WS8.1 Lung transplant referrals and outcomes: experience of a regional adult CF centre 2008–2013

C. Etherington[1], A. Boland[1], I. Clifton[1], P. Whitaker[1], D. Peckham[1]. [1]*St. James's University Hospital, Regional Adult Cystic Fibrosis Centre, Leeds, United Kingdom*

Objectives: Lung transplantation is the most effective means of improving survival and quality of life in patients with end-stage CF. Our aim was to determine outcomes of referrals, waiting times and factors associated with a poor outcome.
Methods: Data were retrieved retrospectively from our electronic patient record system for all adult patient referred to UK Transplant centres over a 6 year period.
Results: 48 patients [27 female, median (range) age at referral 29.7 yrs (16.5–49.5), FEV_1 25% predicted (13–41) and BMI 20.2 kg/m^2 (16.6–29.7) were referred for lung transplantation. Median time from initial discussion to referral was 80 days (13–1090). 3 patients died prior to first appointment and 5 are waiting to be seen. 40 patients were assessed and 26 accepted (4 died after first appointment, 2 not accepted and 8 currently under follow up). Of the 26 accepted, 16 have received a transplant [waiting time on active list 177 days (28–865)], 3 died on the active list, 3 were removed (significant clinical deterioration) and 4 are currently waiting. Total referral process from initial assessment to transplant was 1.6 yrs (0.7–3.2). Fifteen patients died during the referral process; there were no significant differences in clinical severity when compared to patients who were transplanted. However patients who died during the process had a significantly longer time from initial discussion to local assessment [112 (13–1090) vs. 56 (29–440) days, $p = 0.0481$].
Conclusion: Early discussion is important and current waiting times need to be taken into consideration. Increased psychological input is essential as an initial delay in the process may be associated with poor outcome.

WS8.2 Pulmonary arterial hypertension in adult CF lung transplantation candidates

L. Fila[1], A. Grandcourtova[1], L. Valentova Bartakova[1]. [1]*Faculty Hospital Motol, Pulmonary Department, Prague, Czech Republic*

Objectives: To evaluate factors associated with pulmonary arterial hypertension (PAH) in cystic fibrosis (CF) lung transplant (LuTx) candidates.
Methods: Demographic and functional parameters in adult CF LuTx candidates included on waiting list during 2009–2013 period were analyzed.
Results: 19 subjects (10 males and 9 females) with mean (±SD) age 28.9±8.4 years were included into study. Mean (±SD) values of forced expiratory volume in 1 sec. (FEV_1), partial pressure of oxygen and carbon dioxide in arterial blood (p_aO_2 and p_aCO_2), six-minute walking distance (6-MWD) and mean pulmonary artery pressure (MAP) were 23±5% pred., 7.30±0.86 kPa, 5.04±0.76 kPa, 297±86 m and 26±8 mmHg, respectively. Correlations of FEV_1 ($r = -0.495$; $p = 0.031$) and p_aCO_2 ($r = 0.463$; $p = 0.041$) with MAP were found, whereas remaining parameters did not correlate with MAP. In regression analysis, only FEV_1 tended do predict ($p = 0.057$) MAP value. Taken FEV_1 parameter as a cut-off, value <20% pred. had sensitivity, specificity and accuracy of 66.7%, 100.0% and 89.5%, respectively ($p = 0.004$ in Fisher's exact test) for MAP value ⩾25 mmHg. ROC for this FEV_1 cut-off value was 0.699 ($p = 0.11$).
Conclusion: Presence of PAH in adult CF LuTx candidates is associated mainly with very severe airway obstruction (FEV_1 <20% pred.).

WS8.3 Extracorporeal life support as a bridge to lung transplantation: outcome in cystic fibrosis recipients

I. Inci[1], U. Bürgi[2], C. Benden[2], L.C. Huber[2], P. Kestenholz[1], W. Weder[1]. [1]*Division of Thoracic Surgery, University Hospital, Zurich, Switzerland;* [2]*Division of Pulmonary Medicine, University Hospital, Zurich, Switzerland*

Objectives: We investigated the early and mid-term outcome following lung transplantation (LuTX) in patients with end-stage cystic fibrosis (CF) lung disease bridged with extracorporeal life support (ECLS) to LuTX in comparison to CF patients not requiring ECLS to LuTX.
Methods: We reviewed all 186 lung transplants performed between 01/2007 and 10/2013 at our institution. 30 patients required ECLS as bridge to LuTX, 12 of which had CF.
Results: The subgroup of CF patients ($n = 12$) included 8 females, median age 29 years (range 14–56). Venovenous extracorporeal membrane oxygenation (ECMO) was used in 6, venoarterial in 1, interventional lung assist device in 1, and stepwise combination of them in 4 recipients. Primary LuTX was performed in 10 and re-transplantation in 2 cases. Lobar LuTX was performed in 4 recipients. Success rate for bridging was 92% (12/13), only one patient died on ECMO while on the waiting list. Median duration of ECLS was 23 days (range 1–81). Two patients were kept awake on ECLS. Six patients required ECMO in the postoperative period (median 2 days, range 1–9). 30-day, 1-year, and 2-year survival was 92%, 79%, and 79% respectively in CF patients on ECLS in comparison to 100%, 81%, and 78%, respectively in CF patients ($n = 39$) not requiring ECLS ($p = 0.35$, Long Rank).
Conclusion: In carefully selected CF patients ECLS as a bridge to LuTX can produce similar early and mid-term outcomes compared to CF recipients not requiring ECLS a bridge to LuTX.

WS8.4 Extracorporeal life support in cystic fibrosis; a single centre experience

D. Keating[1], F. Finlayson[1], E. Williams[1], D. Edgeworth[1], T. Kotsimbos[1], J.W. Wilson[1]. [1]*Alfred Hospital, Allergy, Immunology and Respiratory Medicine, Melbourne, Australia*

Objectives: To assess our outcomes and indicators of ECMO survival.
Methods: We retrospectively analysed data from six CF patients who had undergone ECMO at our hospital since 2010.
Results: See the table.

Table: ECMO patient details

Patient	FEV1 % Pd	SOFA	Precipitant	Days to ECMO	NIV (hrs)/ Vent (d)	ECMO (d)	Outcome
1	46	9	Infection	11	114/1	32	Died
2	27	8	Infection	3	48/0	8	LTx
3	49	12	Infection & Pregnancy	3	72/1	3	LTx
4	35	12	Infection & Pneumothorax	27	24/1	8	LTx
5	85	10	H1N1	4	192/1	1	Died
6	46	12	Haemoptysis & Pneumothorax	3	0/2	1	Died

Conclusion: APACHEII scores did not identify survivors but low SOFA scores indicate ECMO tolerance. Patients retrieved for ECMO from other hospitals were longer on NIV or intubated and tolerated ECMO poorly suggesting centres with more experience of unwell CF patients identify ECMO candidates better. Only 1 of 6 patients met lung transplant (LTx) assessment criteria meaning ECMO decisions must be considered early if we are to identify recoverable patients. All our patients were exclusively women suggesting their known predilection for rapid clinical decline means they are overrepresented in our ECMO recipients and should be considered for ECMO earlier in the course of their disease.