European Respiratory Society
Annual Congress 2012

Abstract Number: 4680

Publication Number: P683

Abstract Group: 1.5. Diffuse Parenchymal Lung Disease

Keyword 1: Orphan disease **Keyword 2:** Interstitial lung disease **Keyword 3:** No keyword

Title: Rate of FEV1 decline in lymphangioleiomyomatosis associated with tuberous sclerosis complex

Dr. Romain 30028 Lazor romain.lazor@chuv.ch MD [1,3], Prof. Martine 30029 Reynaud-Gaubert MartineLouise.REYNAUD@ap-hm.fr MD [2], Prof. Vincent 30030 Cottin vincent.cottin@chu-lyon.fr MD [3], Dr. Jacques 30031 Lacronique jacques.lacronique@cch.aphp.fr MD [4], Dr. Camille 30032 Taillé camille.taille@bch.aphp.fr MD [5], Prof. Benoit 30040 Wallaert benoit.wallaert@chru-lille.fr MD [6], Dr. Sandrine 30041 Hirschi sandrine.hirschi@chru-strasbourg.fr MD [7], Prof. Bruno 30061 Crestani bruno.crestani@bch.ap-hop-paris.fr MD [5], Prof. Jean-François 30054 Cordier jean-francois.cordier@chu-lyon.fr MD [3] and 30070 the Groupe d Etudes et de Recherche sur les Maladies Orphelines Pulmonaires jean-francois.cordier@chu-lyon.fr . [1] Respiratory Medicine, Lausanne University Hospital, Lausanne, Switzerland, 1011 ; [2] Respiratory Medicine, Marseille University Hospital, Marseille, France ; [3] Reference Center for Rare Pulmonary Diseases, Lyon University Hospital, Lyon, France ; [4] Respiratory Medicine, Cochin University Hospital, Paris, France ; [5] Respiratory Medicine, Bichat-Claude Bernard University Hospital, Paris, France ; [6] Respiratory Medicine, Lille University Hospital, Lille, France and [7] Respiratory Medicine, Strasbourg University Hospital, Strasbourg, France .

Body: Pulmonary lymphangioleiomyomatosis (LAM) is a rare disease characterized by proliferation of abnormal TSC1/2-mutated smooth muscle cells in the lung parenchyma, leading to progressive cystic destruction, lung function decline, and respiratory insufficiency. LAM may be either sporadic (S-LAM) or associated with the tuberous sclerosis complex (TSC-LAM). Cross-sectional studies have suggested that TSC-LAM is less severe than S-LAM, but no longitudinal data are available. We retrospectively compared the rate of FEV1 decline in 16 patients with TSC-LAM and 53 patients with S-LAM diagnosed according to ERS 2010 criteria with a lung function follow-up ≥1 year. Results are shown below.

	TSC-LAM	S-LAM	
	mean ± SD	mean ± SD	p value
n	16	53	
women, %	94	100	
age at LAM diagnosis, yr	33 ± 11	40 ± 10	0.004
initial FEV1, %pred	72 ± 25	70 ± 26	NS
lung function follow-up, yr	5.9 ± 5.2	5.3 ± 3.8	NS
FEV1 decline %pred/yr	-2.8 ± 2.8	-3.4 ± 5.0	NS

FEV1 decline ml/an	-103 ± 89	-114 ± 177	NS

Conclusion: Although diagnosed at an earlier age than S-LAM, patients with TSC-LAM had similar mean FEV1 at diagnosis, and similar mean rate of FEV1 decline. Lung function follow-up similar to S-LAM may be recommended in TSC-LAM.