3rd Middle East Cystic Fibrosis Conference

"Working towards optimal CF care in the Middle East"

21-23 March, 2019

Marriott Hotel Sisli, Istanbul / Turkey

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Scientific Program

Wednesday, March 20, 2019

13:00 -14:30	**Pre-Conference Workshop 1** ***Chairs: Nilay Baş, Ela Erdem***	**Room B**
	Sweat chloride testing *Nico Derichs, ECFS Diagnostic Network WG Coordinator, DEU*	
14:30 - 16:00	**Pre-Conference Workshop 2** **Physiotherapy Workshop** ***Chairs: Zeynep Seda Uyan, Mehmet Köse***	**Room B**
	Christopher Tapley, USA *Sharyn Dagher, USA* *Özge Keniş, TUR*	
16:00 - 16:30	Coffee Break	
16:30 - 18:00	**Nutrition Workshop** ***Chairs: Engin Tutar, Güzin Cinel***	**Room B**
	Sandra Bouma, USA	
12:00 - 18:00	**MECFA Annual Business Meeting** **MECFC 2020 Organizing Committee Meeting**	**Room C**

Thursday, March 21, 2019

08:00 - 17:00	**Registration**	**Lobby**
08:45 - 10:05	**Session I: CF: An International Approach** *Chairs: Esen Demir, Elif Dağlı*	**Room A**
08:45 - 09:00	Conference opening and welcome *Bülent Karadağ, TUR*	
09:00 - 09:15	MECFA – Road to the future *Christine Noke, MECFA CEO, USA*	
09:15 - 09:40	The Cystic Fibrosis Foundation (CFF) – Adding tomorrow and living today *Preston Campbell, USA and Bruce Marshall, USA (video)*	
09:40 - 10:05	European Standards of Care How did we make and maintain them? *Carlo Castellani, ITA*	
10:05 - 10:40	Coffee Break	
10:40 - 12:00	**Session II: Where and How to Start- Standards of Care** *Chairs: Nural Kiper, Derya Altıntaş*	**Room A**
10:40 - 11:00	Survey of CF care in Middle East – Results *Hussein Al Kindi, OMN*	
11:00 - 11:30	Developing CF Centers in the region – The MaMi project *Samya Nasr, USA*	
11:30 - 12:00	Combating cross contamination in clinical care-the rules of hygiene *Bülent Karadağ, TUR*	
12:00 - 13:30	Lunch Break	

Thursday, March 21, 2019

13:30 - 14:00	**Recent Advances in CF** ***Chairs: Demet Can, Deniz Doğru*** CRMS/CFSPID *Carlo Castellani, ITA*	**Room A**
14:00 - 14:30	Update on ongoing studies in CF *Samya Nasr, USA*	
14:30 - 15:10	Lobbying for CF care in the Middle East *Ibrahim Al Janahi, QAT* *İlknur Özgür Görgün- President of KIFDER, TUR*	
15:10 - 15:25	CF and Asthma: How to treat? *Bülent Karadağ, TUR*	
15:25 - 15:45	Coffee Break	
15:45 - 17:35	**Session III: Genetics and Lobbying for CF** ***Chairs: Uğur Özçelik, Ayşe Tana Aslan***	**Room A**
15:45 - 16:35	CF genetics in the Middle East *In Oman, Hussein Al Kindi, OMN* *In Qatar, Atqah Adulwahab, QAT* *In Turkey, Pınar Ata, TUR*	
16:35 - 17:05	Optimizing CF diagnosis by personalized characterization of inconclusive CFTR genotypes *Nico Derichs, DEU*	
17:05 - 17:35	Genetics of CF – current status *Milan Macek, CZE*	

Friday, March 22, 2019

09:00 - 10:30	**Session IV: Emerging topics in CF** *Chairs: Haluk Çokuğraş, Fazilet Karakoç*	**Room A**
09:00 - 09:30	Therapeutic drug monitoring and pharmacokinetics in CF *Daniel Touw, NLD*	
09:30 - 10:00	The important role of patient organizations: CF Europe *Jacquelien Noordhoek, NLD*	
10:00 - 10:30	Update on new drugs for CF and the Rainbow project *Hendrikus Heijerman, NLD*	
10:30 - 11:00	Coffee Break	
11:00 - 12:10	**Oral Presentation Session 1** **"Clinical Characteristics of CF in the Middle East"** ***Chairs: Arif Kut, Nagehan Emiralioğlu***	**Room A**
11:00 - 11:10	**OP-01** Caregiver burden of the children with cystic fibrosis *Özge Keniş Coşkun, Kardelen Gençer Atalay, Evrim Karadağ Saygı, Ela Erdem, Yasemin Gökdemir, Bülent Karadağ*	
11:10 - 11:20	**OP-02** Forced Expiratory Volume in the first second (FEV1) decline in siblings with cystic fibrosis *Mehdi Alizadeh, Hoseinali Ghaffaripour, Marayam Hasanzad, Noushin Baghaee, Habib Emami, Korush Fakhimi Derakhshan*	
11:20 - 11:30	**OP-03** Prospective evaluation of vascular changes in acute respiratory infections in patients with cystic fibrosis *Gökçen Kartal Öztürk, Aykut Eşki, Figen Çelebi Çelik, Seçil Conkar, Ahmet Keskinoğlu, Figen Gülen, Esen Demir*	

Friday, March 22, 2019

11:30 - 11:40 **OP-04** Infection control knowledge and practices among cystic fibrosis patients and their families
Can Akın, Ayça Çakan, Eren Sarıoğlu, Batuhan Başçı, Gamze Alçı, Ayşegül Karahasan

11:40 - 11:50 **OP-05** Non-tuberculous mycobacterial infections in children with cystic fibrosis- a cystic fibrosis center experience in Turkey
Cansu Yılmaz Yeğit, Almala Pınar Ergenekon, Emine Atağ, Nilay Baş Ikizoğlu, Yasemin Gökdemir, Ela Erdem Eralp, Berrin Ceyhan, Kaya Köksalan, Ayşegül Karahasan, Bülent Taner Karadağ

11:50 -12:00 **OP-06** Cystic fibrosis and sinonasal problems
Erdal Sakallı, Ayşe Ayzit Kılınç, Haluk Çokuğraş

12:00 - 12:10 **OP-12** Our experience in cystic fibrosis newborn screening programme
Tugba Ramasli Gursoy, Zeynep Reyhan Onay, Tugba Sismanlar Eyuboglu, Ayse Tana Aslan, Busra Sultan Kibar, Sevgi Pekcan, Melih Hangul, Mehmet Kose, Isil Irem Budakoglu

12:10 - 13:10 **Thematic Poster Session** **Poster Area**
"Microbiology and Immunology in CF"
Chairs: Saniye Girit, Nazan Çobanoğlu

PP-01 Evaluation of microorganisms and their resistance in the respiratory tract specimens of patients with cystic fibrosis and the correlation of antibiotic resistance with clinical findings and pulmonary function tests
Mukaddes Kılıç Sağlam, Canan Baydemir, Zeynep Seda Uyan

PP-02 Plasma IgG4 levels in clinically stable adult cystic fibrosis colonized with pseudomonas aeruginosa
Atqah Abdulwahab, Mona Allangawi, Merlin Thomas, Ilham Bettahi, Sivaraman K Siveen, Jayakumar Jerobin, Prem Chandra, Abdul Badi Abou Samra, Manjunath Ramanjaneya

Friday, March 22, 2019

PP-03 Mycobacterium simiae infection in cystic fibrosis: A case report
Hakan Yazan, Lina Al Shadfaan, Erkan Cakir

PP-04 Epidemiology of pseudomonas aeruginosa among cystic fibrosis patients
Gamze Alci, Aysegül Karahasan, Can Akin, Ayça Cakan, Eren Sarioglu, Batuhan Basci, Beyza Oncel, Yasemin Gökdemir, Ela Erdem Eralp, Bülent Karadağ

PP-05 Genotypic relatedness of pseudomonas aeruginosa strains isolated from sputum samples of Cystic Fibrosis patients in Marmara University Hospital CF center
Gamze Alci, Ayça Çakan, Can Akin, Batuhan Başçı, Eren Sarioglu, Beyza Öncel, Ayşegül Karahasan

PP-06 Non-tuberculosis mycobacteria in cystic fibrosis patients
Dilber Ademhan Tural, Nagehan Emiralioğlu, Ebru Yalçın, Deniz Doğru Ersöz, Uğur Özçelik, Nural Kiper

Friday, March 22, 2019

12:10 - 13:10 **"Different Aspects of CF"** **Poster Area**
Chairs: Şebnem Özdoğan, Sedat Öktem

PP-07 A case of allergic fungal rhinosinusitis at cystic fibrosis
Figen Çelebi Çelik, Sercan Göde, Zübeyde Yıldırım Ekin, Gökçen Öztürk Kartal, Aykut Eşki, Hüdaver Alper, Figen Gülen, Esen Demir

PP-08 Evaluation of bone metabolism in children and adults with cystic fibrosis by using falangeal quantitative ultrasound
Velat Sen, Abdurrahim Dusak, Hadice Selimoglu Sen, Müsemma Karabel

PP-09 Evaluation of the factors that affecting quality of life of children with cystic fibrosis
Zeynep Kayra Tanrıverdi, Velat Sen, Ismail Yıldız, Hadice Selimoglu Sen

PP-10 Allergic bronchopulmonary aspergillosis in patients with cystic fibrosis: 13 yeas experiment at one center
Pakize Cennetoglu, Cansu Yilmaz Yegit, Almula Pinar Ergenekon, Nilay Bas, Ela Erdem Eralp, Yasemin Gokdemir, Tamer Bulent Karadag

PP-11 Cystic fibrosis presenting as arthropathy: A case report
Yazan Zeyad Abed, Omar Khaleel Abu Shamma, Nisreen Rumman, Anas Muhanna, Ahmad Qaaqour

PP-12 Presentation of four cases of allergic bronchopulmonary aspergillosis in cyctic fibrosis patients
Aykut Eşki, Gökçen Kartal Öztürk, Figen Gülen, Esen Demir

Friday, March 22, 2019

12:10 - 13:10 **"Diffuculties in management of CF in the Middle East"** **Poster Area**
Chairs: Erkan Çakır, Tuğba Şişmanlar

