177 Management and outcomes following pneumothorax in cystic fibrosis patients

J. Gallagher[1], D. Nazareth[1], S.M.H. Kazmi[1], K. Mohan[1], J. Greenwood[1], M. Ledson[1], M. Walshaw[1]. [1]*Liverpool Heart & Chest Hospital NHS Foundation Trust, Adult CF Unit, Liverpool, United Kingdom*

Objectives: Spontaneous pneumothorax (PTx) is a well recognised complication in CF and is associated with recurrence and increased morbidity and mortality. To inform future management, we looked at the treatment and outcome of PTx in CF patients attending our large adult centre.
Method: We reviewed 64 episodes of spontaneous PTx in 28 CF patients (mean FEV_1 32% predicted [range 16–68%], 36% male), focussing on modalities of treatment and ultimate outcomes.
Results: Immediate treatments are shown in the table. Nine patients (32%) sustained bilateral PTx and 50% had ipsilateral recurrence (mean interval 4 months). Twenty four have died (63% at 1 year, 80% at 2 years, median survival 7 months [0–68]) after first PTx, with the remaining 4 alive at 6–34 months post PTx.
Conclusions: We have confirmed that spontaneous PTx is a poor prognostic factor in CF with poor survival rates and a recurrence or a contralateral PTx increases mortality. A multidisciplinary approach towards the treatment modalities for the management of recurrent or persistent PTx is essential when dealing with this complication.

Pneumothorax treatment and outcomes

	Episodes (n)	Resolution (%)	2nd line treatment	Complications	Late recurrence (%)
Observation	13	69	Chest drain 23% Pleurodesis 8%	Nil	31
Chest Drain	23	52	Surgery 39%	Death 17%	43
Pleurodesis (talc/blood)	18	72	Surgery 11%	Death 11%	39
Surgery	10	50	NA	Death 50%	20

178 Prognostic impact of recurrent pneumothoraces in adults with cystic fibrosis

J. Chapron[1,2], C. Martin[1,2], E. Audureau[2,3], R. Kanaan[1,2], I. Honoré[1,2], D. Dusser[1,2], D. Hubert[1,2], P.-R. Burgel[1,2]. [1]*Hôpital Cochin, Pulmonary Department, Paris, France;* [2]*Université Paris Descartes, Sorbonne Paris Cité, Paris, France;* [3]*Hôpital Hotel Dieu, Biostatistics, Paris, France*

Rationale: To identify risk factors and long-term outcomes associated with spontaneous pneumothorax in adults with cystic fibrosis.
Methods: Consecutive adult CF subjects who experienced at least one spontaneous pneumothorax at Cochin Hospital (Paris, France) were individually matched (1:3) for age, gender and FEV_1 with CF subjects who did not have pneumothorax. Multivariate conditional logistic regression was used to identify independent factors associated with pneumothorax. Transplant-free survival was analysed using Kaplan-Meier analysis. Cox proportional hazards model were used to examine impact of pneumothorax on survival.
Results: From 1995 to 2010, 51 subjects had pneumothorax: 29 subjects had one pneumothorax and 22 had $\geqslant$2 pneumothoraces. These subjects were matched with 125 CF subjects without pneumothorax. No risk factor for pneumothorax was identified. Adult CF patient who experienced pneumothorax had shorter transplant-free survival ($p = 0.03$; log rank test). However, this difference was due to shorter transplant-free survival in subjects who had $\geqslant$2 pneumothoraces ($p = 0.004$), whereas subjects who had only one pneumothorax had comparable outcome compared to subjects without pneumothorax ($p = 0.52$). In Cox analysis, pneumothorax was an independent predictor associated with shorter survival: 1 pneumothorax (HR 2.10; 95% CI 0.94–4.69; $p = 0.07$) and $\geqslant$2 pneumothoraces (HR 5.35; 95% CI 2.31–12.37; $p < 0.0001$).
Conclusion: No risk factor for pneumothorax was identified. Recurrent pneumothoraces are associated with a major decrease of transplant-free survival, whereas single occurrence of pneumothorax has little effect on survival.

