failure to thrive and bronchiolitis history.He was thought to be CF; treated with PERT and skin lesions resolved within 3 weeks.High sweat test and homozygous DeltaF508 mutation confirmed the diagnosis.
Case3: Two-month-old male patient was admitted with scaling erythematous rash, puffy feet, failure to thrive, diarrhea, hypoalbuminemia, and anemia.We could not perform sweat test because of diffuse edema and rash.He was diagnosed as CF with clinical and laboratory findings.Despite supportive therapies, we lost him becuse of staphilococci sepsis as the skin integrity was impaired.Sequence analysis revealed G576A/R668C compound heterozygous mutation.
Acrodermatitis enteropathica-like skin lesions can be seen in CF because of malabsorbtion and insufficient nutrition that lead to hypoproteinemia, zinc and essential fatty acids deficiencies.In patients with these skin lesions, CF must be kept in mind and sweat test must be repeated after the resolution of edema.

P1123
The use of Pezzer catheter in cases of persistent airleak in advanced cystic fibrosis
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Objective: Spontaneous pneumothorax with persistent airleak is a complication that is commonly reported in patients with cystic fibrosis (CF). There is an attributable mortality and considerable morbidity to the complication, resulting in increased health-care utilization and a measurable decline in lung function. We present a series of 11 patients with CF presenting with recurrent pneumothoraces and persistent airleak treated with a Pezzer catheter.
Material & method: All 11 patients presented with large pneumothoraces; a 28 French Argyle intercostal catheter was inserted in the 5th or 6th intercostal space, mid axillary line, and connected to an underwater seal drain, resulting in re-expansion of the lung. However, there was persistent air leak on coughing in 7 cases while in the rest 4 cases, the intercostal tube became inactive and smaller pneumothoraces presented as local ones, impossible to be drained by the already in place intercostal catheter. In both groups either with persistent pneumothoraces or recurrent ones we decided to use Pezzer catheters made of Latex instead of Argyle ones.
Results: All pneumothoraces were resolved within 3 days after the insertion of a Pezzer catheter. There were no complications recorded and in a follow up of 2 years no recurrence was observed.
Conclusion: Our experience supports the use of Pezzer catheter connected to water seal in cases of advanced cystic fibrosis with prolonged air leak, since it promotes pleurodesis. It reduces significantly the duration of the intrapleural drainages and the length of the in-hospital stay. The procedure is cost-effective, safe, easy to perform.