PP-13 A profile of children with cystic fibrosis in Gaza
Bakr Abu Jarad, Afnan Abufoul, Hanan Abukumail, Rania Abusamra

PP-14 The effect of ivacaftor on adult cystic fibrosis patients at Royal Hospital in Oman
Zakia Ali Al Rashdi, Nasser Al Busaidi

PP-15 Association between CFTR mutation classes and cystic fibrosis severity markers in children
Abdurrahman Erdem Başaran, Ayşen Başaran, Dilara Fatma Kocacık Uygun, Özgül Alper, Ayşen Bingöl

PP-16 Characteristics of cystic fibrosis patients diagnosed after false negative cystic fibrosis newborn screening results
Pinar Ergenekon, Emine Atag, Nilay Bas Ikizoglu, Cansu Yilmaz Yegit, Ellada Rzayeva, Yasemin Gokdemir, Ela Erdem Eralp, Bulent Karadag

PP-17 Clinical characteristics of cystic fibrosis patients at our clinic
Aybüke Kacır, Füsun Ünal, Tuba Koçkar Kızılırmak, Gökhan Baysoy, Sedat Öktem

PP-18 *Clinical and socio-demoghraphic evaulation of cystic fibrosis*
Hakan Yazan, Lina Alshadfaan, Erkan Çakır

13:10 - 14:00 Lunch Break

Friday, March 22, 2019

14:00 - 16:00	**Session V: CF in Middle East** ***Chairs: Figen Gülen, Sevgi Pekcan***	**Room A**
14:00 - 15:30	CF in Iran, Mohammadreza Modaresi, IRN *CF in Egypt, Nader Fasseeh, EGY* *CF Jordan, Samah Awad, JOR* *CF Turkey, Yasemin Gökdemir, TUR* *CF in Omani and Pakistan/Indian patients, Hussein Al Kindi, OMN*	
15:30 - 16:00	Home-made therapies for CF; a recipe for lower cost treatments *Daniel Touw, NLD*	
16:00 - 16:30	Coffee Break	
	Chairs: Figen Gülen, Bülent Karadağ	
16:30 - 17:00	The challenges of treating CF without essential drugs and equipment *Samah Awad, JOR*	
	CF-IDEA, A nonprofit solution to high cost therapies *Christine Noke, USA*	
17:00 - 18:00	Physiotherapy in CF-US and Middle East Experience *Christopher Tapley, USA* *Özge Keniş, TUR*	

Saturday, March 23, 2019

09:00 - 10:30	**Session VI: CF Care in Adults** *Charis: Sait Karakurt, Ayşen Bingöl*	**Room A**
09:00 - 09:30	CF in Adults – What is different ? *Hendrikus Heijerman, NLD*	
09:30 - 10:10	CF Adult Care, challenges in the Middle East *Derya Kocakaya, TUR* *Mona Al Lingawi, QAT*	
10:10 - 10:30	GI and nutrition *Deniz Ertem, TUR*	
10:30 - 10:50	Coffee Break	
10:50 - 11:50	**Top 6 Abstract on Cystic Fibrosis** **Oral Presentation Session-2** ***Chairs: Bülent Karadağ, Ela Erdem Eralp***	**Room A**
10:50 - 11:00	**OP-07** Managing the respiratory care of children with CF in Gaza *Bakr Abu Jarad, Afnan Abufoul, Hanan Abukumail, Rania Abusamra*	
11:00 - 11:10	**OP-08** Distribution of microorganisms isolated from cystic fibrosis patients *Gamze Alci, Aysegül Karahasan, Can Akin, Ayça Cakan, Eren Sarioglu, Batuhan Basci, Beyza Oncel, Yasemin Gökdemir, Ela Erdem Eralp, Bülent Karadağ*	
11:10 - 11:20	**OP-09** Respiratory virus analysis in cystic fibrosis patients with acute respiratory exacerbations *Pinar Ergenekon, Rabia Can Sarinoglu, Cansu Yilmaz Yegit, Yasemin Gokdemir, Ela Erdem Eralp, Guner Soyletir, Bulent Karadag*	

Saturday, March 23, 2019

11:20 - 11:30 **OP-10** Importance of isoforms of immunoreactive trypsin in newborn screening for cystic fibrosis
Bengü Ergenoğlu, Esin Akçael, Ş. Şeyda Pirinççi, Ali İhsan Manav, Erdal Ataç, Fatıma Yücel

11:30 - 11:40 **OP-11** The effect of the family empowerment program on disease management skills of parents with children with cystic fibrosis: A randomized controlled trial
Hatice Donmez, Fatma Taş Arslan, Sevgi Pekcan

11:40 - 11:50 **OP-13** Impact of Body Composition and Nutritional Status on Health Outcomes in Adult Cystic Fibrosis Patients
Nazlı Zeynep Uslu, Derya Kocakaya, Şehnaz Olgun Yıldızeli, Emel Eryüksel, Şeyma Görçin Karaketir, Berrin Ceyhan

11:50 **Closing Remarks,**
Bülent Karadağ, TUR

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Oral Presentations

OP-01 [Clinical Characteristics of CF in the Middle East]

CAREGIVER BURDEN OF THE CHILDREN WITH CYSTIC FIBROSIS

Özge Keniş Coşkun[1], Kardelen Gençer Atalay[1], Evrim Karadağ Saygı[1], Ela Erdem[2], Yasemin Gökdemir[2], Bülent Karadağ[2]
[1]Department of Physical Medicine and Rehabilitaton, Marmara Universtiy, Istanbul, Turkey
[2]Department of Pediatric Pulmonology, Marmara Universtiy, Istanbul, Turkey

OBJECTIVE: To define the state of caregiver burden in the parents of children with cystic fibrosis (CF)

MATERIAL AND METHOD: The study has been done in Marmara University Physical Medicine and Rehabilitation and Pediatric pulmonology department between May 2018 and December 2018. Patients between the ages of 6-13 who had been diagnosed and treated for CF and their parents have participated in the study. Patients' age, sex, respiratory function tests, mutations, psudomonas aueruginosa (Psa) colonization status and the time period since their last exacerbation had been recorded. Patients have filled out the Turkish version of Cystic Fibrosis Questionnaire-Revised (CFQ-R) appropriate for their age. The parents have filled out the Turkish version of the Zarit Caregiver Burden (ZCB) inventory.

RESULTS: A total of 44 children and 58 caregivers (28 mothers, 3 fathers and 14 mother-father dyads) have participated in this study. Patients mean age was 10.4±2.00 (minimum7 – maximum13) and mean FEV1 was 83.1±22.6 (minimum30-maximum118) percent. 28 (62.2%) of the patients were male and 12 (26.7%) had Psa colonization. The mean ZCB scores for mothers were 30.51 ± 10.7 while it was 27.52 ± 9.21 for fathers and were not significantly different (p=0.32). There were significant negative correlations between the mothers' ZCB scores and FEV1, CFQ-R physical, emotional, body image and treatment scores ($p< 0.05$).

CONCLUSION: Both mothers and fathers of the children with CF are under significant burden. This caregiver burden is inversely correlated with quality of life and severity of the disease.

Keywords: caregiver burden, cystic fibrosis, quality of life

OP-02 [Clinical Characteristics of CF in the Middle East]

FORCED EXPIRATORY VOLUME IN THE FIRST SECOND (FEV1) DECLINE IN SIBLINGS WITH CYSTIC FIBROSIS

Mehdi Alizadeh, Hoseinali Ghaffaripour, Marayam Hasanzad, Noushin Baghaee, Habib Emami, Korush Fakhimi Derakhshan
Pediatric Respiratory Disease Research Centre, NRITLD, Masih Daneshvari Hospital, Shahid Beheshti University of Medical Sciences, Tehran, Iran

OBJECTIVE: The end-stage pulmonary disease is the leading cause of death in cystic fibrosis patients but the rate of pulmonary function decline is different among the patients. there are many factors that can change patients' prognoses such as age at diagnosis, sex, disease-causing mutations, body mass index, sputum culture and pancreas sufficiency. The aim of this study is comparing FEV1(forced expiratory volume in one second) decline in siblings with cystic fibrosis.

MATERIAL-METHOD: twenty siblings with the diagnosis of cystic fibrosis and at least two spirometry and one lung CT included in the study and FEV1 decline evaluated by linear regression and compared between males and females.

FINDINGS: Ten boys with mean age of 18 (95% CI = 14.64 to 21.36) and ten girls with mean age of 13.7 (95% CI = 9.41 to 17.99) have mean annual FEV1 decline of -2.6% and -4% respectively (P value = 0.002).

CONCLUSIONS: Females with cystic fibrosis have a more rapid decline in FEV1 compared with males and pulmonary insufficiency appears earlier in their life. By comparing male and female siblings most of confounding variables such as genetic mutation can be controlled and the difference can be attributed to sex.

Keywords: FEV1, Bronchiectasis, Sex, Siblings, Cystic fibrosis

OP-03 [Clinical Characteristics of CF in the Middle East]

PROSPECTIVE EVALUATION OF VASCULAR CHANGES IN ACUTE RESPIRATORY INFECTIONS IN PATIENTS WITH CYSTIC FIBROSIS

Gökçen Kartal Öztürk[1], Aykut Eşki[1], Figen Çelebi Çelik[1], Seçil Conkar[2], Ahmet Keskinoğlu[2], Figen Gülen[1], Esen Demir[1]
[1]Ege University Faculty of Medicine, Division of Pediatric Pulmonology
[2]Ege University Faculty of Medicine, Division of Pediatric Nephrology

Cystic Fibrosis(CF) is a chronic progressive lung disease with systemic inflammation.Although there is an increased risk of cardiovascular disease(CVD) in chronic inflammatory diseases, there is no proven association between CVD and CF.The increase in CVD risk in chronic obstructive pulmonary disease exacerbations was shown to be related to the severity of respiratory tract infection and the increase in systemic inflammation.In our study, we planned to compare augmentation index(AIx) and pulse wave velocity(PWV),which is a marker of CVD, in patients with acute respiratory infection.
Cystic Fibrosis patients aged three years and older who were diagnosed with acute respiratory tract infection in Ege University Division of Pediatric Pulmonology between 1 January-30 June 2019 will be included in the study.

