Burden of comorbidities in cystic fibrosis patients with long term survival following lung transplantation

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Background: Lung transplantation (LTX) is the ultimate treatment option for patients with end-stage cystic fibrosis (CF) lung disease; however, morbidity and mortality are considerable. Nevertheless, the subgroup of patients with CF achieves the best overall outcome post LTX compared to other primary indications. Survival following LTX is dependent on the development of chronic lung allograft dysfunction (CLAD) and complications like infections, cardiovascular events, chronic kidney disease (CKD) and cancer. We investigated a subgroup of patients with CF and a minimum of 10 years post-LTX survival, aiming to determine possible predictors of long-term survival and characterize comorbidities.

Methods: All CF LTX patients at the University Hospital Zurich from 11/1992 to 6/2017 with survival >10 years were included. *Evaluated parameters*: gender, BMI, diabetes, bacterial chronic pulmonary infection (*Burkholderia cepacia complex*) at the time of evaluation for LTX, need for ECLS as bridging to LTX and CMV risk status. *Clinical endpoints*: CLAD-free survival. Additional endpoints included re-hospitalization rate and extra-pulmonary comorbidities: chronic kidney disease, dialysis or kidney transplant, arterial hypertension, cardiovascular disease, diabetes and malignancies. Patients with survival from 1-5 years were compared to patients with survival > 10 years.

Results:

Characteristics at time of listing

CLAD-free survival

	Survival 1-5yrs (n=67)	Surival > 10yrs (n=39)	1.0-	~~	CLAD-no
female n(%)	31 (46)	16 (41)			
BMI median (range)	17.9 (13.0-27.0)	17.4 (13.4-22.8)	0.6-	١	CLAD-yes
BMI <18.5 kg/m2 n(%)	40 (59)	27 (69)	0.2-		L
BCC n(%)	2 (3)	2 (5)	0 5	10 15	20
Preop ECLS n(%)	12 (18)	0 (0)		years	
CMV high risk n(%)	11 (16.4)	20 (51)			

Long-term comorbidities following LTX

	Survival 1-5yrs	Survival > 10yrs
	(n=67)	(n=39)
Diabetes mellitus	25 (37)	20 (51)
Arterial hypertension	27 (40)	32 (82)
Chronic kidney disease	31 (46)	28 (72)
Kidney replacement therapy	1 (1.5)	5 (13)
Kidney transplantation	0 (0)	10 (26)
Cardiovascular disease	4 (6)	9 (23)
Skin cancer	0 (0)	5 (13)
Malignancy other	0(0)	0
PTLD	4 (6)	0
Annual Hospitalisation rate median	1.1 (0-9.2)	0.7 (0-3.2)
(range)		

ventilation or ECLS pre-LTX, low incidence of malignancy and low frequency of hospitalization. Prevalence of diabetes mellitus, arterial hypertension, cardiovascular disease and chronic kidney disease are unrelated to long-term survival.