179 Lung transplantation in cystic fibrosis: waiting list results

L. Fila[1], L. Valentova Bartakova[1], V. Vavrova[2], R. Lischke[3]. [1]*University Hospital Motol, Pulmonary Department, Prague, Czech Republic;* [2]*Charles University, 2nd Faculty of Medicine, Department of Pediatrics, Prague, Czech Republic;* [3]*University Hospital Motol, 3rd Department of Surgery, Prague, Czech Republic*

Objectives: Lung transplantation (LuTx) is a treatment possibility in advanced pulmonary diseases including cystic fibrosis (CF). Careful evaluation of patients before inclusion on waiting list (WL) is based on international guidelines (Orens JB 2006). Care for patients on WL includes regular evaluation of compensation of underlying disease and comorbidities together with assessment of urgency, ability to tolerate and profit from LuTx. In this paper, we present single-centre experience with results in patients included on WL for LuTx.
Methods: During 1998–2011 period, 44 CF patients (22 M, 22 F) were included on WL for LuTx in Prague Transplant Center, Czech Republic (one woman twice, because of the need for re-LuTx). Colonization of airways included *Pseudomonas aeruginosa* in 23, *Stenotrophomonas maltophilia* in 3 and *Burkholderia cepacia* complex/*B.* spec. in 12 cases; remaining patients were without these Gram-negatives. Median (range) age at time on inclusion on WL was 24.6 (12.7–47.5) years. Median (range) time on WL was 0.58 (0.03–2.37) years. 29 patients underwent LuTx with median (range) time on WL 0.31 (0.03–2.37) years, 10 patients died with median (range) time on WL 0.90 (0.10–2.16) years and 3 patiens were removed from WL (colonization with *B. cenocepacia* ST-32 and improvement of lung function in 2 and 1 patient, respectively). Remaining patients (3) are still on WL. Time on WL in deceased patients was longer then in those who were transplanted, but the difference did not reach statistical significance ($p = 0.19$, Cox-Mantel test). Mortality on WL was 22.2%.
Conclusions: LuTx is a viable treatment option in CF patients and increasing donor pool may reduce WL mortality and shorten WL time.

180 Choice of IV vs. non-IV antibiotics for treating pulmonary exacerbations in patients with cystic fibrosis

J.S. Wagener[1], D.R. VanDevanter[2], W. Regelmann[3], L. Rasouliyan[4], W.J. Morgan[5], D.J. Pasta[4], M.W. Konstan[2], Investigators and Coordinators of the Epidemiologic Study of Cystic Fibrosis. [1]*University of Colorado, Pediatrics, Aurora, United States;* [2]*Case Western Reserve University School of Medicine, Pediatrics, Cleveland, United States;* [3]*University of Minnisota, Pediatrics, Minneapolis, United States;* [4]*ICON Late Phase & Outcomes Research, San Francisco, United States;* [5]*University of Arizona, Pediatrics, Tucson, United States*

Objectives: Cystic Fibrosis (CF) patients experience frequent pulmonary exacerbations (PExs). Clinicians manage these episodes of worsening signs and symptoms in a variety of ways, but commonly with antibiotic treatment. A better understanding of why clinicians choose IV vs. non-IV routes for antibiotic delivery may aid in better defining an exacerbation and improving therapy.
Methods: We used data from the Epidemiologic Study of CF to examine the patient characteristics and choice of IV vs. non-IV antibiotics to treat PExs between 2003 and 2005. 45,374 PExs were reported in 13,194 unique patients. 39% of PExs were treated with IV, 15% were treated with inhaled $\pm$ oral, and the remainder received only oral antibiotics. The likelihood of non-IV vs. IV antibiotic treatment was influenced by the patient's age, stage of lung disease, and magnitude of lung function drop prior to the PEx.
Conclusion: Clinicians treat the majority of PExs with oral antibiotics, particularly in younger, healthier patients.