Thirteen patients were included in the study.One patient was excluded from the study because of non-adherence during follow-up.Demographic characteristics of the patients are given in Table1.Antibiotic treatment was chosen according to the clinical condition of the patients and to the colonized microorganism.Eight(66.7%) patients were hospitalized and received intravenous treatment, while four(33.3%) patients received oral antibiotics.Median duration of treatment was 14 days(10-21). Laboratory data and PFTs of the patients before and after the treatment are given in Table2.There was no statistically significant difference between the hemodynamic measurements of the patients before and after treatment(Table3.)

Although according to preliminary results of our study there was no statistically significant difference in hemodynamic measurements,it is thought that the risk of CVD due to chronic inflammation in CF patients may be increased such as other systemic inflammatory diseases.

Keywords: cystic fibrosis, cardiovascular disease, systemic inflammation

Table 1. Subjects characteristics

	Mean ± SD / N (%)
Age	119.16 ± 63.23
Sex	
Female	7 (% 58.3)
Male	5 (% 41.7)
Weight (kg)	26.79 ± 10.78
Height (cm)	128.66 ± 23.70
BMI (kg/m^2)	15.46 ± 1.51
Mutation	
F508del homozygous	2 (%16.7)
F508del heterozygous	4 (% 33.4)
Others	6 (% 49.8)
Colonization	
-	2 (% 16.7)
+	10 (% 83.3)
Microorganism	
Staphylococcus aureus	5 (% 55.5)
Pseudomonas aeruginosa	7 (% 77.7)

Table 2. Laboratory data and pulmonary function tests

Variable	Visit 1	Visit 2	P
White blood cell count (10^3/µL)	12.75 (n:12)	9.35 (n:7)	0.01
Absolute neutrophil count (10^3/µL)	8.45 (n:12)	4.58 (n:7)	0.02
C-reactive protein (mg/dl)	1.4 (n:12)	0.36 (n:7)	0.1
Spirometry, n:8			
FEV1 (%)	51.75 ± 20.54	67.12 ± 28.60	0.01
FVC (%)	50.12 ± 17.63	63.12 ± 25.46	0.02
FEV1/FVC	101.62 ± 11.62	104 ± 11.23	0.27
PEF (%)	72.62 ± 23.37	82.12 ± 34.46	0.32
FEF (%)	61.75 ± 50	78.25 ± 51.30	0.01

Table 3. Haemodynamic data

Variable	Visit 1	Visit 2	P
Heart rate	105.91 ± 19.31	95.66 ± 17.62	0.15
Peripheral			
SBP (mmHg)	113.25 ± 12.89	110.91 ± 14.2	0.58
DBP (mmHg)	64.75 ± 10.93	58.91 ± 11.96	0.28
MAP (mmHg)	86.91 ± 10.81	82.75 ± 10.53	0.34
PP (mmHg)	48.5 ± 9.5	52 ± 15.60	0.58
Central			
SBP (mmHg)	98.91 ± 15	94.75 ± 12.42	0.38
DBP (mmHg)	67 ± 10.77	61.16 ± 11.63	0.24
PP (mmHg)	31 ± 8.5	33.58 ± 10.13	0.4
AP (mmHg)	6.5	6.9 ± 4.18	0.68
AIx (%)	40.41 ± 8.83	32.5 ± 9.8	0.07
PWV (m/s)	4.51 ± 0.48	4.48 ± 0.45	0.85

SBP; systolic blood pressure, DPB; diastolic blood pressure, MAP; mean arterial pressure, PP; pulse pressure, AP; augmented pressure, AIx; heart rate adjusted augmentation index, PWV; pulse wave velocity

OP-04 [Clinical Characteristics of CF in the Middle East]

INFECTION CONTROL KNOWLEDGE AND PRACTICES AMONG CYSTIC FIBROSIS PATIENTS AND THEIR FAMILIES

Can Akın[1], Ayça Çakan[1], Eren Sarıoğlu[1], Batuhan Başçı[1], Gamze Alçi[2], Ayşegül Karahasan[2]
[1]Marmara University Faculty of Medicine,Istanbul
[2]Marmara University Faculty of Medicine, Medical Microbiology Department, Istanbul

AIM: We searched how well patients and families understand the principles of infection control and what are the knowledge, attitudes, and practices of CF patients and their families.

METHOD: A 24 questioned survey was performed to investigate demographic properties and knowledge about infection control in CF patients. SPSS 24.0 was used to analyse data, chi square test was performed as significance measurement (p<0, 05).

RESULTS: Between November 2018 to January 2019, a total of 120 CF patients was included. Demographic features were given in Table 1. Fifty percent of the patients were colonised by Pseudomonas aeruginosa colonization was detected in half of the patients. Vast majority of the patients knew that pathogens can be transmitted between the patients and infections could be reduced by using a mask, handwashing and by cleaning nebulizers (Table 2). Despite the fact that health workers give training on using a nebulizer, 35.3% of the patients do not have a spare nebulizer. Twenty three percent of respondents agreed that CF patients should be at least 2 metres apart, should avoid close contact with others with CF in the clinic.

CONCLUSION: Health workers play a key role in the patient's awareness. Infection measures should be best explained to the patients and their relatives during their hospital stay. It is necessary to organize training seminars on this subject and to distribute brochures. Shortening the time spent in the hospital, avoiding contact with colonized patients and minimizing the risk of cross-transmission will improve the quality of life of patients.

Keywords: Infection Control, Survey, Cystic Fibrosis

Table 1. Infection Control Knowledge of Survey Respondents

Characteristics		PSE+	PSE-	P
Median age of CF patients	10,49	11,36	9,26	
Median age categories (years)				
age<18 (n:100)	8,16	9,2 (n:58)	6,71 (n:42)	
age>18 (n:19)	22,78	21,75 (n:12)	24,57 (n:7)	
Person completing survey				
Mother/Father	84,1%	58	42	
Person	15,9%	12	7	
Monthly income of patients' parents				
1600-2000 TL		22	14	
2000-4000 TL		35	16	
>4000 TL		10	17	
No specific income		3	2	
Educational status of Mother				
Uneducated		2	1	
Primary School		36	18	
High School		21	14	
Associate's Degree		1	5	
Bachelor's Degree		10	11	
Educational status of Father				
Uneducated		0	1	
Primary School		36	16	
High School		21	14	
Associate's Degree		3	2	
Bachelor's Degree		10	16	
Number of CF clinic visits in past year				0,001
3-9	63,1%	35	40	
9+	36,9%	35	9	
Hospitalizations in recent year				0,048
yes	49,5%	40	19	
no	50,5%	30	30	
Time spent in hospital (min)				
0-30	4,2%	3	2	
30-120	16%	7	12	
120+	79,8%	60	33	
Mother had smoked while pregnant	6,7%	4	4	
Smoking in home	26,1%	17	14	
Cystic fibrosis education				
Healthcare worker	62,2%	43	31	
Internet	31,1%	21	16	
Family	4,2%	4	1	
Friends	2,5%	2	1	

Table 2. Infection Control Knowledge of Survey Respondents

	Yes	No	No Idea
Patients with CF are able to transmit microbes to each other.	88,2% (n=105)	9,2% (n=11)	2,5% (n=3)
Use of masks in hospital reduces the risk of transmission among the patients.	85,7% (n=102)	10,1% (n=12)	4,2% (=n5)
Hand washing protects us from diseases.	95,8% (n=114)	3,4% (n=4)	0,8% (n=1)
There is no need to wash hands if there is no visible dirtiness.	97,5% (n=116)	0,8% (n=1)	1,7% (n=2)
Do you wash your hands regularly?	88,2% (n=105)	10,1% (n=12)	1,7% (n=2)
Do you clean up your nebulizer regularly? (After every use)	87,4% (n=104)	8,4% (n=10)	
Do you dry your nebulizer after you clean up?	93,3% (n=111)	2,5% (n=3)	
Do you have an extra nebulizer?	60,5% (n=72)	35,3% (n=42)	
I pay attention to the fact that there should be approximately 2 meters of distance between my CF relative and the other patients.	75,6% (n=90)	23,5% (n=28)	0,8% (n=1)

OP-05 [Clinical Characteristics of CF in the Middle East]

NON-TUBERCULOUS MYCOBACTERIAL INFECTIONS IN CHILDREN WITH CYSTIC FIBROSIS- A CYSTIC FIBROSIS CENTER EXPERIENCE IN TURKEY

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BACKGROUND-AIM: Non-tuberculous mycobacteria are organisms that found in environment and can cause chronic lung infections in the context of structural lung disease such as cystic fibrosis(CF).In this study, we aimed to evaluate the features of 5 patients with CF who were diagnosed non-tuberculous mycobacterial pulmonary disease (NTM-PD).

MATERIAL-METHODS: CF patients diagnosed with NTM-PD between 2005 and 2019 were included. Clinical features of the patients, treatment modalities and response to treatment were evaluated retrospectively.

RESULTS: 5 patients were reported. The mean age of the patients were 24.8±11.4 years and 3 of them were male. All of the patients were colonised with Pseudomonas aeruginosa. The mean age of the detection of non tuberculous mycobacteria was 17.2±6.9 years and Mycobacterium abscessus was isolated from all of the patients. Mean FEV1 before and after treatment were 58.8±16.6 and 61.2±22.4, respectively. In the intensive phase of the treatment; all of the patients had received intravenous amikacin and oral clarithromycin. All of the patients had received nebulised amikacin for the continuation phase of the treatment. Mycobacterium abscessus were eradicated in 3 patients. The mean treatment duration was 2.6±1.5 years. Side effects related with NTM medications were amikacin induced hearing-loss in one patient and elevated liver enzymes in 2 patients.

CONCLUSION: NTM-PD is a major threat for patients with CF and associated with a rapid decline in pulmonary functions. Diagnosis, management and treatment of NTM-PD still remains a major challenge for clinicians and new studies are needed for better outcomes.

