Midland Rheumatology Society
Spring Meeting 2017
Friday 10 March 2017
Fawsley Hall Hotel
Fawsley, Northamptonshire, NN11 3BA
https://www.handpickedhotels.co.uk/fawsleyhall
Free car parking is available on-site

08.45  Coffee and Registration
09.20  Welcome and Introduction
09.30  Clinical Papers (Oral Abstracts)
      1. Clinical Audit: Tocilizumab Induced Neutropenia
         J Pinnell, T Potter
      2. Long term outcomes of daily oral vs. pulsed intravenous cyclophosphamide for ANCA-associated vasculitis in a non-trial setting
         J Royle, J La-Crette, P Lanyon, A Ferraro, A Butler, F Pearce
      3. Optimizing The Assessment Of Spondyloarthropathy Among Ophthalmologists – A Regional Survey
         S Goel, P Kumar, A Moorthy
      4. Is there an Ethnic variation in Acceptance of Biologic therapy? A University Hospital Experience
         H Selvaskandan, H Al-Ani, A Moorthy
10.30  Clinical Cases
11.00  Coffee Break
11.15  Clinical Papers (Oral Abstracts)
      5. Is Hydroxychloroquine associated with decreased mortality among people with Rheumatoid Arthritis?
         H Fleet, F Pearce, P Lanyon, A Abhishek, M Grainge
      6. Tocilizumab for the Management of Rheumatoid Arthritis: Discontinuation due to inefficacy and toxicity – Experience from a large teaching hospital
         E Byrne, P Mark, S Khalid, K Graves, K-P Kuet, R F Kilding, J R Maxwell, M Akil
      7. Use of the DETECT Protocol for early Pulmonary Artery Hypertension diagnosis in patients with Systemic Sclerosis in everyday clinical practice
         C Koutsianas, S Subasinghe, K Douglas
      8. Quality improvement in fibromyalgia and inflammatory arthritis provision: a service evaluation of social deprivation in new fibromyalgia and inflammatory arthritis referrals
         M McCormack, J Bateman
12.15  Cytokines Control of Inflammation  
Professor Simon Jones  
Professor of Immunology, Cardiff Institute of Infection and Immunology School of Medicine

13.00  Lunch and Poster Viewing  
West Midlands Specialised Rheumatology CRG Meeting

14.00  Managing Moderate Disease Activity in Rheumatoid Arthritis  
Professor David L Scott  
King’s College, London

15.00  Applying COM-B to Medication Adherence  
Dr Christina Jackson

15.45  Tea/Coffee

16.00  Clinical Cases

16.30  BSR Update from Regional Chairs and MRS Business

17.30  Close

18.00  Dinner in the Salvin Boardroom

This Meeting is Approved by Royal College of Physicians, London  
(6 CPD points applied for)

Programme kindly supported by  
Celgene Limited, Bristol-Myers Squib, Novartis, Merck, Napp, Abbvie, Sobi, Lilly

The sponsors have provided funding for an exhibition stand/space at this educational meeting and have had no control over the agenda, speaker choice or content of the meeting. In accordance with the ABPI code of practice, the funding obtained is solely for the Midland Rheumatology Society Meeting on the 10th March, 2017 and is not intended for use for the after course dinner.
Clinical Papers
Background
Tocilizumab is an interleukin-6 (IL-6) receptor blocker that is an effective treatment for rheumatoid arthritis (RA) and juvenile idiopathic arthritis (JIA). However, its use can be complicated by neutropenia in over a quarter of patients requiring adjustment of the treatment dose.\(^1\),\(^2\),\(^3\)

Objectives
We audited our use of Tocilizumab to establish whether its dose was being appropriately adjusted following neutropenia and to identify any undesirable consequences of this adjustment.

Methods
We examined the clinical records of all of the patients who have ever received Tocilizumab from the Rheumatology Department at University Hospitals Coventry and Warwickshire NHS Foundation Trust (UHCW), UK. A proforma was used to document any incidents of neutropenia, any subsequent change to the dose of Tocilizumab used, and the effect of any dose adjustment. Use of Tocilizumab was compared to the standard established within the manufacturers Summary of Product Characteristics (SPC).\(^2\),\(^3\)

Results
19 patients were identified as having received Tocilizumab (18 female, 1 male, mean age 50.1). 15 patients had RA and 4 had JIA. Tocilizumab was received intravenously (IV) or subcutaneously (SC).
10 patients developed neutropenia with 38 occurrences of grade 1, 21 grade 2, 4 grade 3, and 0 grade 4 neutropenia. 5 patients had their treatment temporarily withheld due to neutropenia, 1 appropriately. 3 patients had their doses of Tocilizumab reduced in response to neutropenia, 1 inappropriately. 1 patient began treatment on a reduced dose of Tocilizumab but developed grade 3 neutropenia.
3 patients had incidents of neutropenia before receiving treatment with Tocilizumab, 2 of whom developed neutropenia whilst receiving Tocilizumab. Neutropenia was never associated with the development of infection. However, 5 patients did develop infections whilst receiving Tocilizumab, one of whom later died.
Of the 4 patients receiving reduced doses of Tocilizumab, 2 achieved therapeutic effect and 2 required their doses to be increased again. 3 had further episodes of neutropenia.

Conclusions
Neutropenia was common amongst our patients but was typically low grade, transient and not associated with infection. Tocilizumab was withheld or reduced more often than the SPC would require.
Half of the patients receiving reduced dose Tocilizumab still had good clinical effect in keeping with findings in the literature.\(^4\),\(^5\) This raises the possibility of using lower doses of Tocilizumab than we do currently thereby reducing the risk of side effects and the cost of treatment.

