Journal of Cystic Fibrosis

The Official Journal of the European Cystic Fibrosis Society

Supplement:

Abstracts of the
42nd European Cystic Fibrosis Conference
Liverpool, United Kingdom, 5–8 June 2019

Vol. 18 Suppl. 1
June 2019
ISSN 1569-1993

www.ECFS.eu

P323

Pseudo Bartter Syndrome: the most common complication in the Turkish National Cystic Fibrosis Registry

T. Sismanlar Eyuboglu[1], D. Doğru Ersoz[2], E. Cakır[3], N. Cobanoglu[4], S. Pekcan[5], G. Cinel[6], E. Yalcın[2], N. Kiper[2], V. Sen[7], H. Selimoglu Sen[8], O. Ercan[5], O. Keskin[9], S. Bilgic Eltan[9], L. Muhammed Al Shadfan[3], H. Yazan[3], D.U. Altıntas[10], S. Sasihuseyinoglu[10], N. Sapan[11], S. Cekic[11], H. Cokugras[12], A.A. Kılınc[12], T. Ramaslı Gursoy[13], A.T. Aslan[13], A. Bingol[14], A.E. Basaran[14], A. Ozdemir[15], M. Kose[16], M. Hangul[16], N. Emiralioglu[17], G. Tugcu[6], H. Yuksel[18], O. Yılmaz[18], F. Orhan[19], Z.G. Gayretli Aydın[20], E. Topal[21], Z. Tamay[22], A. Suleyman[22], D. Can[23], C.M. Bal[24], G. Caltepe[25], U. Ozcelik[2]. *[1]Dr Sami Ulus Maternity and Children Training and Research Hospital, Pediatric Pulmonology, Ankara, Turkey; [2]Hacettepe University Faculty of Medicine, Pediatric Pulmonology, Ankara, Turkey; [3]Bezmialem University Faculty of Medicine, Pediatric Pulmonology, Istanbul, Turkey; [4]Ankara University Faculty of Medicine, Pediatric Pulmonology, Ankara, Turkey; [5]Necmettin Erbakan University Meram Medicine Faculty, Pediatric Pulmonology, Konya, Turkey; [6]Ankara Children's Hematoloji Oncology Training and Research Hospital, Pediatric Pulmonology, Ankara, Turkey; [7]Dicle University Faculty of Medicine, Pediatric Pulmonology, Diyarbakır, Turkey; [8]Dicle University Faculty of Medicine, Pulmonology, Diyarbakır, Turkey; [9]Gaziantep University Faculty of Medicine, Pediatric Allergy, Gaziantep, Turkey; [10]Cukurova University Faculty of Medicine, Pediatric Allergy and Immunology, Adana, Turkey; [11]Uludag University Faculty of Medicine, Pediatric Allergy and Immunology, Bursa, Turkey; [12]Istanbul University Cerrahpasa Medicine Faculty, Pediatric Allergy and Pulmonology, Istanbul, Turkey; [13]Gazi University Faculty of Medicine, Pediatric Pulmonology, Ankara, Turkey; [14]Akdeniz University Faculty of Medicine, Pediatric Pulmonology, Allergy and Immunology, Antalya, Turkey; [15]Mersin Maternity and Children Hospital, Pediatric Pulmonology, Mersin, Turkey; [16]Erciyes University Faculty of Medicine, Pediatric Pulmonology, Kayseri, Turkey; [17]Gaziantep Cengiz Gökçek Maternity and Children Hospital, Pediatric Pulmonology, Gaziantep, Turkey; [18]Celal Bayar University Faculty of Medicine, Pediatric Pulmonology, Allergy and Immunology, Manisa, Turkey; [19]Karadeniz Technical University Faculty of Medicine, Pediatric Allergy, Trabzon, Turkey; [20]Karadeniz Technical University Faculty of Medicine, Pediatric Infectious Disease, Trabzon, Turkey; [21]Inonu University Faculty of Medicine, Pediatric Allergy, Malatya, Turkey; [22]Istanbul University İstanbul Medicine Faculty, Pediatric Allergy, Istanbul, Turkey; [23]Balikesir University Faculty of Medicine, Pediatric Pulmonology, Balıkesir, Turkey; [24]Ataturk Regional Training and Research Hospital, Pediatric Pulmonology, Erzurum, Turkey; [25]Ondokuz Mayıs University Faculty of Medicine, Pediatric Gastroenterology, Samsun, Turkey*