Keywords: cystic fibrosis, non-tuberculous mycobacteria, pulmonary disease

OP-06 [Clinical Characteristics of CF in the Middle East]

CYSTIC FIBROSIS AND SINONASAL PROBLEMS

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Although the complications of cystic fibrosis (CF) on the lung and pancreas are well known, Its effects on the upper airways system are less known. The sinus diseases (chronic rhinosinusitis (CRS) with or without nasal polyposis) are the most common reason for applying to the otorhinolarygolojists. We evaluated endoscopic examinations, computerized tomographic (CT) findings and sinonasal symptoms in Turkish CF patients and aimed to determine the prevalence of sinonasal pathology and to find the relationship between these three clinical features in Turkish CF patients. CF patients underwent a prospective evaluation about sinonasal symptoms using a questionnare. They also underwent nasoendoscopy and paranasal sinuses CT scan. Ninety CF patients with a positive diagnosis of CF were offered an Ear Nose Throat (ENT) examination between April 2018 and December 2018. Sixty-two CF patients attended for ENT review. Mean age was 15.25 years (range 12–20 years). Thirty-five patients were male and thirty-seven were female. Thirty-six (58%) patients reported no sinonasal symptoms and 26 (42%) patients reported sinonasal symptoms. Forty-five CF patients (72.6 %) had a positive finding on nasal endoscopic examination including eleven CF patients (17.7 %) with nasal polyposis. 24 patients had a CT scan. 96% of patients CT scanned have positive findings on CT scan. There was no significant difference between asymptomatic and symptomatic CF groups with respect to their endoscopic score or their CT score.

Keywords: Cystic fibrosis, sinonasal problems, nasal polyposis,nasal endoscopy

OP-07 [Best 6 Abstracts on Cystic Fibrosis]

MANAGING THE RESPIRATORY CARE OF CHILDREN WITH CF IN GAZA

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OBJECTIVES: Examining the respiratory features of children with cystic fibrosis(CF) in Gaza and highlighting the current lacunae in Gaza's CF treatment

METHODS: This is a retrospective cross-sectional study of children (aged ≤ 18 years) with CF who are managed at 3 hospitals in Gaza. Medical record data, including respiratory pathogens, lung functions, chest CTs and hospital admissions, were analysed via SPSS 25.

RESULTS: The records of 92 CF patients aged ≤ 18 years were reviewed, with 76 < age 12 and 53 being male.When diagnosed, all patients were started on long-term nebulised gentamycin, irrespective of their microbiology results. None were on regular nebulised mucolytic or hypertonic saline treatment, and no respiratory physiotherapy input was available. A sputum culture was obtained from 23 patients, 15 of them colonised as follows: 9 with Pseudomonas aeruginosa, 4 with Streptococcus viridians and 2 with MRSA. Spirometry was performed on 32 patients. The median predicted FEV1% was 65. Of the 15 patients who had chest CTs, 5 showed bronchiectasis. Thirty-one patients required hospital admission over the last year for respiratory exacerbations. The average hospital admission was 15 days/patient. IV antibiotics' choices were mainly based on drug availability. In 20% of the admissions, a single IV antibiotic was used. All patients were admitted to multiple occupancy rooms.

CONCLUSION: Our study highlighted several barriers to managing CF in Gaza, including no national CF management guidelines, a lack of a multidisciplinary team approach, especially regarding the respiratory physiotherapist, poor infection control measures and diagnostic/therapeutic insufficiency, including mucolytic treatment and necessary antibiotics.

Keywords: Cystic Fibrosis, Pediatric, Respiratory

OP-08 [Best 6 Abstracts on Cystic Fibrosis]

DISTRIBUTION OF MICROORGANISMS ISOLATED FROM CYSTIC FIBROSIS PATIENTS

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AIM: Identification of microorganisms present in the Cystic fibrosis (CF) lung is important for microbiological surveillance, which guides the antibiotic therapy that plays an important role in maintaining the CF patients in stable clinical condition. We retrospectively analyzed culture results of respiratory samples from CF patients followed in CF clinic during a one year period.

MATERIAL-METHOD: Respiratory samples including sputum, deep pharyngeal swab (DPS), lavage and aspirate materials were inoculated into medium and identified by conventional methods. Results were scanned through Laboratory Operating System.

FINDINGS: Median age of the patients was 17 with a range of 0-43 (Only 3 patients older than 30 years of age). Among these patients, 114 (45.1%) were female. A total number of 1214 respiratory samples from 253 CF patients were included in the study. Distribution of samples were shown in Table 1, consisting DPS samples in 58.5%. Single pathogen were isolated in 71%, double pathogens in 26% and triple pathogens in 3% of the samples. Microorganisms isolated from respiratory samples were given in Table 2. The most common pathogen was Pseudomonas aeruginosa (45.7%), followed by Staphylococcus aureus (36%). The highest colonization rate for bacterial pathogens was detected in the age groups of 6-15 as shown in Table 3.

CONCLUSION: The main causes for the high morbidity and mortality in patients with CF are recurrent respiratory infections. Respiratory samples from patients followed in CF clinics should be monitored for the presence of microorganisms in order to start proper treatment to prevent chronic colonisation and persistant infection.

Keywords: Cystic Fibrosis, Microorganisms, Etiology, Microbiology, respiratory samples

Table 1. Distribution of respiratory samples from CF patients

Sample Type	(n/%)
Deep Tracheal Aspirate	2 (0.17%)
Bronchoalveolar Lavage	2 (0.17%)
Endotracheal Aspirate	4 (0.35%)
Sputum	495 (40.8%)
Deep Pharyngeal Swab	711 (58.5%)
TOTAL	**1214 (100%)**

Table 2. Number of isolated microorganisms from respiratory samples

Pseudomonas aeruginosa	526
Staphylococcus aureus	261
Methicillin resistant S: aureus	153
Achromobacter spp.	50
H. influenzae	47
Stenotrophomonas maltophilia	37
Escherichia coli	17
Burkholderia cepaciae	12
Serratia marcescens	10
Acinetobacter baumanii	9
Klebsiella pneumoniae	8
Enterobacter spp.	7
Aspergilus fumigatus	5
Candida spp.	5
Streptococcus sp	4

Table 3. The most common bacterial pathogens according to the age groups

	0-6	6-15	15-30
Pseudomonas aeruginosa	145	200	165
Staphylococcus aureus	121	194	100
Haemophilus influenzae	1	31	15

OP-09 [Best 6 Abstracts on Cystic Fibrosis]

RESPIRATORY VIRUS ANALYSIS IN CYSTIC FIBROSIS PATIENTS WITH ACUTE RESPIRATORY EXACERBATIONS

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BACKGROUND-AIM: Acute viral respiratory tract infections in patients with cystic fibrosis (CF) play a significant role as causing exacerbations of the disease, a decrease in lung function and often lead to hospitalisations. In this study, we aimed to evaluate the viral detection results of the CF patients during acute exacerbation.

METHODS: CF patients applied with acute upper respiratory tract infection symptoms and fewer were included to the study. Total 37 respiratory samples from 21 patients with CF were analyzed by using the Film Array Respiratory Panel 2.0 (Biomérieux, France). This assay can detect 20 of the most common upper respiratory pathogens simultaneously by using mutiplex nested PCR based microarray method.

RESULTS: One or more respiratory pathogen was detected in 15 CF patients. Mean age of the patients was 11.13 ± 10.04 years old and 66.7% were girls. The mean FEV1 was 58 ± 16 % predicted. 86.7% of the patients were colonized with Pseudomonas aeroginosa whereas chronic MRSA growth was 33.3%. Twelve (80%) patients had pancreatic insufficiency. One or more viral patogens detected as positive in 51,3% (19/37) of samples. Rhinoviruses/Enteroviruses were the most prevalent pathogens encountered in the study (30,4%), followed by Corona viruses (21.7%), RSV (13,0%), Parainfluenza (13,0%) and Influenza A viruses (13,0%) respectively. The investigated bacterial pathogens (B.pertussis, C.pneumoniae, and M.pneumoniae) were not detected in samples.

CONCLUSION: Rapid identification of the pathogens in respiratory tract infections is important for patient management and isolation measures.

Keywords: viral patogen, cystic fibrosis, exacerbation

Table-1

Respiratory Pathogens	n (%)
Rhinovirus/Enterovirus	7 (30,4)
Coronavirus	5 (21,7)
Respiratory synscytial virus	3 (13,0)
Parainfluenza virus	3 (13,0)
Influenza A virus	3 (13,0)
Human metapneumovirus	1 (4,3)
Adenovirus	1 (4,3)
Total	23 (100,0)

Pathogens detected in respiratory samples

OP-10 [Best 6 Abstracts on Cystic Fibrosis]

IMPORTANCE OF ISOFORMS OF IMMUNOREACTIVE TRYPSIN IN NEWBORN SCREENING FOR CYSTIC FIBROSIS

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Cysticfibrosis(CF)isthemostfrequentautosomalrecessivehereditarydiseaseamongCaucasians(1:2,000to3,500).Immunoreactive Trypsin (IRT) is used for determination and screening in newborns as its level increases in CF. IRT exists as 3 isoforms: IRT1, IRT2 and IRT3 (mesotrypsin), where IRT1 and IRT2 are used as a marker of diagnosis of CF as well as diagnosis of other pancreatic diseases.

IRT1 form is the most used in Newborn screening for CF in most countries. However, in the result of some researches, IRT2 has been more closely aligned with CF. Also, IRT tests have been plagued by a high rate of false-positive results (screen positive and diagnosis negative) and faces the problem of determination of inappropriate cut-off values in multi-ethnic communities. Real risk of a false CF positivity reaches even the 30% of newborns in some ethnic origins. To avoid an unacceptable stress for families and a waste of medical and economic resources, it is important to use optimized systems for identification of correct IRT form and determination of cut-off values for each region.

During this research we have developed ELISA assays specific for IRT1 and IRT2. Our aim is to perform a comprehensive research to determine the profile of IRT values in CF in Turkey.

Acknowledgements: This study was supported by TÜBİTAK 1003-SBAG, project number 115S124

Keywords: Cystic fibrosis, newborn screening, monoclonal antibodies, ELISA, immunoreactive trypsinogen(IRT)

OP-11 [Best 6 Abstracts on Cystic Fibrosis]

THE EFFECT OF THE FAMILY EMPOWERMENT PROGRAM ON DISEASE MANAGEMENT SKILLS OF PARENTS WITH CHILDREN WITH CYSTIC FIBROSIS: A RANDOMIZED CONTROLLED TRIAL

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OBJECTIVE: Parents with children with cystic fibrosis (CF) experience many problems about disease management. Family empowerment programs increase the knowledge and skills of parents on disease management. The aim of this study is to determine the effect of nursing intervention based on the family empowerment program on parents' management of CF.