References
LONG TERM OUTCOMES OF DAILY ORAL VS. PULSED INTRAVENOUS CYCLOPHOSPHAMIDE FOR ANCA-ASSOCIATED VASCULITIS IN A NON-TRIAL SETTING
Jeremy Royle, Jonathan La-Crette, Peter Lanyon, Alastair Ferraro, Amanda Butler, Fiona Pearce
Nottingham University Hospitals

BACKGROUND Cyclophosphamide (CYC) may be given as daily oral tablets (PO), or pulsed intravenous (IV) therapy to induce remission in ANCA-associated vasculitis (AAV). A previous trial (n=149) and extended follow up study (n=134), demonstrated that IV administration was associated with fewer infections but a greater risk of relapse. In an audit of AAV outcomes, we evaluated this in an unselected observational cohort.

METHODS All patients diagnosed with AAV and treated with CYC in our centre between 2006 – 2013 were identified by multiple case ascertainment strategies. We assessed differences in mortality, relapse, neutropenia and infection using Kaplan-Meier methods, cox regression and logistic regression as appropriate. Unadjusted and adjusted (for age, sex and renal function) values were calculated.

RESULTS 114 patients were identified: 57 received PO and 57 received IV treatment. Baseline characteristics were significantly different; patients receiving PO treatment were older (median age 72 vs. 55 yrs, P=<0.05), had higher serum creatinine (mean creatinine 295 vs. 80 µmol/L, P=<0.05) and almost universal renal involvement (98% vs. 46%, P=<0.05). ANCA serology was predominantly MPO in the PO group (56%) and PR3 in the IV group (61%), P=0.02. Median follow up was 4.0 and 5.0 years respectively. Overall, 22 patients died in 542 patient-years of follow up (mortality rate of 40.5 per thousand person-years). One year survival was 86.0% in PO and 98.2% in IV patients. One year relapse-free survival was 80.7% in PO compared to 87.3% in IV patients. Hazard ratios for the whole follow up period are listed in table 1. During the first year after treatment, neutropenia occurred in 10/57 (18%) PO and 3/55 (5%) IV patients, adjusted OR 4.4 (0.5-42.7, P=0.19), whilst infection requiring hospital admission occurred in 16/57 (28%) and 9/55 (16%) respectively, P=0.23. Cancer occurred in 5/57 PO and 3/55 IV patients (P=0.50).

CONCLUSIONS Patients who received PO and IV cyclophosphamide were significantly different at baseline. There was no difference in survival or relapse rates after adjusting for these differences. There was a trend towards more neutropenia and increased admissions with infections in the PO group, but these were not statistically significant – however the sample size was small. In this unselected cohort of patients undergoing remission induction therapy for AAV, rates of admission with infection appear to be higher than those from a previously published RCT. This may help inform clinicians when advising patients of their treatment related risks.

Table 1. Outcomes of patients receiving PO and IV cyclophosphamide.

<table>
<thead>
<tr>
<th>Outcome</th>
<th>Unadjusted HR/OR for PO CYC</th>
<th>Adjusted HR/OR* for PO CYC</th>
<th>P Value (Adjusted HR/OR)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Death (during follow up)</td>
<td>3.8 (1.4-10.4)</td>
<td>1.3 (0.1-15.2)</td>
<td>P=0.84</td>
</tr>
<tr>
<td>Relapse (during follow up)</td>
<td>0.4 (0.2-0.9)</td>
<td>1.0 (0.3-2.9)</td>
<td>P=0.10</td>
</tr>
<tr>
<td>Neutropenia (first year)</td>
<td>3.7 (1.0-14.2)</td>
<td>4.4 (0.5-42.7)</td>
<td>P=0.19</td>
</tr>
<tr>
<td>Admission with infection (first year)</td>
<td>2.0 (0.8-5.0)</td>
<td>2.2 (0.6-8.6)</td>
<td>P=0.23</td>
</tr>
</tbody>
</table>

* Adjusted for age, sex and renal function (eGFR)
OPTIMIZING THE ASSESSMENT OF SPONDYLOARTHROPATHY AMONG OPHTHALMOLOGISTS – A REGIONAL SURVEY

S. Goel* 1, P. Kumar2, A. Moorthy3

1.Fourth Year Medical student, University of Leicester
2.Consultant Ophthalmologist, University Hospitals of Leicester NHS Trust, UK
3.Consultant Rheumatologist, University Hospitals of Leicester NHS Trust, UK

Background: Spondyloarthritis (SpA), is an under diagnosed condition with an average delay in diagnosis of up to 8 years. This delay is due to non-specific symptoms whose onset is in young patients who are unlikely to present to healthcare services and presentations to varied specialties due to extra-articular manifestations. 30% of Ankylosing Spondylitis (AS) patients have acute anterior uveitis (AAU) at some point due to the link with HLA B-27. AAU may precede the diagnosis of AS by several years. Previous studies have shown that up to 40% people with AAU have undiagnosed SpA, and are not routinely seen in rheumatology services due to lack of symptom and risk recognition (1). Early diagnosis in this group is essential to reduce the delay in diagnosis and improve the outcome of SpA.

Aim: 1. To identify the current assessment of inflammatory back pain, in patients with recurrent AAU, by local ophthalmologists.
2. To identify ways to improve cross specialty referral and reduce delay in diagnosis of SpA.

Methods: This is a prospective, questionnaire based semi qualitative study. The target population includes specialist ophthalmology trainees, consultants and GPs with a special interest in ophthalmology across the East Midlands. The questionnaire was piloted locally at a regional uveitis meeting. The improved questionnaire was then electronically communicated to ophthalmologists across East Midlands. The data was collated and analyzed using smartsurvey software.

Results: We received 61 responses with a response rate of 51%. 57 responses (n=57) were included for analysis. The respondents largely comprised doctors working at university hospitals (74%) and consultants (45%). 17% of the respondents were not confident with taking an inflammatory back pain history. Of the remaining 83%, only 67% respondents would normally think to ask about all the main symptoms of inflammatory back pain. Therefore 44% respondents were not competent in taking an inflammatory back pain history. 54% of respondents would routinely ask about symptoms of SpA in patients with recurrent AAU, even though 79% would test for HLA-B27. However, 42% of respondents would never refer a recurrent AAU patient to a rheumatologist for any reason. Most respondents felt that work pressure was the most likely reason for deficit of proper SpA assessment and referrals, followed by lack of support by local and national evidence based guidelines.

