461* Clinical features of a cystic fibrosis patient chronically colonized with *Pandoraea pnomenusa* identified using MALDI-TOF mass spectrometry

A. Fernandez-Olmos[1], M.I. Morosini[1], M. García-Castillo[1], L. García-García[1], A. Lamas[2], R. Cantón[1], L. Máiz[2]. [1]*Hosp. Univ. Ramon y Cajal, Servicio de Microbiología, CIBER Epidemiología y Salud Publica, Madrid, Spain;* [2]*Hosp. Univ. Ramon y Cajal, Unidad Fibrosis Quística, Madrid, Spain*

Data from the clinical course and outcome of CF patients colonized/infected with *Pandoraea* spp. are scarce, partially due to the difficulties in its identification. MALDI-TOF MS (MT) has been recently introduced for Clinical Microbiology use improving bacterial identification.
Objetive: To describe the clinical features of a patient, chronically colonized along 3 years with *Pandoraea pnomenusa*, after re-examination of bacterial identification results.
Methods: Eight isolates recovered from sputum cultures (2006 to 2009) of a 28-year-old female patient were re-identified using MT (MALDI BioTyper 2.0, Bruker Daltonik, Germany). PCR of 16S rDNA and sequencing were also performed. Genetic relatedness of isolates was assessed (*Xba*I-PFGE*)*.
Results: The patient firstly acquired *P. pnomenusa* in Oct-2006. Her pulmonary function remained stable, FVC 2.87 L (85%) and FEV_1 2.22 L (75%), until Jan-2008 when her clinical condition worsens, suffering from 7 pulmonary exacerbations that required antibiotic treatment. Her lung function declined to FVC 2.36 L (72%) and FEV1 1.85 L (65%) (Oct-2009). Laboratory records indicated that 8 isolates were identified, by conventional methods, as various "non-fermentative Gram-negative bacilli". MT identified all these isolates as *P. pnomenusa.* PCR of 16S rDNA and sequencing could establish the genus. PFGE results revealed indistinguishable patterns.
Conclusions: The presence of *Pandoraea* spp. can be underestimated in CF patients when using conventional identification methods. MT provides an accurate identification of this organism which may have a potential role in pulmonary function deterioration when chronically colonizes CF patients.

462 Temocillin: time to rediscover it?

A. Amaddeo[1], F. Poli[2], E. Barbi[2], I. L'Erario[2]. [1]*IRCCS Burlo Garofolo, Cystic Fibrosis Centre, Trieste, Italy;* [2]*IRCCS Burlo Garofolo, Trieste, Clinica Pediatrica, Trieste, Italy*

I. is a 22-year-old female affected by cystic fibrosis, diagnosed at birth for the presence of meconium ileus. She also suffers from CF related diabetes and mild CF hepatopathy. Since 2003 *B. cenocepacia* was constantly isolated from her sputum. In January 2009 she had a respiratory exacerbations with increased sputum production, increased dyspnoea and decreased FEV1 (from 48% to 36% of predicted at admission) requiring a course of intravenous antibiotics. Laboratory tests at admission shown increased erythrocyte sedimentation rate (ESR 66 mm/h) and C-reactive protein (CPR 5.56 mg/dl). Last available sputum culture of *B. cenocepacia* was resistant to all commonly used antibiotics. While awaiting multiple combination bactericidal antibiotic test results, IV meropenem, tobramicin and ceftazidime were administered. After four days of therapy her clinical condition began to deteriorate, with increased CRP (8.73 mg/dl), decreased spirometric results (FEV1 18% of predicted), intermittent fever and increased need for oxygen supplementation (from 1 to 4 l/min). Meanwhile multiple combination tests did not prove bactericidal activity. We decided to introduce Temocillin, an intravenous antibiotic not used in Italy, but proven to be efficacy in the treatment of *B. cenocepacia* multiresistant exacerbation. From the 16th days of hospitalisation temocillin was administered. After two days of treatment the fever disappears and CRP started to diminish (from 9.8 mg/dl to 0.5 mg/dl at days 21). After two weeks of intravenous temocillin therapy. To this days she experienced 4 more exacerbations, all successfully treated with IV temocillin; she is currently awaiting for lung transplantation.