MATERIAL-METHOD: A single-blinded, parallel group randomized controlled trial with a pretest and posttest design was used. The participants were randomly assigned to the experimental (n=24) and control (n=24) groups. Nursing interventions applied to the experimental group for ten weeks with the aim of empowering families include providing an individualized training and a booklet concerning disease management, making phone calls and providing consultancy services for the purpose of reinforcing the training and increasing the motivation of parents. A routine polyclinic follow-up was applied to the control group. As the data collection tool, an information evaluation questionnaire prepared by the researcher by obtaining expert opinion was used. The Wilcoxon Signed Ranks Test and the Mann-Whitney U Test were used for in-group and inter-group analyses, respectively.

FINDINGS: The comparison between the experimental group and the control group after the intervention showed that the correct response mean (46.0 ± 2.54) of the experimental group increased compared to the control group (35.3 ± 8.95). It was further revealed that the nursing interventions were effective (p<0,001) and the effect size was high (d:1.627 [CI: 0.934, 2.305]).

CONCLUSIONS: Family empowerment interventions increased the disease management skills of the parents of children with CF.

Keywords: cystic fibrosis, child, parents, disease management, Nursing interventions, empowerment

OP-12 [Best 6 Abstracts on Cystic Fibrosis]

OUR EXPERIENCE IN CYSTIC FIBROSIS NEWBORN SCREENING PROGRAMME

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OBJECTIVE: Cystic fibrosis (CF) newborn screening programme (NSP) began in 2015 in Turkey. Newborns with high immunoreactive trypsinogen (IRT) values are identified and directed to the centers for sweat chloride testing. In this study, we aimed evaluate sweat test results of patients with high IRT values.

MATERIAL-METHOD: The data of children with high IRT values between 2015-2018 were evaluated retrospectively in three pediatric pulmonology centers. Patients' age, IRT values and sweat test results were noted.

FINDINGS: Between January 2015-December 2018, 1432 children admitted to the pediatric pulmonology departments with high IRT values. Sixty (4.1%) patients were diagnosed with CF. The mean age of the patients was 70.9±57.9 days. The mean of the first IRT values were 227.5 ± 111.4 (min: 90, max: 565.8) and the second IRT values were 194.9 ± 104.3 (min: 63.2, max: 479) in the CF patients. The mean sweat test results of the patients were Cl: 82.4 ± 16.7 mEq/L.

CONCLUSIONS: The ideal dried blood spots time for NSP in newborns is days 3 and 7. In our study, the time of arrival of the patients with high IRT values to our centers was approximately between 22-180 days. It was found that the patients reached the centers for sweat chloride testing quite late despite the determination of the IRT results and it causes late diagnosis of these patients.

Keywords: child, cystic fibrosis, newborn screening programme

OP-13 [Clinical Characteristics of CF in the Middle East]

IMPACT OF BODY COMPOSITION AND NUTRITIONAL STATUS ON HEALTH OUTCOMES IN ADULT CYSTIC FIBROSIS PATIENTS

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AIM: Cystic fibrosis (CF) is a multi-systemic disease. Chronic inflammation and malabsorption may affect body composition, respiratory function and health outcomes. Our aim was to examine anthropometric measures, microbiologic data, spirometric data, comorbidities and how body composition may impact health outcomes in adult CF patients.

METHODS: 31 adult CF patients (15 female, 16 male) with mean age 25 (18-42) and 23 controls were enrolled. The modified Medical Research Council (mMRC) scale, 6 minute-walk-test and pulmonary function test were performed. Anthropometric data including body mass index (BMI), fat-free mass index (FFMI) and total body fat percentiles were measured. Clinical indices, presence/absence of pancreatic insufficiency and method of nutrition were recorded.

RESULTS: Mean duration of disease was 18.6±7.5 years. 24 of the patients (77%) were below targeted BMI. Mean BMI of the patients was 20.6±3.1 kg/m2 and mean FFMI was 16.4±1.6 kg/m2. When compared to control group, both BMI and FFMI were lower in CF patients (p<0.05 and 0.00) and both were correlated with age (p: 0.008, r: 0.468 and p: 0.036, r:0.377). Female patients had lower FFMI (15.7±1.3 vs17.1±1.6, p:0.01) and higher fat percentile (22.7% vs 12.5%, p:0.00) than male patients. FFMI was found to be positively correlated with FEV1L and FVC L (p:0.017, r: 0.025 and p:0.05 and r: 0.490, respectively).

20 patients (65%) had pancreatic insufficiency and all were under pancreatic enzyme replacement medication. This group of patients had lower fat percentile then the group without pancreatic insufficiency (15.0±6.7% vs 23.2±9.0%; p:0.011)

16 patients (52%) had anemia and FFMI positively correlated with hemoglobin values (p:0.03, r: 0.390). 17 patients (55%) were receiving oral nutritional support and these patients had lower BMI then those who were not receiving (19.4±2.6 vs 22.1±2.9, p: 0.013).

CONCLUSION: The prevalence of malnutrition was high in adult CF patients and lung function is correlated with nutritional status. Periodic measurement of anthropometrics and close monitoring of nutritional status may improve respiratory function and health outcomes in adult CF patients

Thematic Poster Presentations

PP-01 [Microbiology and Immunology in CF]

EVALUATION OF MICROORGANISMS AND THEIR RESISTANCE IN THE RESPIRATORY TRACT SPECIMENS OF PATIENTS WITH CYSTIC FIBROSIS AND THE CORRELATION OF ANTIBIOTIC RESISTANCE WITH CLINICAL FINDINGS AND PULMONARY FUNCTION TESTS

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OBJECTIVE: Antibiotics have an important role in cystic fibrosis (CF) lung disease. Antibiotic resistance, especially multi-drug resistance (MDR) increases in CF. This study aimed to determine whether there is a change in antibiotic resistance in years and to evaluate the relationship between the antibiotic resistance and patients' clinical status and pulmonary function tests.

MATERIALS-METHODS: CF patients followed at our pediatric pulmonology clinic who had significant microorganisms in respiratory cultures from August 2010 through December 2016 were included in the study. The findings were analyzed retrospectively.

FINDINGS: 333 respiratory cultures taken from 39 CF patients were analyzed. MSSA was isolated from 122 cultures, P. aeruginosa from 97 cultures and MRSA from 22 cultures. From 2010 to 2016 while MSSA and MRSA incidence increased, P. aeruginosa and other microorganisms' incidence didn't increase. MDR-MSSA and MDR-P. aeruginosa incidence didn't change, but MRSA incidence increased in years. There was a significant relationship between antibiotic resistance development and acute pulmonary exacerbations, pulmonary examination findings, loss in weight and height percentile. As the antibiotic resistance increased, FEV1, FVC, FEV1/FVC, PEF, FEF25-75 were significantly decreased. The use of azithromycin or inhaled antibiotics and the use of oral antibiotics in the last three months were not associated with the development of antibiotic resistance.

CONCLUSIONS: Pulmonary disease remains the leading cause of morbidity and mortality in patients with CF. Therefore; it is important to take respiratory tract cultures with 3-months intervals and at the time of acute pulmonary exacerbation and give rational antibiotic treatment according to the culture results.

Keywords: cystic fibrosis, antibiotic resistance, microorganism

PP-02 [Microbiology and Immunology in CF]

PLASMA IgG4 LEVELS IN CLINICALLY STABLE ADULT CYSTIC FIBROSIS COLONIZED WITH PSEUDOMONAS AERUGINOSA

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BACKGROUND: Chronic cystic fibrosis (CF) lung infection by Pseudomonas aeruginosa (P aeruginosa) increases inflammation and destruction of lung tissue. Plasma immunoglobulin G4 (IgG4) has been associated with chronic infections.

OBJECTIVE: To determine plasma IgG4 level in clinically stable adults CF patients, and examine its correlation with P aeruginosa and lung function.

METHODS: A cross-sectional study. Clinical samples were collected from 19 CF patients and 17 healthy controls. Plasma IgG4 levels were determined using a commercially available Sandwich ELISA kit (Cat# MBS2019882, MyBioSource, Inc., CA, USA)

RESULTS: Patient's age ranged from 18 to 40 years with a mean age of 24.63 ±5.41 years. Seventeen CF patients with homozygous CFTR I1234V and remaining two with other mutations. None of CF patients had CF related liver disease. The mean plasma IgG4 in CF patients 1697.57±786.53 (median 1449.2, range 611.3 to 3308.1) μg/ml significantly higher than healthy control 838.32±534.89μg/mL (median 658.9, range 273.6 to 1844.4), P=0.001. Fifteen CF patients (78.9%) colonized with P aeruginosa. No correlations between the parameters of lung function or P aeruginosa and plasma IgG4 levels. Seven CF subjects (38.8%) had high levels of Plasma IgG4 > 2000 μg/ml. No statistically significant differences were observed in the IgG4 high patients than IgG4normal patients with respect to age, sex, body mass index (BMI), exocrine pancreatic sufficiency, CF related diabetes, P aeruginosa, staphylococcus aureus and FEV1.

CONCLUSION: These findings may suggest a neutral effect of IgG4 as an anti-inflammatory immunoglobulin without effect on P aeruginosa or lung function.