Conclusions: Our survey demonstrates a clear knowledge gap among ophthalmologists regarding inflammatory back pain and the importance of assessing it in recurrent AAU patients. This can be improved by educating ophthalmologists about SpA and developing local guidelines about patients with HLA B-27 recurrent AAU. Combined with effective communication between rheumatologists and ophthalmologists, this will increase cross specialty referrals and hopefully reduce the delay in diagnosis of SpA.

IS THERE AN ETHNIC VARIATION IN ACCEPTANCE OF BIOLOGIC THERAPY?
A UNIVERSITY HOSPITAL EXPERIENCE

Dr. H. Selvaskandan, Dr. H. Al-Ani, Dr. A. Moorthy
Department of Rheumatology, University Hospitals of Leicester, Leicester, United Kingdom

Background: Ethnic variation in medication adherence & preference is well documented (1). While usually a reflection of patient autonomy, the issue takes significance when it impacts on a clinician’s ability to provide effective evidence based care. Indeed, race has an effect on the outcome of rheumatological disease (2), likely for both biological and psychosocial reasons. Cross sectional studies from United States of America (USA) found ethnic minorities were less likely to be on a biologic for a rheumatological disease compared to Caucasians, consistent even after adjustment for education & insurance (3). UK based studies found similar results (4), although few investigated the disparity in the acceptance of biologics between ethnicities. Leicester has an ethnically diverse population, where addressing such disparities is crucial in delivering effective and equal health care.

Methods: Our objective was to determine any disparity in acceptance of biologic therapy exists, when offered in person, in a healthcare system free at the point of access, between White British and other ethnicities. Data was collected from nurse led Biologics therapy clinics, from October 2016 to December 2016. All patients referred were deemed suitable for biologic therapy as per NICE guidelines by a Rheumatologist, and were attending the clinic for counselling and assessment prior to consenting for biologics therapy. Proformas were prepared and piloted. Improved proforma with information including demographic, disease & treatment details, as well the outcome of the consultation (biologic accepted or rejected) was used to collect data. The collated data were then analysed using EXCEL spreadsheet.

Results: Data was collected from 55 patients (n=55). It is interesting to note Sex distribution was nearly equal (54% female). 57% of the total sample were White British. The remaining 43% cohort included; Indian, Bangladeshi, Pakistani, White Other, Asian other, African Caribbean and Any other mixed race. The most common disease necessitating referral for a biologic was rheumatoid arthritis (53%), followed by psoriatic arthritis (23%). In total 16% of patients rejected a biologic drug, of which 66% were ethnic minorities. The rejection rate among ethnic minorities was thus 24% compared to 10% in the White British cohort. The highest rejection rate was within the Any Other Mixed Ethnicity cohort (100%), followed by the Bangladeshi cohort (50%). Of note, all patients who rejected biologic therapy from an ethnic minority background did not speak English as their first language. Rejection rates were highest in the Spondyloarthropathies (21%).

Conclusions: Our results demonstrate a disparity between the White British population and other ethnicities in the acceptance of biologics, despite one to one counselling. This can have detrimental impacts on treat to target concept and disease progression, and thus will be further investigated & addressed.

Background: Rheumatoid Arthritis (RA) patients have increased mortality compared to the general population. Hydroxychloroquine prescription associates with reduced risk of cardiovascular events and other morbidities in people with RA. The aim of our study was to determine if Hydroxychloroquine decreases mortality in people with RA.

Methods: We included incident cases of RA in the Clinical Practice Research Datalink (CPRD) 2006-2014. To ensure cases had definite RA they needed a record of both a Read code for the diagnosis of RA and GP prescription of a DMARD. We calculated a propensity score for the likelihood of receiving hydroxychloroquine depending on baseline characteristics, and used this to create two cohorts of RA patients exposed to and not exposed to hydroxychloroquine with similar baseline characteristics, thus avoiding confounding by indication. We used a landmark approach to avoid immortal time bias, i.e. patients who live longer have a greater chance of being prescribed hydroxychloroquine. We defined exposure to hydroxychloroquine as receiving ≥1 GP prescription within 6 months of diagnosis, and began follow up for death, the outcome of interest at 6 months onwards. We used Cox-regression to determine whether Hydroxychloroquine reduced mortality, and compared cause of death between the exposed and unexposed cohorts using chi-square.

Results: 1,420 RA patients exposed to hydroxychloroquine were matched to 1,420 unexposed RA patients. Using Cox-regression, no association was found between Hydroxychloroquine prescription and overall risk of mortality after adjusting for ...(hazard ratio 1.25, 95% confidence intervals 0.90-1.74). However, there was a significant difference in the cause of death between the exposed and unexposed cohorts P=0.044. Deaths in the exposed cohort were less likely to be due to cardiovascular disease, cancer or Idiopathic Pulmonary Fibrosis.

Conclusions: In this study, we did not find that Hydroxychloroquine had a significant effect on overall mortality, but reduced deaths due to cardiovascular disease, cancer or Idiopathic Pulmonary Fibrosis. Our decision to classify exposure solely on whether patients were prescribed hydroxychloroquine in the 6 months following diagnosis may have introduced bias towards no effect. The difference in the cause of death data between exposed and unexposed cohorts supports existing evidence that Hydroxychloroquine may have beneficial effects on reducing comorbidities associated with RA.
Background: Tocilizumab (TCZ) is a humanised anti interleukin-6 receptor antibody licensed for use for the treatment of moderate to severe Rheumatoid Arthritis (RA) as monotherapy or in combination with methotrexate (MTX).

