6.01 Prevalence and Clinical Characteristics of LAM in an Irish Cohort

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Rationale: Lymphangioleiomyomatosis (LAM) is a metastasizing neoplastic disease that results in cystic lung disease. It is rare, with a reported prevalence of 3-8 cases per million women. Characteristic HRCT Thorax plus; serum VEGF-D ≥800pg/ml, history of tuberous sclerosis (TSC), angiomyolipomas (AML), chylothorax or lymphangioleiomyomas are diagnostic.

Methods: Data for 53 patients with a diagnosis of LAM attending our Rare Lung Diseases Clinic was reviewed. Patient demographics, TSC diagnosis, hormonal status, presentation, diagnostic criteria, PFTs and numbers on treatment and oxygen were collected.

Results: Mean age was 53.3 years (SD+/-12.3). Nine percent (n=5) have a diagnosis of TSC. Twenty eight percent (n=15) had angiomyolipoma, nine percent (n=5) a history of chylothorax and 24% (n =13) a history of pneumothorax. Forty-five percent of our cohort (n=19) were pre-menopausal. Average VEGF-D level was 567.34pg/ml (SD+/-544.18pg/ml). Twenty one percent (n=9) are currently on treatment with an mTOR inhibitor and four patients are on long-term oxygen. Average FEV1 is 89.76% (SD+/-20.85%) and DLCO is 69.92% (SD+/-26.44%).

Discussion: 53 patients with LAM have been identified to date in Ireland equating to 21 cases per million females; much higher than the reported worldwide prevalence. With effective therapy available it is important to correctly identify those with LAM.

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