318 *Candida blankii*: New agent in cystic fibrosis airways?

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A 14-year-old teenage boy with increase in pulmonary exacerbations and worsening in pulmonary function and nutritional status is presented. He was diagnosed with meconium ileus when he was born (F508del/2183AA→G). He had intermittent *P. aeruginosa* sputum isolation and chronic MRSA infection since 2010. In 2012 chronic *S. maltophilia* infection began while MRSA disappeared.
At the end of 2012 yeast isolation appeared although it could not be identified by Vitek2 system. As the patient began to show clinical and nutritional deterioration, a BAL was performed in order to find microbiological agents. During that procedure, a yeast was isolated again and it was finally identified by using molecular methods as *Candida blankii*. Another strain isolated later was also identified as *C. blankii*.
MICs of amphotericin B, fluconazole, voriconazole, itraconazole, posaconazole, anidulafungin and caspofungin were determined using EUCAST E.Def 7.2 method. Although no breakpoints are established for this species, both isolates presented low MIC values against all the drugs tested (⩽0.13 mg/L).
Antifungal treatment with itraconazole 200 mg/d began in August 2014. Serum concentration was monitored after 2nd week treatment, and according to the result the dose was decreased to 100 up to now. As of December 2014 the patient is improving. He gained weight (5 kg) and is feeling better, and although his FEV_1 is stable, he has no pulmonary exacerbations.
We report the first case of *Candida blankii* infection in humans. This finding reinforces the need to identify the fungal isolates to species level in order to know the current epidemiology of yeast isolates and their antifungal susceptibility profile.

319 Nebulised temocillin for prophylaxis against *Burkholderia vietnamiensis* in an adult with cystic fibrosis

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Objectives: To illustrate the use of nebulised temocillin for prophylaxis in CF and chronic *Burkholderia* infection in a 29-year-old male.
Methods: Retrospective data collection from medical notes, included spirometry, CRP, frequency of IV antibiotics (14-day courses), and symptomatology. Data was collected for pre and post temocillin periods.
Results: Baseline spirometry: FEV1 66% pred., FVC 89% pred. Between ages 23 and 27 years, he had 2 courses of IV antibiotics yearly for exacerbations. FEV1 fell to a minimum of 55% pred., and CRP (<10 baseline) reached a maximum of 50. Between ages 27 and 29, he suffered increased frequency/severity of exacerbations requiring 13 courses of IV antibiotics, with a raised CRP (>100 on 3 occasions), and worsened spirometry (minimum FEV1 37% pred. and FVC 40% pred.). Worsening symptoms were recorded. Following onset of nebulised temocillin alternate months (7 months), frequency of IV antibiotics decreased to 4 courses (3 courses whilst off temocillin). Maximum CRP 42, and minimum FEV1 50% pred., significant symptom improvement was reported. Continuous nebulised temocillin was then started. No IV antibiotics were given in the following 4-month period.
Conclusion: Temocillin is a narrow spectrum penicillin stable to most β-lactamases and ESBLs. It demonstrates in vitro activity against *B. cepacia*. It does not induce β-lactamase production. Clinical data suggests benefit of IV temocillin for CF patients with *B. cepacia*. This case illustrates that temocillin may be a well tolerated and effective nebulised antibiotic for CF patients with *Burkholderia*.

320 The use of ivacaftor in CF mutations with residual functioning protein

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Objectives: Ivacaftor was shown to decrease FEV1 by 5% in patients with classIV R117H mutation. We report the use of ivacaftor in four CF adults with various Class IV mutations.
Methods: Ivacaftor was prescribed to 4 patients with deteriorating clinical status. We obtained pre and post administration FEV1, weight, sweat chloride (in 3/4), and CFQ-R.
Results: Patient 1, a 26yo woman, CF genotype of 2 class IV mutations (R347P & L1065P). She had four pulmonary exacerbations, FEV1 dropped from 85% to 75%. Six months following ivacaftor, FEV1 improved to 81%. She gained 10 lbs. Sweat chloride decreased from 96 to 86 mmol/L and CFQ-R increased from 55 to 100. Discontinuation of ivacaftor showed marked worsening in all parameters.
Patient 2 is a 61yo male with W1282X and the class IV mutation D1152H. Six months into ivacaftor, FEV1 improved from 67% to 87% and he gained 17 lbs. Sweat chloride decreased from 30 to 22 mmol/L and CFQ-R improved from 55 to 100 points.
Patient 3 is a 35yo male with the class IV D579G mutation and S912X. After ivacaftor, FEV1 improved from 27% to 31%. He gained 7 lbs. Sweat chloride decreased from 91 to 74 mmol/L and CFQ-R doubled.
Patient 4 is a 72yo male with CF genotype G542X and the class IV mutation D1152H. Following ivacaftor, FEV1 improved from 39% to 51%. He gained 7 lbs and CFQ-R doubled.
Conclusion: We noted improvement in all patients and in all parameters at 6 months compared to baseline. One patient was retested at one month after stopping ivacaftor with worsening parameters. These data support the beneficial effects of CFTR potentiators in CF mutations producing residual functioning protein.

321 Changes in lung function and airway microbiology following ivacaftor therapy in an adult G551D homozygote

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Objectives: Ivacaftor has increased lung function and decreased pulmonary exacerbations and intravenous antibiotic use in patients with CF carrying the G551D mutation. This case study describes the effects of ivacaftor on airway microbiology and lung function in a G551D homozygote with chronic *Pseudomonas aeruginosa* (PA) infection.
Methods: All sputum culture reports prior to ivacaftor therapy were reviewed. Following ivacaftor initiation sputum samples were sent for standard culture at each clinic visit. 16S gene rRNA pyrosequencing was performed on paired sputum samples collected immediately prior to and after 6 months of ivacaftor therapy. FEV_1 was measured at baseline and at each follow up visit. Sweat chloride was assessed pre-treatment and at 6 months.
Results: Mucoid PA infection was acquired in 1983. The pre-ivacaftor baseline sputum sample and all preceding 26 samples taken over 8 years were culture positive for PA. 7 consecutive samples collected over 16 months following ivacaftor were culture negative for PA. A later sample was culture positive for the original infecting strain of PA (using variable number tandem repeat profiling). 16S gene rRNA pyrosequencing showed a drop in relative abundance of PA from 98.2% to 4.8% and an increase in microbial diversity over the 6 month sample period. FEV_1 improved from 68% predicted (baseline) to 90% predicted (20 months post ivacaftor). Sweat chloride reduced from 105 mmol/L (baseline) to 52 mmol/L (6months).
Conclusion: Bacterial diversity and relative abundance of typical CF pathogens may be significantly altered by modulation of both CFTR alleles. Sputum culture clearance of mucoid PA on should be interpreted with caution.