Aims: To describe the use of TCZ for RA in a large UK teaching centre.

Method: A retrospective case note review of all adult patients receiving TCZ either alone or in combination with other DMARDS, for the treatment of RA between April 2009 and January 2017 in Sheffield, UK.

Results: 131 patients received TCZ for RA: 44 (33.5%) as a monotherapy, 87 (66.5%) in combination with other DMARDS. Overall 47 (35.8%) discontinued TCZ because of inefficacy or adverse events. 4 (3%) patient discontinued TCZ due to primary failure, 2 were on MTX, 1 on Leflunomide (LEF), 1 monotherapy; 2 were seronegative, 2 RF and CCP positive; all 4 had prior anti TNF and 1 had rituximab (RTX). 9 patients (6.8%) stopped TCZ because of secondary failure: mean treatment duration 26.5 months (14-50 months); 7 CCP +ve, 4 RF +ve, 2 double negative; 3 took TCZ as monotherapy, 4 with MTX, 1 with Sulfasalazine (SZP), 1 with Hydroxychloroquine (HCQ); all had prior anti TNF, 7 RTX, 1 Abatacept. 24 (18.3%) patients stopped TCZ because of adverse events: 7 (5.3%) due to recurrent infections, mean treatment duration 6 months [2-36 months] - 5 had prior anti TNF and 1 had anti TNF and RTX; 5 (3.8%) due to abnormal LFTs , mean treatment duration 25 months [2-66 months] 3 on MTX, 1 on HCQ and 1 monotherapy; 4 patients (3%) cancer; 4 (3%) rashes. There were 4 deaths in our cohort: 2 due to cancer and 2 unknown cause. We have not seen any cases of infusion reaction, diverticular perforation or reactivation of tuberculosis.

Conclusions: our real life data on the safety profile of TCZ in the treatment of adult patients with RA is consistent with clinical trial data and is similar to other biological drugs used in the treatment of RA. We have seen a relatively low rate of withdrawal due to primary and secondary inefficacy.
USE OF THE DETECT PROTOCOL FOR EARLY PULMONARY ARTERY HYPERTENSION DIAGNOSIS IN PATIENTS WITH SYSTEMIC SCLEROSIS IN EVERYDAY CLINICAL PRACTICE

Christos Koutsianas, Sunari Subasinghe, Karen Douglas
Department of Rheumatology, The Dudley Group NHS Foundation Trust, UK

Background: The survival rate of Systemic Sclerosis (SSc) patients with Pulmonary Arterial Hypertension (PAH) is significantly lower than SSc patients without PAH and delay in diagnosis contributes significantly to this outcome. The DETECT study provides a sensitive tool for early identification of patients at risk of PAH and advocates evaluation with Right Heart Catheterisation (RHC). Few published data look into its implementation in everyday clinical practice and compare it to annual echocardiographic (TTE) screening.

Methods: We aimed to apply the DETECT algorithm (DA) on 57 consecutive patients with a diagnosis of SSc that had at least one visit to Russells Hall Hospital Scleroderma clinic from February to November 2016. Data was prospectively collected on Excel (clinical history, physical examination, immunological status, treatment, PFTs, NT-proBNP, urate, ECG, TTE, CXR and HRCT results). DETECT scores were obtained using the online calculator (www.detect-pah.com) and compared with the TTE probability for PAH as per the 2015 ESC/ERS Guidelines. Chi Square, Spearman’s and ANOVA tests (SPSS version21) were used to compare variables between screening strategies.

Results: 31 patients with a full set of data were included in the final analysis. 93.5% were female with a mean age of 64.4±12.9 years, 87% had limited cutaneous (lc) SSc, 58.1% positive anticientromere antibody and mean time since diagnosis was 6.2±3.2 years. The majority of patients (80%) were asymptomatic for PAH. Mean DLCO was 60.9±18.3% predicted (the DETECT study included on patients with DLCO< 60%). We did not exclude patients with renal insufficiency; mean eGFR was 74 ±17.7ml/min.

The implementation of the DA recommended RHC in 18/31 patients (DETECT step 2 score ≥ 35, 58%, see table 1) compared to just 4/31 (13%) based on TTE by 2015 ESC/ERS guidelines. The additional 14 patients identified by the DA were found to have no statistically significant differences in SSc type, dyspnea symptomatology, immunosuppressive treatment, presence of interstitial lung disease (ILD), age, time from diagnosis or symptom onset, eGFR and CRP compared to patients where RHC was not recommended. From the 18 patients identified by DETECT, only 4 reported shortness of breath. 2/18 had existing cardiac conditions (aortic stenosis, left ventricular hypertrophy), while 9/18 had ILD (5 mild, 2 moderate, 2 severe as per HRCT).

Conclusions: Implementation of the DA in a non-selected SSc patient population (the majority asymptomatic) is more sensitive for RHC referral than annual TTE screening in clinical practice. Recommendations for RHC per DETECT score are difficult to predict on symptom and other traditional clinical parameters. The use of the DA will lead to increased RHC referrals. Cardiologists may need education on this new method for screening and although the ESC/ERS 2015 guidelines discuss the DA, they conclude that its cost-effectiveness has not been clarified as compared with symptom-based detection.