Keywords: Cystic fibrosis, adult, Pseudomonas aeruginos, lung function

PP-03 [Microbiology and Immunology in CF]

MYCOBACTERIUM SIMIAE INFECTION IN CYSTIC FIBROSIS: A CASE REPORT

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Nine years old man was diagnosed as cystic fibrosis at the age of 2 years old. In his last outpatient clinic visit he was hospitalized with symptoms of cough, increased sputum and shortness of breath. In physıcal examination bilateral inspiratory rales were determined. In PA graphy rıght middle and lower lobe ınfıltratıon was seen and piperacilin-tazobactam, amikacin treatment was started. İn his thorax CT examination, there were ground- glass opacity, acinar involvement, air bronchogram, Ghon complex images in the lower and middle lobes of right lung. Atypical mycobacterium was determined in culture. Flexible bronchoscopy was performed. Mycobacterium Simiae was determined in his sputum. İn the antibiogram, it was susceptible to clarithromycin, borderline resistant to amikacin and ethionamide, resistant to rifabutin, ethambutol, streptomycin, rifampicin, linezolid and moxifloxacin. His treatment was planned as amikacin, trimethoprim- sulfamethoaxazole, moxifloxacin and clarithromycin. As the symptoms of the patient were relieved he was discharged and followed up from outpatient clinic. Non tuberculous mycobacteria (NTM) are generally not an infectious agent for healty individuals. Prevalence is not known exactly, however it has been increasing in recent years. NTM are usually found in environment (water, soil, animal etc). The diagnosis is made by culture of at least two samples from the patients with clinical and radiological findings. In treatment at least two of the anti-mycobacterial drugs are used at least 18-24 months. NTM infections are rare in patients with cystic fibrosis. Routine ARB and mycobacterium culture in sputum should be performed, especially in patients with poor prognosis.

Keywords: cystic fibrosis, non tuberculous mycobacteria, mycobacterium simiae

PP-04 [Microbiology and Immunology in CF]

EPIDEMIOLOGY OF PSEUDOMONAS AERUGINOSA AMONG CYSTIC FIBROSIS PATIENTS

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OBJECTIVE: Pseudomonas aeruginosa is a microorganism responsible for high morbidity and mortality in patients with Cystic Fibrosis (CF). In order to reduce the severity of the infection in CF, it is crucial to start empirical antibiotic therapy before getting the antibiotic susceptibility results and facility-specific antibiograms are very useful for clinicians in this manner.

MATERIAL-METHOD: Respiratory samples including sputum, deep pharyngeal swab (DPS), lavage and aspirate materials from 253 CF patients were inoculated into culture plates and identified by conventional methods. Antimicrobial susceptibility tests were performed according to EUCAST.

RESULTS: P.aeruginosa was isolated from 522 samples of 154 (60.3%) patients between January-December 2018 (Table 1). Median age of the P. aeruginosa isolated patients was 17 with a range of 0-43 (Only 3 patients older than 30 years of age). Among these patients, 51 (33.2%) were female. P. aeruginosa was isolated as a single pathogen in 61, 8% of the patients. High rates of resistance was detected to third-generation cephalosporins, aminoglycosides and quinolones that are often preferred in treatment and prophylaxis (Table 2).

CONCLUSIONS: Antimicrobial treatment play one of the most important roles in the survival of CF patients. In our institution, the first choice antimicrobials in severe acute exacerbations is the combination of ceftazidime-amikacin. Oral ciprofloxacin is commonly used for prophylaxis and treatment in mild exacerbations. The susceptibility to ceftazidime and amikacin decreased significantly in years, probably due to excessive usage. Close monitoring of the patients and wise usage of antimicrobials is essential for CF patients.

Keywords: Pseudomonas aeruginosa,Cystic Fibrosis, Antimicrobial susceptibility

Table 1. Distribution of *Pseudomonas aeruginosa* according to sample types

Sample types	n (%)
Sputum	301 (57)
Deep pharyngeal swab	220 (42.1)
Bronchoalveolar lavage	1 (0.1)
Total	522

Table 2. Antimicrobial susceptibility rate *Pseudomonas aeruginosa* strains

	%S									
No strains	PIP	TZP	CAZ	AN	GN	NN	CIP	FEP	MEM	IMP
522	%76,5	%70	%60	%48,6	%53,6	%72,1	%54,8	%65,8	%66,7	%57,6

PIP: Piperacillin, TZP: Piperacillin-Tazobactam, CAZ: Ceftazidime, AN: Amikacin, GN: Gentamicin, NN: Tobramycin, CIP: Ciprofloxacin, FEP: Cefepime, MEM: Meropenem, IMP: Imipenem

PP-05 [Microbiology and Immunology in CF]

GENOTYPIC RELATEDNESS OF PSEUDOMONAS AERUGINOSA STRAINS ISOLATED FROM SPUTUM SAMPLES OF CYSTIC FIBROSIS PATIENTS IN MARMARA UNIVERSITY HOSPITAL CF CENTER

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OBJECTIVE: Persistent airway infection by Pseudomonas aeruginosa among patients with cystic fibrosis (CF) is accompanied by deteriorating pulmonary function and reduced survival. The most likely source is believed to be from the environment where individual patients acquire their own unique strains. Molecular methods with high reproducibility and good discriminatory power must be used to compare genotyping relatedness.

MATERYAL-METHOD: During a 3 months period between November 2018-January 2019; 67 P. aeruginosa strains were isolated from 120 patients followed in our clinic. Conventional culture media was used for inoculation of samples and bacterial identification was done by MALDI-TOF (Biomerioux,France) method. Antimicrobial susceptibility test was performed according to EUCAST criteria. In order to detect genotyping similarity between patients, we have chosen 37 multidrug resistant strains and one isolate per patients was included. DNA was extracted by using the DNA purification kit (GENMARK). ERIC-PCR method with ERIC-1 and ERIC-2 primers was used for genotyping. PCR products were visualized in agarose 1,5% gels stained with ethidium bromide.

RESULTS: Antimicrobial susceptibility results is given in Table. We have an increasing rate of resistance to antibiotics that are commonly used in treatment. As we seen in Figure,there was no predominance between patients genotypically.

CONCLUSION: Cross-infection between patients has been detected in some CF clinics, suggesting the emergence of transmissible strains Nevertheless, these observations are regarded as unusual and insufficient to warrant additional infection-control measures We suggested that cross-infection is not common in our outpatient clinic and a common source of acquisition is unlikely.

Keywords: Pseudomonas aeruginosa, Genotyping, ERIC-PCR, Epidemiology, Cross-infection

Figure : Genotyping relatedness among *Pseudomonas aeruginosa* strains

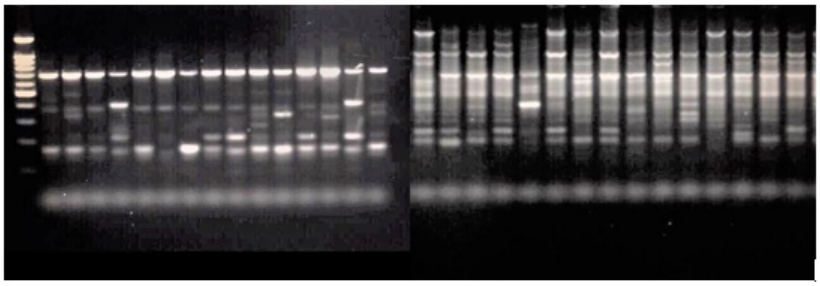

Table 1. Antimicrobial susceptibility rate *Pseudomonas aeruginosa* strains

	%S								
No strains	TZP	CAZ	AN	GN	NN	CIP	FEP	MEM	IMP
67	56,1%	63.6%	40.1%	42.4%	65.1%	53.1%	54.5%	45.4%	49.4%

TZP: Piperacillin-Tazobactam, CAZ: Ceftazidime, AN: Amikacin, GN: Gentamicin, NN: Tobramycin, CIP: Ciprofloxacin, FEP: Cefepime, MEM: Meropenem, IMP: Imipenem

PP-06 [Microbiology and Immunology in CF]

NON-TUBERCULOSIS MYCOBACTERIA IN CYSTIC FIBROSIS PATIENTS

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OBJECTIVE: Non-tuberculous mycobacteria(NTM) can cause chronic lung infection particularly in patients who have structural lung disease or immunodeficiency.We evaluated the prevalence and management of NTM infections in CF patients.

Material-METHOD: A retrospective analysis was carried out on the data collected between 2000-2018.CF patients having at least one positive NTM isolate were included and patients' laboratory, spirometry results, sputum culture, chest radiology findings, comorbidity status, along with demographic characteristics and past treatment history were reviewed.

RESULTS: NTM grew in the sputum of 9 CF patients.Acid-Fast Bacilli smear was positive in 6 out of 9 patients at first isolation of NTM.The mean age of the patients at the time of first NTM-positive culture was 18.89 years.Eight patients had Mycobacterium abscessus growth, one patient had Mycobacterium szulgai in their sputum.Five NTM-positive CF patients received antibiogram directed therapy.Two patients received antimycobacterial therapy for 12 months, one patient for 6 months and one patient for more than 12 months.HRCT scans obtained after the first NTM-positive culture were available in 8 patients.Consolidation, multifocal bronchiectasis, tree-in-bud findings and hilar, mediastinal lymphadenopathy were the most common HRCT findings. The mean of FEV1 at first NTM-positive culture, at the end of the treatment were 65%, 70%, respectively. NTM eradication was observed in 2 patients, chronic colonization was observed in 2 patients, one patient has still been receiving therapy for 4 months.

CONCLUSIONS: Diagnosis and treatment of NTM infections are important for proper functioning of lungs and prognosis of CF patients.Further studies are needed for effective and appropriate management of NTM infected CF patients.

Keywords: Non-tuberculosis Mycobacteria, NTM, cystic fibrosis, Mycobacterium abscessus

PP-07 [Different aspects of CF]

A CASE OF ALLERGIC FUNGAL RHINOSINUSITIS AT CYSTIC FIBROSIS

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Allergic fungal rhinosinusitis (AFRS) is a subtype of chronic rhinosinusitis that develops as a result of fungal sensitization. A 14-year-old boy with cystic fibrosis and allergic rhinitis was admitted with productive cough. On physical examination, serous post-nasal discharge and fine rales were detected. Chest X-ray revealed similar results with previous radiographs such as peribronchial thickening and hyperaeration. The patient received antibiotic treatment consider of pulmonary exacerbation due to low pulmonary function. In follow-up, the patient had nasal congestion and green nasal secretions. In rhinoscopy, nasal polyps were seen in the left middle meatus and mold hyphae were seen in the pathology of the nasal secretions. Paranasal sinus CT revealed high-density masses concordant with fungal sinusitis. On the maxillofacial MRI, low signalization was observed in all sinuses, more prominently in the left sphenoidal sinus. In laboratory, TIgE and A. Fumigatus-spIgE were found to be 2678 IU/mL and 40.1 kUA/l, respectively. A. fumigatus skin prick test (SPT) enduration 9mm. Presence of nasal polyp, IgE elevation, fungal susceptibility in SPT and characteristic radiological findings were concordant with AFRS. Endoscopic sinus surgery was performed. In histopathological examination, fungal hyphae embedded in fibrin was not invading the mucosa. Postoperative systemic and nasal steroid treatment was started to prevent recurrence. In CF patients with nasal symptoms, nasal polyps and AFRS should be considered, although the latter is quite rare. For optimal diagnosis and treatment of AFRS involve surgical approaches. AFRS frequently recurs in patients not treated with glucocorticoids after surgery.

