lung function (available in 9 patients) demonstrated a mean improvement in% predicted FVC and FEV_1 of +33 (p = 0.009) and +37 (p = 0.006), respectively, following cessation of nitrofurantoin. 44% of patients were also prescribed oral prednisolone. Comparing these two groups (cessation + steroid vs cessation alone) showed no significant difference in mean% predicted FVC (p = 0.47) or FEV_1 (p = 0.87), gender, age or imaging at diagnosis. Following treatment, there was no significant difference in% predicted FVC (p = 0.87) or FEV_1 (p = 0.93) between groups. The mean% predicted FVC improvement was 31% in the steroid group and 34% in the cessation only group, showing no significant difference between groups (p = 0.86).

Conclusions With increased nitrofurantoin prescribing, the prevalence of NL will continue to rise throughout the UK and heightened awareness of the condition will be required in primary and secondary care. Our data demonstrates that significant improvements in lung function occur on cessation of nitrofurantoin and suggests no benefit is conferred by additional use of corticosteroid in patients with chronic NL.

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S8 RITUXIMAB THERAPY FOR REFRACTORY MYOSITIS RELATED INTERSTITIAL LUNG DISEASE UNRESPONSIVE TO CONVENTIONAL IMMUNOSUPPRESSION: THE BRISTOL INTERSTITIAL LUNG DISEASE SERVICE EXPERIENCE

C Sharp, N Dodds, A Edey, H Adamali, H Gunawardena, A Millar. *North Bristol NHS Trust, Bristol, UK*

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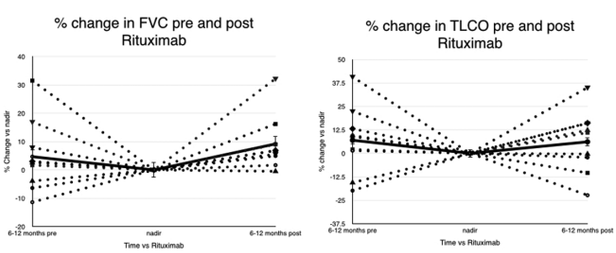
Introduction Rituximab is a chimeric monoclonal antibody against CD20 that depletes B-lymphocytes. There is increasing evidence for its use in Scleroderma ILD.[1] Recently it has been reported as rescue therapy in patients with connective tissue disease related severe fibrotic lung disease who have failed conventional immunosuppression.[2] It remains unclear which patients are most likely to benefit from this potent immunosuppressive treatment. We review here the experience of the Bristol Interstitial Lung Disease service in use of Rituximab in a subset of patients with myositis (Anti-synthetase syndrome and Dermatomyositis).

Methods We retrospectively reviewed the case notes of 10 patients with severe and progressive ILD despite immunosuppression with Cyclophosphamide and Mycophenolate Mofetil, who had received salvage treatment with Rituximab. Serial pulmonary function tests, 6 min walk distances and HRCT appearances (as assessed by a Thoracic radiologist) were compared in the year before and after Rixtuximab therapy. Changes in physiological variables compared to nadir at treatment were compared with paired-samples T-Test.



Abstract S8 Figure 1

Results The average age of the patients was 49.8 (range 26.9–72.99), with 7/10 female. 4 patients had dermatomyositis, while 6 had Anti-Synthetase Syndrome (2 Anti-Jo1, 2 Anti-PL12, 1 Anti-PL7, 1 Anti-PM-Scl). There were complete lung function data available for 9 patients and 6MWD data for 6 patients.

CT appearances stabilised in all 9 patients with follow-up scans available, with significant improvement in 2 (1 after a second pulse of Rituximab).

FVC improved after treatment by an average of 9.2% (p = 0.023, 95% CI 1.67–16.76), with TLCO improving by an average of 6.1% (NS). Figure shows% change in FVC and TLCO leading to and after therapy. 6MWD remained stable.

There were no adverse events reported.

Summary Our experience adds to the growing evidence to support the use of Rituximab in severe CTD-ILD, and suggests that a subset of patients with myositis may show good therapeutic response.

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S9 ACUTE INFLAMMATORY PRESENTATION ASSOCIATES WITH SURVIVAL IN INTERSTITIAL LUNG DISEASE AND EXTRACORPOREAL MEMBRANE OXYGENATION-REQUIRING SEVERE RESPIRATORY FAILURE: A SINGLE CENTRE CASE SERIES

L Starsmore, B Lams, S Agarwal, A Nair, R Preston, N Barrett, G Glover, N Ioannou, C Langrish, D Wyncoll, CIS Meadows. *Guy's and St Thomas' NHS Foundation Trust, London, UK*

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Introduction Patients with interstitial lung disease (ILD) and severe respiratory failure (SRF) requiring mechanical ventilation are widely perceived to have poor outcomes. A therapeutic strategy incorporating extracorporeal membrane oxygenation (ECMO) improves all cause SRF survival. There exist no data on the use of ECMO in severe ILD. ECMO may offer lung rest, reduce the inflammatory burden associated with mechanical ventilation and allow time for effective immunosuppression. We hypothesised that the use of ECMO and early immunosuppression increases survival in patients with ILD in whom mechanical ventilation was failing.

Methods Retrospective interrogation of a single centre ECMO database for patients with ILD between 2011 and 2014. Variables collected included diagnosis; immunosuppression regimen; duration of symptoms prior to ECMO initiation; serum biochemistry; clinical severity score (SOFA) and survival to ECMO decannulation, ICU discharge and at 6 months. ECMO centre admission computed tomography (CT) thorax scans were independently analysed for pattern and degree of abnormality by two radiologists. Variables were compared between responders (those who survived without lung transplant) and non responders (composite group of those who died and one patient who survived with lung transplantation). Two-tailed t-tests were used for all comparisons.

Results 12 patients with an ILD diagnosis who received ECMO were identified. ECMO and ICU survival was 58.3%. The group of responders had a shorter duration of symptoms prior to ECMO (p = 0.04), a higher CRP (p = 0.046), a higher SOFA