References:
<table>
<thead>
<tr>
<th>DETECT step 2 score</th>
<th>Age (years)</th>
<th>eGFR (ml/min)</th>
<th>CRP (mg/L)</th>
<th>DLCO (%)</th>
<th>Disease duration (years)</th>
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<tbody>
<tr>
<td>&lt;35</td>
<td>63.8 ±13.5</td>
<td>76 ±15</td>
<td>5 ±3</td>
<td>68 ±19</td>
<td>7 ±2</td>
</tr>
<tr>
<td>35-39</td>
<td>60.6 ±12.2</td>
<td>79 ±12</td>
<td>5 ±3</td>
<td>60 ±13</td>
<td>6 ±4</td>
</tr>
<tr>
<td>≥ 40</td>
<td>69.0 ±12.7</td>
<td>65 ±23</td>
<td>9 ±15</td>
<td>51 ±19</td>
<td>6 ±4</td>
</tr>
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</table>

<table>
<thead>
<tr>
<th>DETECT step 2 score</th>
<th>Interstitial lung disease</th>
</tr>
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<tbody>
<tr>
<td></td>
<td>No</td>
</tr>
<tr>
<td>&lt;35</td>
<td>10</td>
</tr>
<tr>
<td>35-39</td>
<td>5</td>
</tr>
<tr>
<td>≥ 40</td>
<td>3</td>
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**Tables 1 & 2**

DETECT step 2 score ≥ 35 recommends RHC. The range 35-39 represents a lower risk range, but up to 15% of patients have undiagnosed PAH.
QUALITY IMPROVEMENT IN FIBROMYALGIA AND INFLAMMATORY ARTHRITIS PROVISION: A SERVICE EVALUATION OF SOCIAL DEPRIVATION IN NEW FIBROMYALGIA AND INFLAMMATORY ARTHRITIS REFERRALS

McCormack M.¹, Bateman J.²

¹. Year four Medical Student, University of Birmingham Medical School
². Consultant Rheumatologist, Royal Wolverhampton NHS Trust, WV10 OQP

Background
Previous work has shown a relationship between social deprivation and musculoskeletal symptom severity¹. Audit data shows FMS represents >25% of new patient referrals to our service, which operates across two main sites, to a diverse population across several CCGs. Accessible open access government data now provides unprecedented detail across 32,844 English neighbourhoods, categorising every neighbourhood into deciles with a global deprivation index, the Index of Multiple Deprivation (IMD) into deciles, (1-10 most to least deprived) drawn from 7 components². To develop our service we seek to understand epidemiological factors in both fibromyalgia syndrome (FMS) and inflammatory arthritis (IA). We report initial findings using this dataset.

Methods
We conducted a service evaluation of consecutive new patient referrals diagnosed with FMS between 1.1.16 and 1.10.16. Routine metrics were collected (age, gender, postcode, clinic site, and disease indices [2010 ACR Diagnostic criteria]). FMS cases were anonymously age and gender matched against new patients presenting to the same clinics diagnosed with IA (all causes) over 18 months. We derived the IMD scores from patient postcodes, used descriptive statistics and non-parametric statistical analysis to explore differences between IMD in the two groups. We used open access data visualisation techniques to form an interactive map of our population.

Results
We matched 55 new patients diagnosed with FMS to 55 new patients with IA. The groups were similar (Table 1), with high FMS morbidity scores, presenting the full range of IMD deciles for both FMS and IA. There were no significant differences for IMD overall, or any subcomponents (Table 1). We produced an interactive topographical population map which visualises our service provision (catchment 700km², distance 31km N-S, 51km E-W; max. travel: FMS 25km, IA 26 km).

<table>
<thead>
<tr>
<th></th>
<th>FMS (mean, SD)</th>
<th>IA (mean, SD)</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td>45.5 (11.1)</td>
<td>46.5 (10.7)</td>
<td>.631*</td>
</tr>
<tr>
<td>Gender</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Female (53)</td>
<td>Male (2)</td>
<td></td>
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<tr>
<td>Deprivation indices</td>
<td></td>
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<tr>
<td>Selected IMD</td>
<td>Mean ‘IMD’ decile (SD, median, range)</td>
<td>Mean ‘IMD’ decile (SD, median, range)</td>
<td>p-value</td>
</tr>
<tr>
<td>FMS disease scores</td>
<td>Mean ACR 1990 TP, ACR2010 WPI, 2010 SS Score</td>
<td>Mean ACR 1990 TP, ACR2010 WPI, 2010 SS Score</td>
<td>.624*</td>
</tr>
<tr>
<td>Gender</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Female (53)</td>
<td>Male (2)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>4.8 (2.6, 5, 1-10)</td>
<td>5.1 (2.6, 5, 1-10)</td>
<td></td>
</tr>
</tbody>
</table>

Table 1. Population metrics for Fibromyalgia (FMS) and inflammatory arthritis (IA)

Discussion
This approach to combining disease categories, disease metrics, with deprivation measurements allows us to better understand the population we serve, and its needs in terms of education, employment, and geography. We found no significant differences in IMD between referrals with FMS or IA. These data underline the need for us to provide accessible services to cater for all socio-economic groups. This approach offers the potential to examine treatment presentation and response by IMD scores between and within diseases, allowing targeting and impact assessment of patient-tailored management. This innovative data visualisation of our service will be used to plan future service development. This utilisation of open access data offers potential for wider application in service planning across the region, and may be of interest to other providers.
References:
Posters
REVIEW OF PREGNANCY OUTCOMES IN SPONDYLOARTHROPATHY IN A UNIVERSITY TEACHING HOSPITAL

A. Shajpal 1, M. Khare 2, A. Moorthy 3

1Specialist registrar in Obstetrics and Gynaecology, 2Maternal and Fetal Medicine Consultant, 3Rheumatology Consultant, University Hospitals of Leicester, Leicester, United Kingdom

Background: Spondyloarthritis (SpA) is a chronic inflammatory condition of the spine affecting mainly the male population however the incidence amongst the female population is increasing. The peak incidence of SpA is most common in the reproductive age group. There is a lack of focus on pregnancy in SpA as compared to other autoimmune condition such as Lupus and Rheumatoid arthritis, due to a male predominance. There is a paucity of information (1) on fertility and pregnancy outcomes in this condition compared to other diseases and this may lead to inequality in healthcare delivery.

Objectives:

• To review the pregnancy outcomes in women with SpA in our unit
• To review ankylosing spondylitis activity during pregnancy.
• To improve the quality of care in this group of patients by developing local pathways.

