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REGISTRY OF RARE LUNG DISEASES - OUR FIRST SINGLE CENTER RESULTS

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Objective: Objectives: The aim is to present first single center results of our first Registry of rare lung diseases. Purpose of our Registry is to monitor outcomes and study best practices in care or treatment and it can be used for improving and monitoring the quality of health care.

Methods: In our registry we collected data for patients that are controlled and treated in our Center for one of the rare diffuse lung diseases. Registry includes demographic data, symptoms and signs of the disease, comorbidities, presence of professional exposure, lung function test results, diagnostic tools and management/treatment of the patients.

Results: A total of 48 patients were included. 31 (65%) were male and 17 (35%) female with median age 64.5 years (30±89). 27 (56%) patients were diagnosed with idiopathic pulmonary fibrosis, 6 (12%) patients had NSIP, 5 (10%) had lymphangioleiomyomatosis, 4 (8%) had Langerhans histiocytosis, 3 (6%) had eosinophilic pneumonia, 2 (4%) had lipid pneumonia and 1 (2%) had microlithiasis. Regarding symptoms 34 (63%) patients reported dyspnoea, 25 (52%) reported dry cough, 2 (4.2%) patients reported no symptoms. Median time between onset of symptoms and diagnosis was 11 months (1±48). 12 (25%) patients had associated COPD, 6 (12%) had pulmonary hypertension, 2 (4%) had GERD, 2 (4%) had pneumothorax, 1 (2%) had tuberous sclerosis and 1 (2%) was obese. Other comorbidities that was reported included diabetes mellitus, arterial hypertension, cardiomyopathy,

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coronary disease and depression.

Conclusion: Knowing the importance of data contained in registries, we have presented first such data on national level regarding adult rare diffuse lung disease population in Croatia. These efforts would be used to improve outcomes in people with rare lung diseases and ultimately to recruit patients for clinical trials to learn about a particular disease or condition and will lead to developing research hypotheses.