Objective: Pseudo bartter syndrome (PBS) is a known complication of cystic fibrosis (CF) which is usually seen in infancy and warm weather conditions. First data of Turkish National CF Registry was documented and PBS was found as the most common complication in the patients. We aimed to investigate the clinical features of CF patients with PBS.
Method: Data of CF patients with PBS evaluated in terms of demographics, mutation analysis, pulmonary function tests, colonisation and other complications.
Results: Totally 1170 CF patients were included into registry in 2017 and 120 of them (10%) had PBS. Seventy one (59%) of them were male and 49 of them were girl. Mean age of diagnosis was 0.73 ± 1,67 years (min:0.08; max:11). Five of them had meconium ileus history and 110 of them had pancreatic insufficiency. Chronic liver disease accompanied in 5 patients and diabetes in 1 patient.
Thirty three of the patients with PBS had *Staphylococcus aureus*, 23 *Pseudomonas aeruginosa*, 4 *Stenotrophomonas maltophilia* colonisation. Forty five different mutations were detected in 65 patients in 120 alleles. The most common mutation was DF508 in 21 alleles and it was homozygous in 5 (4%) patients. N1303K, D110H, G542X and E92K were the other common mutations, respectively. The most common mutations were class 1 and 2 mutations.
Discussion: Young age of our patients and warm climate of our country may cause the pseudo-bartter as the most common complication and severe mutations may predispose it.

P325

Multivitamins – the way forward: optimising vitamin supplementation in adults with cystic fibrosis

J. Al-Siaidi[1], A. Keele[1], D. Slim[1]. *[1]Bristol Adults Cystic Fibrosis Centre, Bristol, United Kingdom*

Objectives: To compare supplementation using a new combined vitamin preparation with individual vitamin supplements in terms of patient acceptability, cost and meeting current European nutrition guidelines for vitamin K (ECFS, 2016).
Methods: Dietitians at Bristol Adult Cystic Fibrosis Centre have worked with ParaPharm Development Ltd to develop a multivitamin preparation that met European guidelines for vitamin supplementation in CF. Paravit-CF was developed: a taste free, odourless formulation containing high levels of vitamins A, D, E and K.
Paravit-CF was approved by the Bristol, North Somerset & South Gloucestershire Joint Formulary in September 2018. CF patients requiring vitamins A, D, E and K are being transferred on to Paravit-CF 1 or 2 capsules daily.
To date, 18 patients have completed a short questionnaire on the acceptability of Paravit-CF.
Cost was compared with individual vitamin supplements using basic NHS net prices from the British National Formulary.
Results: Treatment burden is typically reduced from 5 to 2 capsules per day. Survey results show good acceptability: 89% of patients were satisfied with the switch to Paravit-CF, 78% of patients thought it would make it easier to take their vitamin supplements.
Cost of vitamin supplementation is reduced by approximately 50% when using Paravit-CF compared to a typical multivitamin regimen.
To date, 92 of 234 patients have changed to Paravit-CF. 57% of patients now meet the ECFS guidelines for vitamin K supplementation compared with 27% prior to starting Paravit-CF.
Conclusion: Paravit-CF is more acceptable to patients than individual vitamin supplements due to the reduced treatment burden.
Increased adherence to vitamin supplementation would be expected in view of better acceptability. It is tasteless, odourless, easy to swallow and saves money.
More patients are meeting ECFS nutrition guidelines.
Further work is needed to look at vitamin status on Paravit-CF.

P326

Are combined vitamin preparations useful in a paediatric cystic fibrosis population?

K. Stead[1], C. Patchell[1], L. Paskin[2], P. Nagakumar[3], M. Desai[3]. *[1]Birmingham Children's Hospital, Dietetic Department, Birmingham, United Kingdom; [2]Birmingham Children's Hospital, Pharmacy Department, Birmingham, United Kingdom; [3]Birmingham Children's Hospital, Respiratory Department, Birmingham, United Kingdom*

Objectives: A number of combined vitamin preparations (CVP) designed for use in CF are available, including vitamins A,D,E & K in one preparation. Our current practice uses separate vitamin A,D,E & K preparations to allow individualised treatment based on annual serum vitamin monitoring, resulting in bespoke vitamin combinations. We aimed to assess the suitability of CVP, considering prescribed vitamin doses against our local CF clinical guidelines.
Methods: Data was collected on vitamins prescribed in CF clinic Jan-Feb 2018. Doses were compared to the local CF clinical guidelines.
Results: 96 patients (54 male) age8.3yrs (6 m–16.5 yrs) were identified. They were prescribed a mean of 1.7 (range 1–3) vitamin preparations a day with multiple administration (range 1–7) required for 48/96 (50%)patients to get desired serum levels. 2 patients were prescribed IM vitamin D & 2 oral vitamin K. 20 patients (20.8%) were prescribed vitamin doses in accordance with local guidelines.
Conclusions: 79.2% of patients reviewed had individualised prescriptions based on vitamin levels. Further research is required to understand potential benefit of individualised regimes instead of a "blanket" guideline. Patients with individualised regimes may have low serum levels resulting from poor compliance & CVP may manage this, reducing the need to individualise doses. All CVP contain Vitamin K as recommended in national