Methods: This is a retrospective case review of pregnancies in women with SpA booked at a large tertiary teaching hospital over three years between January 2014 and December 2016. We have an annual delivery rate of 11,000 maternities. Maternity electronic database and clinic diaries were used to identify the cases. A standardised proforma was used to collect and collate the data for demographics, pre pregnancy counselling, disease activity and pregnancy outcome.

Results: Six pregnancies were identified in the study period. All patients were under the care of Rheumatology. The maternal age range was between 28 and 35 years. The BMI ranged between 18 and 37. The patients ethnicity included 5 caucasian and one Asian. Five women had previous pregnancies and one was in her first pregnancy. Two of the multiparous women had previously delivered by caesarean section. Three of the six women suffered from anxiety and/or depression and one had fibromyalgia. Two of the six patients were not on any medication at the start of pregnancy and didn’t require any during pregnancy. Four women needed various analgesics and one patient was on sulfasalazine but stopped this at 5 weeks’ gestation. NSAID was stopped in 3 women after confirmation of pregnancy. One patient who was on Anti TNF therapy discontinued the drug preconception. We observed 50% of the patients attended specialist maternal-fetal medicine clinics and had anaesthetic input during the pregnancy. One patient saw a physiotherapist and accessed hydrotherapy during pregnancy.

Two of six patients delivered preterm (<37 weeks) and 4 delivered at term (>37 weeks). Of the preterm deliveries, 1 went into spontaneous labour not related to disease flare and the other was delivered electively for fetal concerns. All 6 women were delivered by caesarean section. One of these was planned as an elective caesarean for maternal request due to difficulty abducting legs. All the remaining caesarean deliveries were for obstetric indications not related to SpA.

Conclusions: This small observational study did not highlight any worsening SpA disease activity or poor pregnancy outcome. However, there is a possible need for care pathways for managing this group of patients which would help standardise the care during pregnancy. Good and effective communication between Rheumatologist and Obstetricians is essential to improve the quality of care for this group of patients.

References:

A CASE OF LEFT SIDED ACCESSORY SACROILIAC JOINT MIMICKING SPONDYLOARTHROPATHY

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Background
Accessory sacroiliac (SI) joints are not rare, occurring in 10-30% of the population. They are formed between the sacral and iliac articular surfaces at the posterosuperior portion of the SI joint, from the level of the first to the second sacral foramen. Their aetiology remains unclear, as it is not certain if the ASIJ is a congenital condition or an acquired joint. They are often affected by degenerative changes, with findings of reduced joint space, sclerosis, osteophytes and ankylosis common. These changes may manifest with symptoms of low back, and buttock pain and may mimic focal sacroiliitis on plain film imaging. We present the case of a 35 year old lady who presented with inflammatory-sounding back pain, whose MRI Spine revealed the cause as an accessory sacroiliac joint rather than an axial spondyloarthropathy.

Case
A 35 year old lady presented with a 2 year history of low back pain. She had early morning stiffness lasting 2 hours, stiffening after inactivity and buttock pain. She reported recent history of increasing lethargy. Her mother had Psoriasis and her sister had Lupus. On examination, there was no peripheral synovitis. She had several Smythe fibromyalgic tender points. There was mild restriction in range of movement of her lumbar spine and investigations revealed a CRP of 1, with ANA, Rheumatoid Factor, CCP and HLA B27 negative. Lumbar Spine and Sacroiliac joint x-ray showed sclerosis of both sacroiliac joints, suggestive of mild bilateral sacroiliitis. Subsequent MRI Spine with STIR sequences demonstrated the presence of a left accessory sacroiliac joint. This finding was associated with mild subchondral marrow oedema and sclerosis. She was booked for radiologically guided steroid injection to the left sacroiliac joint.

Discussion
It is still a controversy whether accessory SI joint is a congenital or acquired disorder. Evidence for the accessory SI joint as an acquired lesion is the presence of fibrocartilage around the articular surface in cadaveric specimens. Furthermore, a significant increase in the incidence of accessory SI joints has been reported with advancing age. They are also associated with obesity and multiparity. What is clear however, is that they are frequently symptomatic. One series reported that low back pain was noted in 65 out of 102 patients with accessory SI joints. This same cohort also exhibited a high frequency of radiographic abnormalities with 52 out of 102 having abnormal radiographs showing sclerosis (41/52), osteophytes (10/52), and ankylosis (8/52). One hypothesis to account for the propensity for degenerative change of accessory sacroiliac joints is that the angle of these joints is more angulated and oblique than the normal sacroiliac articulation.

Conclusion
We present a case of a 35 year old lady with inflammatory sounding back pain. MRI imaging revealed the presence of an accessory sacroiliac joint. Accessory SI joints are easily misinterpreted as sacroiliitis on radiographs, especially when they are associated with degenerative arthritis. Our case highlights that a confident diagnosis of sacroiliitis can be difficult using plain film radiographs alone. It also highlights the superiority of MRI for patients with inflammatory back pain because as well as being a sensitive test for sacroiliitis it could reveal another cause for a patient’s pain.
AN UNUSUAL CAUSE OF A SWOLLEN LEG
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Background
The presentation of a Baker’s cyst arising in the knee being confused with DVT is well known but the phenomenon that a bursa originating from the hip causing swelling of the whole leg being confused with DVT is not widely reported in the literature. It is important that this is recognised as part of the differential diagnosis for a swollen leg.

Case
A 66-year-old lady presented with a progressively swollen and tender left leg. She has a background of small cell lung cancer, and a previous DVT. She had an ultrasound scan to exclude DVT but results were inconclusive due to vessel wall thickening from her previous DVT. The sonographer noted an incidental finding of a collection anterior to the hip joint. She was treated as DVT with rivaroxiban. She revisited A&E 3-weeks later with increased swelling and tenderness. Ultrasound scan was repeated but it did not show any new findings. The rheumatologist reviewed her and noted a palpable but painless swelling in her left groin. The radiologist then reviewed her previous pelvic CT scan, carried out for staging of malignancy, and noted the presence of an iliopsoas bursa compressing on the common femoral vein. The bursa was aspirated and injected with steroid under ultrasound guidance. The swelling in her leg resolved after the aspiration.

