2.20 Characteristics of Diffuse Cystic Lung Diseases in a National Referral Centre

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Background: Diffuse cystic lung diseases (DCLD) are defined by the presence of multiple, irregular, thin-walled cysts within the pulmonary parenchyma. The knowledge of DCLD has increased due to widespread availability of HRCT. Despite this, the diagnosis of DCLD can be a challenge. **Methods**: This was a single centre retrospective observational study of patients referred to the National Rare Lung Disease Clinic (RLDC) from its initiation in March 2019 to the end of December 2023. Data was collected from the inhouse electronic database (Clinical Portal) **Results:** 217 patients were referred to the RLDC during the study period. There was a mean of 39 ± 20 new referrals seen per year. 145 patients ultimately carried a diagnosis of DCLD. The mean age of this cohort was 53 ±15 years. 22% (n=32) were male and 14% (n=18) were smokers. The most common DCLDs seen in our clinic were Lymphangioleiomyomatosis (36%, n=53) Birt-Hogg-Dubé Syndrome (35%, n=51), Pulmonary Langerhans Cell Histiocytosis (7%, n=10) and Lymphocytic Interstitial Pneumonia (7%, n=10). 1 patient had a diagnosis of Congenital Pulmonary Airway Malformation. Pneumothorax is a common presentation in 32% (n=46). The mean FEV1/FVC (%) was 58±29 and DLCO predicted (%) was 56±36. **Conclusion**: Our study highlights the importance of access to specialist services to accurately characterise DCLD and its mimics, particularly those which can be screened, may be progressive or have malignant potential. **Conflicts of Interest:** The authors declare that they have no conflicts of interest.