O-082

RARE INDICATIONS FOR LUNG TRANSPLANTATION: A EUROPEAN SOCIETY OF THORACIC SURGEONS (ESTS) SURVEY

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Objectives:
Due to the paucity of evidence supporting recommendations, lung transplantation (LT) guidelines are based mainly on consensus. Knowledge on rare ("orphan") diseases that can be treated by LT is even more difficult to collect and consolidate. The aim of this study was to collect and analyze clinical data of patients transplanted for rare diseases.

Methods:
Members of the ESTS Lung Transplantation Study Group were invited to collect data on transplant patients; the exclusion criteria comprised combined transplants, retransplantation and the most common underlying diseases (pulmonary fibrosis, emphysema, cystic fibrosis, pulmonary hypertension, and GVHD). Data were collected in a single anonymized database and described using mean and standard deviation or absolute frequencies and percentage, Kaplan-Meier estimates and death hazard ratio were used to depict survival. The data were analyzed using R package.

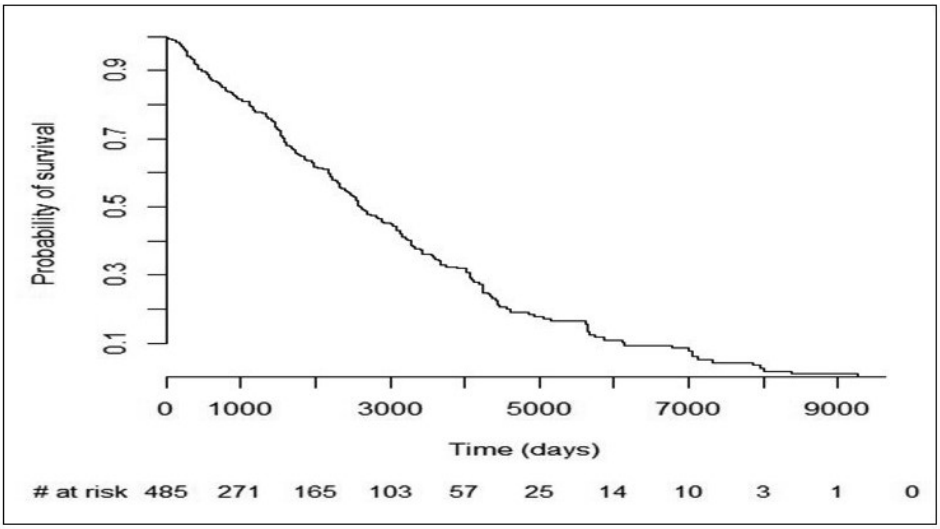
Results:
Four-hundred-and-eighty-six records were collected from 12 centers; 38 different underlying diagnoses were reported. The table reports essential clinical parameters. One and 3-year overall survival probabilities were 0.928 (95%CI 0.904-0.953) and 0.808 (95%CI 0.769-0.848) whereas ISHLT registry survival matched for age category were 0.861 (95%CI 0847–0.875) and 0.738

(95%CI 0.716–0.76) respectively. The most frequent underlying diseases were scleroderma (83), sarcoidosis (80), lymphangioleiomyomatosis (67) and hypersensitivity pneumonitis (50). One and 3-year survival rate of the mentioned cohorts were 0.945 and 0.776, 0.942 and 0.862, 0.948 and 0.843, 0.863 and 0.690 respectively. The figure reports the comprehensive actuarial survival curve. Overall risk hazard for death constantly increased over time while each cohort had its particular profile.

Conclusions:
Thirty-eight rare indications to LT were collected; the four above mentioned pathologies constitute 58% of all the rare diagnoses. Overall survival was similar to that documented by the ISHLT registry for adult patients. Risk hazard profiles for death were different among diagnosis cohorts. LT can be proposed as a successful therapy even in "orphan" diseases.

Total patients with follow-up, n	486
Age, year ,mean (sd)	48.0 (12.1)
Gender, female, n (%)	262 (53.9)
FEV1 % predicted, mean (sd) – (431 patients)*	41.2 (21.6)
FVC % predicted , mean (sd) – (403 patients)*	50.2 (22.1)
DLCO % predicted, mean (sd) – (278 patients)*	33.2 (17.6)
Bilateral transplantation, n (%)	399(82.1)
Induction therapy, n (%)	215 (44.2)
CLAD, n (%)	183 (37.7)
Retransplantation, n (%)	16 (3.3)

n: number; sd: standard deviation; *: patients with available data.



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