Discussion
The iliopsoas bursa is the largest synovial bursa in the body. It communicates with the hip joint by a defect in the thinnest part of the capsule between the pubo-femoral and iliofemoral ligaments. Amongst its commonest associations are osteoarthritis and rheumatoid arthritis of the underlying hip, although trauma, gout, pseudogout, tuberculosis and avascular necrosis have been reported as well. The earliest and the most common symptom associated with iliopsoas bursitis is hip pain. It may give rise to signs and symptoms which may be due to femoral vein or nerve compression, or compression of lower limb lymphatics. A palpable groin mass is not an uncommon mode of presentation. There have been a few of cases reported in the literature of iliopsoas bursitis mimicking deep vein thrombosis. The investigation of this condition is by a combination of ultrasound and CT or MRI scanning. Ultrasound helps to confirm the cystic nature of the condition and can be used to perform needle aspiration. The definitive treatment for iliopsoas bursitis surgical excision, although previous studies have shown a very low rate of recurrence after aspiration and injection of steroids in the bursa.

Conclusion
Iliopsoas bursa is a rare cause of swelling in the groin and should also be considered in the differential diagnosis of a patient with unilateral leg swelling. Urgent imaging combining US and MRI will exclude venous occlusion and thrombosis, whilst affording an accurate diagnosis and aid planning of management.
AXIAL SPONDYLOARTHRITIS IN NOONAN’S SYNDROME – A PREVIOUSLY UNREPORTED ASSOCIATION

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Noonan’s syndrome is an autosomal dominant genetic syndrome with variable penetrance, where affected individuals have multiple congenital anomalies. Various musculoskeletal abnormalities have been described in past publications, including short stature, chest wall deformities (most commonly pectus excavatum and/or pectus carinatum), spinal deformities (scoliosis, kyphosis, abnormal lordosis, cervical stenosis), hip dysplasia, facial dysmorphism, cubitus valgus, and joint hyperextensibility. However, here we describe the case of a male patient with Noonan’s syndrome who has been diagnosed with ankylosing spondylitis, an association that has previously been unreported in the literature to date.

Our patient is a 29 year-old male with Noonan’s syndrome, which was treated with growth hormone injections from the ages of 6 to 18 years, and musculoskeletal abnormalities in the form of cervical spondylosis and severe spinal fusion, which had previously been attributed to his Noonan’s. He was referred to Rheumatology in mid-2016 for advice on optimising management of his significantly reduced spinal mobility, inflammatory back pain and increasing hip pain, for which X-rays of his pelvis were done exhibiting marked reduction of both acetabular joint spaces, bony overgrowth of the femoral necks and obliteration of both sacroiliac joints.

Clinical assessment revealed an advanced degree of spondyloarthropathy with significantly reduced lumbar flexion in the Schober’s test of +1 cm, restricted chest expansion of 1-2cm, hip abduction of 56cm, markedly increased tragus-to-wall distance of 19cm, and cervical spine rotation of only 18°. Subsequent blood results showed him to have a positive HLA-B27 status with a raised C-reactive protein of 48. This, in combination with review of his MRI and plain film images in the Rheumatology-Radiology MDT demonstrating presence of spinal inflammation leading to spinal vertebral and sacroiliac joint fusion and loss of normal curvature of the spine, confirmed a diagnosis of Ankylosing Spondylitis.

He was initially managed with Naproxen for four months and multi-disciplinary team input, including physiotherapy and occupational therapy, but as his BASDAI and VAS scores have remained high (6.8/10 and 8/10 respectively), he is currently being treated with a trial of an alternative NSAID, in the form of Eterocoxib, and hydrotherapy. Considering future management options, should his symptoms remain refractory to NSAIDs, the next step would be to commence him on biologics, either Anti-TNF or Anti-IL17 therapy.

One of the key learning points from this case is that when a patient has a congenital syndrome, there is a danger of clinicians attributing all of their symptoms to that one condition rather than considering an additional diagnosis, as was the case here. Furthermore, although in 2013 a case report of peripheral spondyloarthritis in a patient with Noonan’s syndrome was published in Brazil, after an exhaustive search through the literature in the PubMed, Medline and Cochrane databases, no case has yet been described of a Noonan’s syndrome patient with an established diagnosis of Ankylosing spondylitis. Therefore we are keen to highlight this unusual case for the education of our Rheumatology colleagues and their patients.
Zoledronic acid (ZA), a bisphosphonate administered as annual intravenous injections to treat osteoporosis, is primarily excreted through the kidneys without further metabolism. In the Summary of Product Characteristics (SPC) of ZA, it is clearly stated that for non-oncological uses of this drug, such as in osteoporosis, its use is contraindicated if estimated creatinine clearance (eCrCl) is less than 35ml/minute, calculated with the Cockroft-Gault equation using the actual body weight of patients. However, given that UK biochemistry laboratories provide an MDRD-derived eGFR, which only takes age, gender, serum creatinine and ethnicity into account, many clinicians use this result to estimate renal function when prescribing ZA instead of actively calculating the eCrCl. Unfortunately, in those of advanced age, who often have reduced muscle bulk, eGFR can be overestimated as it does not take the weight of the patient into account, unlike the Cockroft-Gault equation for estimating creatinine clearance.