Keywords: Allergic, fungal, sinusitis

PP-08 [Different aspects of CF]

EVALUATION OF BONE METABOLISM IN CHILDREN AND ADULTS WITH CYSTIC FIBROSIS BY USING FALANGEAL QUANTITATIVE ULTRASOUND

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OBJECTIVE: Low bone mineral density is common in children and adults with cystic fibrosis (CF). Cystic fibrosis transmembrane regulatory protein (CFTR) dysfunction is known to have direct effects on bone cell activity. The aim of this study was to investigate the relationship between bone density by using ultrasonographic bone densitometer measurement in in children and young adults with CF.

METHOD: This prospective case-control study included 40 patients with CF and 35 healthy controls. In the patient and control groups, amplitude-bound volume (Ad-SoS), wide-band ultrasound attenuation (BUA) and T-score levels were measured and the hardness index (SI) was calculated.

RESULTS: Ad-SoS levels were significantly lower in patients with CF than in healthy controls ($p < 0.001$). In patients with CF, BUA ($p < 0.001$), T-score ($p < 0.001$) and SI ($p < 0.001$) were significantly lower compared to the healthy control group. There was a positive correlation between Ad-SoS age and age in patients with CF ($r = 0.514$, $p < 0.001$).

CONCLUSION: Our study results suggest that bone ultrasound measurements may be a reliable marker for CF-related bone diseases and may reflect the quality and quantity of the skeletal system.

Keywords: Bone ultrasound, bone density, cystic fibrosis

PP-09 [Different aspects of CF]

EVALUATION OF THE FACTORS THAT AFFECTING QUALITY OF LIFE OF CHILDREN WITH CYSTIC FIBROSIS

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BACKGROUND AND AIMS: Cystic fibrosis (CF) is a chronic disease affecting both the child and the family system. The aim of this study was to evalute the factors that affect the quality of life of children with CF.

METHOD: The study included 40 children with CF and the mothers of 40 children with CF. The sociodemographic information and Cystic Fibrosis Questionnaire-Revised (CFQ-R;), form were used in the evaluation of the subjects.

RESULTS: The CFQ-R scores of the parents of the CF patients who were admitted to the hospital were higher than the mothers of non-hospitalized children. Additionally; there was a statistically significant difference in the treatment and nutritional scores in the between two groups. Also there was a statistically significant difference only in the physical functioning parameters between the patients who were hospitalized and non-hospitalized.

CFQ-R scores of the parents of CF patients who were diagnosed with PA infection were higher than the mothers of children without PA infection. There was a significant correlation in vitality, treatment, health, body image, and digestion scores. The CF patients with BMI below 25th percentile were lower physical functioning, vitality, body image, weight, nutritional symptoms and school performance scores than the patients with BMI above 25th percentile.

CONCLUSION: Pseudomonas aeruginosa infection, hospitalization and malnutrition negatively affected the quality of life of the CF patients and mothers of children with CF. There is a need for further studies and developing programs that will provide psychosocial support for mothers of children with CF in our country.

Keywords: Cystic fibrosis, quality of life, children and parents

PP-10 [Different aspects of CF]

ALLERGIC BRONCHOPULMONARY ASPERGILLOSIS IN PATIENTS WITH CYSTIC FIBROSIS: 13 YEAS EXPERIMENT AT ONE CENTER

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BACKGROUND: With new diagnosis and treatment methods, life expectancy of patients with cystic fibrosis (CF) is prolonged, however complications are increased. One of these complications is allergic bronchopulmonary aspergillosis (ABPA), which is a hypersensitivity reaction to the antigens of Aspergillus fumigatus. Our aim was to evaluate the clinical features, treatment modalities and outcomes of patients with cystic fibrosis who were diagnosed with ABPA in our clinic.

MATERIALS-METHODS: 12 patients, who were followed up from our Cystic Fibrosis Center between 2005 and 2018 years with diagnosis of ABPA were included. Clinical and demographic features of the patients, treatment modalities, complications and outcomes of the treatment were evaluated retrospectively.

RESULTS: The mean FEV1 scores before the diagnosis, at the diagnosis and after the treatment were 81.64±20.7, 65.9±16 and 68.1±19.7, respectively. Oral and pulse steroid, voriconazole, posakonazole, itraconazole and omalizumab were used for treatment. Adverse effects related to steroid were seen in 9 patients; included hyperglicemia, diabetes mellitus, myopathy and cataract. Atypical mycobacterium infection was detected in 2 patients. Side effects related to antifungal medications such as headache, vomiting and visual problems were seen in 2 patients. Remission was seen in 4 patients, 5 patients had multipl ABPA relapses and 2 patients were died because of severe pulmonary exacerbation. 3 patients are still on ABPA treatment.

CONCLUSION: Treatment of ABPA in cystic fibrosis patients is still a major problem because of drug side effects and difficulty of curative treatment. New treatment modalities are needed.

Keywords: Cystic fibrosis, allergic bronchopulmonary aspergillosis, steroid, antifungal, side effects

Table 1.: Demographic features of patients

Female/Male	6/6 (50%/50%)
Average Age	192±48 months
BMI Z Score	-1,8±0,65
Age at Diagnosis of ABPA	129±45 months
Positivity of SPT for ABPA	7/12 (68%)
Colonization with Pseudomonas Aeruginosa	12/12 (100%)
Pancreatic Insufficiency	12/12 (100%)
Azithromycin Prophylaxis	3/12 (25%)
Inhale Steroid Usage	9/12 (75%)
Dornase Alpha Usage	12/12 (100%)
Diabetes Mellitus	1/12 (8%)

Table 2.: Evalution of Respiratory Function Test, Total IgE, Aspergillus Fumigatus Antigen and Radiological findings of Patients

	At the Time of Diagnosis	At the End of the Treartment
FEV1 scores	65.9±16	68.1±19.7,
Total IgE	2451±468	576±147
Aspergillus Fumigatus Antigen	57.2±44	21.6±22.2

PP-11 [Different aspects of CF]

CYSTIC FIBROSIS PRESENTING AS ARTHROPATHY: A CASE REPORT

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INTRODUCTION: Historically, the diagnosis of cystic fibrosis (CF) was made after the patients present with signs and symptoms. But since the introduction of newborn screening, the diagnostic algorithm has changed and patients are being diagnosed earlier with improved survival. This survival gain has lead to emergence of delayed complications like musculoskeletal disorders. However, in countries with limited resources, such disorders can be the presenting features.

Case description: We present a case of arthropathy in a 24 year old male patient who was initially misdiagnosed as juvenile rheumatoid arthritis (JRA) in his childhood and was managed with immunosuppressive drugs for several years. The correct diagnosis of CF was made 10 years later when he was admitted for an arthritis exacerbation and CT scan showed bronchiectasis. This warranted a sweat chloride test (90 mmol/L, 120 mmol/L), and genetic testing (homozygous for W1282X).

DISCUSSION: Approximately 12-20% of CF patient will experience some form of joint disease. The prevalence of musculoskeletal disorders in general increases with age and with severity. Two distinct types of arthritis are described in CF; CF-related arthropathy (CFA) and hypertrophic osteoarthropathy (HPOA). The improved survival of CF patients nowadays might lead to increase in the prevalence of such musculoskeletal disorders. The lack of knowledge about the whole spectrum of CF manifestations can lead to misdiagnosis as in our patient. Therefore, in countries where newborn screening for CF is not available, clinicians should keep a high index of suspicion in order to diagnose patients earlier and not to miss cases.

Keywords: cystic fibrosis, CF-related arthropathy (CFA), hypertrophic osteoarthropathy (HPOA)

PP-12 [Different aspects of CF]

PRESENTATION OF FOUR CASES OF ALLERGIC BRONCHOPULMONARY ASPERGILLOSIS IN CYCTIC FIBROSIS PATIENTS

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Allergic bronchopulmonary aspergillosis (ABPA), a progressive fungal allergic lung disease, is a common complication of cystic fibrosis. We presented four cases of ABPA with cystic fibrosis (CF) We diagnosed these patients according to diagnostic criteria defined by CF Foundation. Median age of our patients was 11.5 (5-15) years. Initial median pulmonary function test results were 38% (29%-58%) for FEV1 of predicted and 35% (34%-51%) for FVC of predicted. Baseline total IgE and Aspergillosis specific IgE levels were 843 (353-1267) IU/ml and 35.0 (0.15-90.0) kUA/l Rast Mtd. Three of the patients were treated with oral prednisolone 2 mg/kg/day as a starting dose and oral ıtraconazole. Steroid dose was decreased according to clinical findings, IgE levels and continued for three months. Fourth patient required pediatric intensive care admission and initialy he was treated iv voriconazole, methylprednisolone and two weeks later treatment switched to oral prednisolone and ıtraconasole. Following treatment there was an improvement in FEV1 and FVC predicted. Median pulmonary function test results were 47% (42%-73%) for FEV1 of predicted and 41% (40%-63%) for FVC of predicted. Posttreatment total IgE and Aspergillosis specific IgE levels were 581 (327-627) IU/ml and 25.0 (14.1-36.4) kUA/l Rast Mtd. The possibility of ABPA should be considered in all CF patients with pulmonary exacerbations who are unresponsive to routine antibiotic treatment.

Keywords: allergic bronchopulmonary aspergillosis, cyctic fibrosis, treatment

PP-13 [Difficulties in management of CF in the Middle East]

A PROFILE OF CHILDREN WITH CYSTIC FIBROSIS IN GAZA

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OBJECTIVES: Examining cystic fibrosis(CF) in children in Gaza, including its epidemiological and genetic features.