463 Non invasive ventilation (NIV) and pregnancy

S. Bresci[1], S. Gambazza[1], D. Innocenti[1], F. Mecacci[2], P. D'Onofrio[3], M.P. Rambaldi[2], C. Braggion[1]. [1]*AOU Meyer, Cystic Fibrosis, Florence, Italy;* [2]*AOU Careggi, Gynecology, Florence, Italy;* [3]*AOU Careggi, Anaesthesia, Florence, Italy*

FEV1 < 40% of predicted value is a relative contraindication for pregnancy. Here we introduce a 29 ys old woman who decided to carry out her pregnancy despite a FEV1 of 25% pred. at 2 weeks' gestation.
She had a severe bronchopneumopaty with a FEV1 < 30% in past 4 ys with chronic infection by *P. Aeruginosa*, MRSA. During last exacerbations, she needed oxygen therapy (OT) at rest and during efforts. She has pancreatic insufficiency but a good BMI (20) and a CFRDM insulin-dependent. Once pregnant, our team regularly assessed clinical status, cardio-respiratory function, glucose blood levels and microbiological findings. She required 3 hospitalizations at 17th, 26th and 28th week's gestation due to acute exacerbations; she was treated with i.v. ceftazidime, inhaled colistine, steroids and OT, first nocturnal then daytime. To prevent respiratory deterioration and dyspnoea we started to acclimatize her to NIV since the first admission, with no recorded hypercapnia. Though her reluctance, it was then well tolerated and she used it increasingly, especially during sleep. Spirometry, SpO_2 and gas exchange remained stable throughout pregnancy. A joint decision was made to deliver the baby at 31st week gestation by caesarian section, using spinal-anesthesia and being assisted by physiotherapists for NIV in operating theatre. The outcome was successful for her and the newborn.
A close surveillance, a sub-continuous antibiotic therapy, a tailored respiratory physiotherapy and a multidisciplinary work let her be in fairly good health to give birth. According to our findings and patient's judgment, the key point of this success has been the early but gradual use of NIV.

464 Renal hyperechogenicity: a newly recognized complication of severe dehydration in infants with cystic fibrosis

A. Katelari[1], P. Galina[2], A. Kapi[1], J. Lympari[1], M. Zarifi[2], S. Doudounakis[1]. [1]*Cystic Fibrosis Department, 'Aghia Sophia' Children's Hospital, Athens, Greece;* [2]*Radiology Department, 'Aghia Sophia' Children's Hospital, Athens, Greece*

Introduction and Aim: Dehydration is a common complication of cystic fibrosis. Its frequency mainly depends on external factors as a rise in environmental temperature, fever, excess loss and/or inadequate intake of salt and fluids. As dehydration progresses, hypovolemic shock ultimately ensues, with a detrimental effect on the vital organs. At the present study, we aimed to explore the influence of dehydration on kidneys of infants with cystic fibrosis. Specifically, we studied the ultrasonographic changes of kidneys after an episode of severe dehydration.
Patients and Methods: 12 infants with cystic fibrosis were admitted to the hospital with dehydration over the last year. There were 6 boys and 6 girls (mean age 17 months). All the infants were admitted during summer months (June to August). For 4/12 dehydration was the presenting symptom of cystic fibrosis. All infants were investigated with renal ultrasound at the time of dehydration. The severity of dehydration was correlated with ultrasound findings.
Results: 4/12 infants with cystic fibrosis and dehydration had abnormal renal ultrasound at the time of dehydration. Specifically, the ultrasound examination demonstrated echogenic kidneys with preservation of the corticomedullary differentiation. The kidneys were equally involved. All 4 children had markedly abnormal electrolytes with chloride below 70 mmol/l and urea above 100 mg/dl.
Conclusion: Severe dehydration causes renal changes as detected by ultrasound scan. It is important to take this event into consideration and avoid concomitant use of nephrotoxic drugs, which may have an add-on effect on vulnerable kidneys and cause irreversible damage.