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

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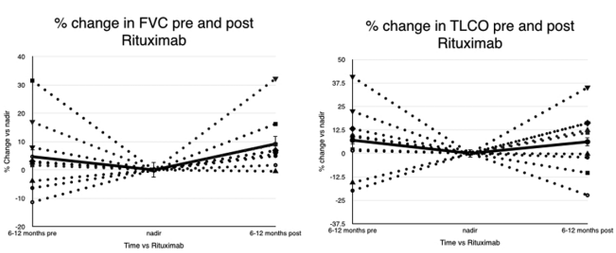
Introduction Rituximab is a chimeric monoclonal antibody against CD20 that depletes B-lymphocytes. There is increasing evidence for its use in Scleroderma ILD.[1] Recently it has been reported as rescue therapy in patients with connective tissue disease related severe fibrotic lung disease who have failed 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

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10.1136/thoraxjnl-2014-206260.15

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

Abstract S9 Table 1

Case Number	Diagnosis	Immunosuppression Regimen	Age	Onset of symptoms to ECMO initiation (days)	CT (%)	DADCT Total Abnormality (%)	SOFA Score	CRP
	Eosinophilic pneumonia;							
1	Hypereosinophilic syndrome	IVMP	30	21	100	82.5	7	344
3	Eosinophilic pneumonia	IVMP	26	12	0	80.0	9	285
10	Organising pneumonia	IVMP + CYC	52	30	10	66.8	12	226
11	Acute interstitial pneumonitis	IVMP + RTX	44	17	100	88.8	11	497
	Anti-Jo 1 associated acute interstitial							
12	pneumonitis	IVMP + CYC + RTX	42	25	10	82.1	11	350
	DD: AIP, eosinophilic pneumonia,							
13	Organising pneumonia	IVMP	35	19	90	91.7	13	266
Mean			**38.2**	**20.7**	**51.7**	**82.0**	**10.5**	**328**
	NON-RESPONDERS							
2	Acute interstitial pneumonitis	IVMP + CYC	54	129	88.4	68.4	6	70
4	Idiopathic pleuropulmonary fibroelastosis	None	45	140	81.3	81.2	7	117
	Acute Interstitial pneumonitis;	IVMP + CYC + RTX plus heart lung						
5	Organising pneumonia	transplant	46	26	50	85.9	9	153
6	Unclassifiable	IVMP + CYC + RTX	20	46	100	96.7	7	66
7	Bleomycin-induced fibrosis	IVMP	46	29	82.5	85.8	9	111
	Clinically amyopathic							
	Dermatomyositis with acute interstitial							
8	pneumonitis	IVMP + RTX	29	172	100	95.6	4	71
Mean			**40**	**90.3**	**83.7**	**85.6**	**7.0**	**98**
P-values			**0.79**	**0.04**	**0.19**	**0.53**	**0.01**	**0.046**

Key: IVMP: Intravenous Methylprednisolone, RTX: Rituximab, CYC: Cyclophosphamide

score (p = 0.01) and a lower preponderance of diffuse alveolar damage (DAD) on CT (p = 0.19) although there was no difference in overall extent of CT abnormality. (Table 1).

Conclusions The use of ECMO and early immunosuppression led to a 58.3% survival in a group of ILD associated SRF who would otherwise have been highly likely to die. The responders were characterised by a more acute and more inflammatory presentation. We suggest that ECMO and immunosuppression should be considered in patients with ILD and SRF who are failing mechanical ventilation.

S10 COST BURDEN OF N-ACETYLCYSTEINE (NAC) IN ADULT PATIENTS WITH IDIOPATHIC PULMONARY FIBROSIS

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Introduction and objectives New data from the US IPFNET PANTHER Study[1] has failed to demonstrate efficacy of NAC in adult IPF patients with mild to moderate disease. However, use of NAC in adults with Idiopathic Pulmonary Fibrosis (IPF) is commonplace in the UK[2] despite weak clinical evidence and limited support from clinical guidelines. NICE recently estimated that between 30 and 45% of patients with moderate IPF are treated with NAC monotherapy at an annual cost of £158 per patient[3]. We wanted to estimate the cost burden of NAC prescribing in England based on the actual acquisition cost to the NHS.

Methods We obtained the actual prices of NAC at a dose of 600 mg TDS from 11 different sources in England including IPF specialist centres, UK Medicines Information and guidance from Area Prescribing Teams and applied the average price into the NICE IPF costing template assuming 45% of moderate IPF (just over 3000 patients) patients receive NAC and 90% are still taking treatment at 52 weeks.

Results The average annual cost of NAC from 11 different sources was £675.63 (425% greater than NICE cost assumptions) with costs ranging from £144–£1078 per annum. This equates to an estimated annual cost of NAC in England of £2,070,266.

Conclusion NAC is unlicensed with a recent trial demonstrating limited benefit in treating IPF. The estimated annual cost burden of NAC to the NHS in England is very high. In light of the current financial position of the NHS more should be done to reduce the use of ineffective treatments that offer poor value for money. Physicians should re-evaluate the use of NAC in the management of IPF.

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S11 PIRFENIDONE POST-AUTHORISATION SAFETY REGISTRY (PASSPORT)–INTERIM ANALYSIS OF IPF TREATMENT

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Introduction Pirfenidone (Esbriet®) is approved for mild/moderate idiopathic pulmonary fibrosis (IPF). PASSPORT is a post-authorisation safety registry required by the European Medicine Agency.

Objective To present interim data from PASSPORT.