METHODS: This is a cross-sectional analysis of children (aged ≤ 18 years) with CF who are managed at 3 hospitals in Gaza. Data, including mortality, diagnostic tools, consanguinity, growth parameters and smoking status, were collected retrospectively from medical records and personal phone calls/interviews. Data were analysed via SPSS 25.

RESULTS: In total, 107 children with CF were identified, with 15 having died within the last 5 years from respiratory failure. Of the 92 living patients studied, 76 were < age 12 and 53 were male.The median age at diagnosis was six months. All patients had an elevated sweat chloride value > 60 mEq/L. Genetic testing was conducted on 45 patients using PCR kit testing for 30 mutations. The most common mutations included homozygous delta F508 in 19 patients.Ninety percent of our sample had either first- or second-cousin parents.All patients were started on Creon at diagnosis.The medians of weight and BMI centiles were 11.5 and 14.5, respectively.At least 1 family member who smokes was identified in 41.7% of our population.

CONCLUSIONS: High premature CF mortality was found, with Delta F508 being the most common gene mutation. Half our sample had no genetic testing due to few resources.The high rate of consanguinity and the lack of newborn screening suggest an under-diagnosed population, making genetic counselling essential. Identification of the CFTR mutation spectrum/frequency among CF patients in Gaza is needed to establish adequate.Introducing faecal elastase testing will support CF diagnosis and determine Creon requirements.

Keywords: Cystic Fibrosis, Diagnosis, Gaza

PP-14 [Difficulties in management of CF in the Middle East]

THE EFFECT OF IVACAFTOR ON ADULT CYSTIC FIBROSIS PATIENTS AT ROYAL HOSPITAL IN OMAN

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OBJECTIVE: Assess role of ivacaftor in CF patients who had specific gated mutation by evaluating response of clinically relevant markers & obtain local data in Oman

METHODS: This study was institutional, single center, retrospective, medical record review of cases age 12 and older. All patients with mutation (P.ser 549 Arg Del at chromosome 7) are eligible for the study. The assessments were performed at baseline (1 year before) and 12 months after initiation of treatment in same group (act as their own control by comparing pre & post treatment). The primary outcome was FEV1%. Sub studies were FVC%, BMI and number of hospitalization due to infective exacerbation

RESULTS: 15 of patients were included in the study. 14 have homogenous and 1 had heterogonous (P.ser 549 Arg & P.phe 508 Del) mutations. All of them have bronchiectasis, 12 have pancreatic insufficiency and 1 diabetic. Mean FEV1% improved from 54.27±25.46 to 59.27±25.25 (p= 0.007). Mean FVC% improved from 63.60±20.02 to 66.93±19.72 (p=0.052. Mean BMI improved from 16.20±3.21 to 17.87±3.34 (p= 0.001). Median number of hospitalization decreased from one to zero admission (p= 0.001) following ivacaftor. The positive sputum cultures with pseudomonas decreased from 78.6% to 70%. Although this drug appeared safe in most of patient, 2 of the patients developed liver enzyme derangement without need to stop drug

CONCLUSION: this study showed all parameters were statically significant and proved effectiveness of ivacaftor for patient with (P.ser 549 Arg del) mutation at chromosome 7 in Oman regardless to severity or complication of the disease.

Keywords: Cystic fibrosis, Ivacaftor, Oman, adult

PP-15 [Difficulties in management of CF in the Middle East]

ASSOCIATION BETWEEN CFTR MUTATION CLASSES AND CYSTIC FIBROSIS SEVERITY MARKERS IN CHILDREN

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BACKGROUND: Cystic fibrosis is a multiorgan disease with great phenotypic heterogeneity and there are many studies that attempt to reveal the relationship between disease genotype and phenotype. The aim of this study was to investigate the effect of class I or class II mutation in any allele irrespective of the other allele on cystic fibrosis disease severity markers.

METHODS: This was a cross-sectional prospective study. Thirty-eight cystic fibrosis patients were evaluated. The patients CFTR genotype was divided into 2 group as the ones with (group I) and without (group II) class I or class II mutation in any allele. Cystic fibrosis disease severity markers were analysed between groups.

RESULTS: There was a significant difference observed in pancreas insufficiency (83.3% vs. 35.7%; p=0.005), chronic pseudomonas aeruginosa infection (58.3% vs. 7.1%; p=0.002), cough severity score (1.75±1.11 vs. 0.92±1.54; p=0.029), number of severe exacerbations treated with intravenous antibiotics during the previous year (0.91±1.01 vs. 0.28±0.82; p=0.032), number of outpatient control in the last year (5.39±2.8 vs. 2±1.1; p<0.001) and sweat chlorid levels (76.7±15.2 vs. 61.04±22.35; p=0.02), all being higher in group I patients. The parental consanguinity (12.5% vs. 50%; p=0.02) and mean BMI (15.8+2.2 vs. 17.6+2.8; p=0.03) was lower in group I.

CONCLUSION: Having class I or II mutation in any allele is a useful marker to predict the clinical course and severity of the disease in cystic fibrosis patients.

Keywords: cystic fibrosis, CFTR mutation classes, cystic fibrosis phenotype, disease severity markers,

PP-16 [Difficulties in management of CF in the Middle East]

CHARACTERISTICS OF CYSTIC FIBROSIS PATIENTS DIAGNOSED AFTER FALSE NEGATIVE CYSTIC FIBROSIS NEWBORN SCREENING RESULTS

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OBJECTIVE: The aim of this study is to report the clinical and genotype characteristics of patients who were diagnosed as cystic fibrosis (CF) after false negative cystic fibrosis newborn screening (CFNBS) results since January 2015.

MATERIAL-METHODS: Patients with first IRT value greater than 90 ng/ml and second IRT value greater than 70 were accepted as CFNBS test positive and referred to a sweat test.The sweat test value above 60 mmol/L was considered positive.

FINDINGS: 63 patients were diagnosed as CF in our clinic between 2015-2018. While 84.2% (n = 53) of the patients were diagnosed by positive CFNBS result, 15.8% of the patients (n = 10) had false negative results of IRT values. Of the 10 patients included in the study, 50% were girls, with a mean age of 30.7 ± 8.8 months (12-39 months); The mean age of diagnosis was 152 ± 147 days (28-480 days). Genetic mutation was detected in all patients. The first IRT level was above 90 µg/l in 40% (n = 4) of the patients. 20% of the patients had respiratory symptoms; Respiratory symptoms and diarrhea were present in 10% and growth failure was observed in 30% of the patients. 30% of the patients were presented with meconium ileus and 10% of the patients were diagnosed due to history of CF in their brother or sister.

CONCLUSION: It should be kept in mind that IRT negativity may also occur in mutations that cause severe CF.

Keywords: Cystic fibrosis, screening, false negativity, genetic

PP-17 [Difficulties in management of CF in the Middle East]

CLINICAL CHARACTERISTICS OF CYSTIC FIBROSIS PATIENTS AT OUR CLINIC

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AIM: The aim of this study is to determine the clinical and demographic characteristics of Cystic Fibrosis (CF) patients at our clinic.

METHOD: Cystic fibrosis patients data were collected in the pediatric pulmonology clinic of Medipol University Medical Faculty.

RESULTS: Sixteen children were evaluated at study period (9 girls and 7 boys), the median age was 30.5 months and median age at diagnosis was 60 days. Sixty two percent of patients (n=10) were diagnosed with newborn screening test. The median number of outpatient visits to our center per year was 5.5 and the median number of respiratory culture per year was 4. Chronic colonization was evaluated in 5 patients, intermittant colonization in 9 patients. Fifty percent of patients had colonization with pseudomonas, 35.7% with MSSA, 14.3% with MRSA. The median number of pulmonary exacerbations per year was 1. Inhaled bronchodilator and pulmonary physiotherapy were advised to all CF patients. Dornase alpha (81.3 %), hypertonic saline (68.8 %), inhaled corticosteroids (12.5 %), inhaled colistin (62.5 %) and inhaled tobramycin (12.5 %) were prescribed. *Table1* Eighty one percent of patients had pancreatic exocrine insufficiency (fecal elastase < 200µg/g). Two patients were underweight (BMI Z score <-1,8). Eleven patients were need to take high calorie intake, 5 patients with nasogastric feeding tube, 1 patient with gastrostomy tube. Three patients had meconium ileus and 2 patients nasal polyp. Five patients over 4 years old were going on formal education.

CONCLUSION: Intensive periodic follow up of CF patients is important for preventing mortality and morbidity.

Keywords: Cystic fibrosis, Colonization, Pulmonary Medications

PP-18 [Difficulties in management of CF in the Middle East]

CLINICAL AND SOCIO-DEMOGHRAPHIC EVAULATION OF CYSTIC FIBROSIS

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Cysticfibrosisisanautosomalrecessiveinheriteddiseasecaused byCTFRgenemutation.Itinvolvesprimarilythelungandmanyother organs.Theaimofthisstudyistoevaluatethesociodemographicandclinicalcharacteristicsofpatientswithcysticfibrosisinourclinic. The study included 106 patients, 52 of whom were girls (49%). The mean age at diganosis was 1,1 ±0,98 years old.. Twenty two patients (20,7 %) were diagnosed by neonatal screening program. The most common CFTR gene mutations were delta F508 (53,2%), G542X (11,6%), E92K (10,3%), G85E (6,4 %). When the patients were evaluated microbiologically; 26 of them had pseudomonas aeruginosa and 29 patients had staphylococcus aureus. Non tuberculous mycobacteria was seen in one patient. Two patients were diagnosed as allergic bronchopulmonary aspergillosis. We also evaluated the treatment that patients were on. All of patıents were usıng dornaz alpha, 91 of patients were having pancreatic enzyme replacement (85,8 %), 34 were having hypertonic saline (32 %), 35 were having inhaled salbutamol (33,1 %), 9 were having inhaled steroid (8,4 %) and 20 were having inhaled antibiotic (18,8 %). There was no patient who used inhaled mannitol. Respiratory physiotherapy had regulary been performed by all of our patients. 54% of patients were receiving enteral nutrition support. Cystic fibrosis is a chronic disease that requires lıfe long and regular treatment. A multidiciplinary approach and treatment of the disease related complications is important to prolong the survey and increase life quality of patients.

Keywords: Cystic fibrosis, bronchiectasis, pseudomanas

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