In this study, we compared these two methods of measuring the renal function of all patients of 80 years of age or above who have received ZA to treat osteoporosis over the last two years at New Cross Hospital, Wolverhampton. This was done by recording the pre-dose eGFR provided by the Pathology TD-Web system, and then independently calculating the pre-dose eCrCl, using the same blood results and weight recorded at the time of the infusion, via MDCalc online which adopts the Cockroft-Gault equation to do this and also gives an adjusted eCrCl for those who are overweight. Out of a total of 44 patients, six had to be excluded due to lack of documentation available of the parameters needed to calculate the eCrCl. Out of the final group of 38 patients, 10.5% of patients (n=4) had an eGFR > 35ml/min/1.73m² and so had been prescribed ZA at the time, but on retrospective calculation were found to have an eCrCl < 35ml/min. Furthermore a further 10.5% (n=4) who were overweight had an adjusted eCrCl <35ml/min, calculated using their adjusted body weight. Consequently, more than 1 in 5 of our patients had been incorrectly been deemed eligible for treatment with ZA according to its SPC recommendations, as eGFR had been used instead of eCrCl.

Although in the long-term renal function has been found to remain unaffected by ZA, transient decline in renal function post-infusion has been noted in the literature, and this is more pronounced in patients with renal impairment. As ZA can have nephrotoxic implications in the short-term, this study highlights that clinicians should avoid relying on the MDRD-derived eGFR; rather to ensure safe prescription practice, the Cockroft-Gault method of assessing renal function should be used to determine eligibility of patients for ZA to treat osteoporosis, especially as many of these patients are elderly and/or may be at extremes of body mass.
Calcinosis is more common in juvenile-onset dermatomyositis than adult-onset dermatomyositis although in both cases it can prove resistant to treatment. We describe two patients with dermatomyositis with refractory calcinosis who have shown improvement with biologic drugs.

**Case 1** A 15-year-old girl presented with classic juvenile-onset dermatomyositis with rash, weakness and raised muscle enzymes. Five years later her inflammatory myopathy was well-controlled but she continued to have significant problems with calcinosis despite treatment with oral and IV steroids, IV immunoglobulin and methotrexate. She was given adalimumab and pamidronate with a good response and continues on treatment and follow-up.

**Case 2** A 50-year-old woman initially presented with an inflammatory arthritis and interstitial lung disease. She had rash, muscle weakness and elevated muscle enzymes. She later developed subcutaneous calcification at the elbows, knees and thighs. The calcinosis progressed despite oral and IV steroids, IV immunoglobulin and IV cyclophosphamide. She was intolerant of methotrexate and azathioprine and was given mycophenolate but continued to develop further calcinosis. She was given rituximab, this led to an improvement in the subcutaneous calcification and she went on to receive two further cycles. Unfortunately after the third cycle she was admitted with neutropenia and pneumonia. After remaining stable for 2 years the calcinosis has recently started to worsen and retreatment with lower dose of rituximab is planned.

Over the years, a number of different drugs have been used in the treatment of calcinosis, including bisphosphonates, colchicine and diltiazem. Success with biologic drugs is starting to be more widely described, although this remains limited to case reports and series. These two cases show further success with biologic drugs in managing this difficult problem.

A 73 year old female with a background of hiatus hernia, previous vasovagal syncope underwent a knee replacement after which her walking improved but there was progressive deterioration in her walking with pain in her right leg. She then noticed sensory symptoms in her feet and tingling in the little finger of her left hand radiating up to her forearm which then spread across the other fingers such that the hand became progressively weak and floppy. Recently she also noticed problems with the right shoulder which she can't move greatly. It was a fall that prompted her admission to hospital. On examination she has a complete left wrist drop, difficulty moving fingers of her left hand, an inability to elevate her right arm or flex the right elbow.

Blood tests were unremarkable: ANA-ve, ANCA-ve, ENA-ve, HIV-ve, Hep B+C -ve, complements normal, ESR 2. Proceeded to have an LP that showed 7 leucocytes (40% polymorphs, 60% lymphocytes), CSF cytology: increased numbers of uniform small mononuclear cells in keeping reactive lymphocytes, no malignant cells seen, normal CSF protein and glucose at 256 and 3.8 respectively.

MRI spine showed thickening and enhancement of C7 nerve roots on the left and an abnormal appearance to T1 root on the left which is thickened and enhanced. Nerve conduction studies demonstrated asymmetric abnormalities within the sensory and motor studies in the upper limbs in keeping a multiple mononeuritic process likely mononeuritis multiplex.

Proceeded to have CT abd/pelvis to rule out malignancy that showed a homogenous soft tissue mass at the head of the pancreas with enlarged coeliac lymph nodes. This was biopsied and the results were in keeping with diffuse large B cell lymphoma, non-germinal centre type. This is a case of mononeuritis multiplex likely secondary to lymphoma, she was treated with IV methylprednisolone initially but she responded poorly.

Learning points:
-different causes of mononeuritis multiplex
-how to investigate mononeuritis multiplex

Note: A video eliciting limb weakness will be shown as well as MRI and CT images of neck and abdomen.
A CURIOUS CASE OF GANGRENE

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A 60 year old male with a past medical history of 'antiphospholipid syndrome', CVA aged 40 on warfarin, CKD 3, HTN, T2DM presented with gangrene of the great toe, 2nd, 3rd and 4th toes, red discolouration of the forefoot as well as an absent dorsalis pedis pulse of the left leg with brittle nails and dry scaly skin. On the right leg there was only an absent dorsalis pedis pulse and red discolouration of the lower leg. Prior to admission patient sustained trauma to left leg developing cellulitis which responded poorly to antibiotics. MRA showed stenosis at the origin of the common iliac and external iliac arteries, patient proceeded to have an angioplasty but the gangrene worsened further despite intervention. Patient also developed confusion during admission. A doctor noticed 'mottling' of the skin therefore a rheumatology referral was made. A series of investigations were done which showed obscure ANCA, positive ANA homogenous 1/1600, +ve anticardiolipin, low complement levels, raised CRP at 86, deteriorating renal function with a PCR (protein creatinine ratio) of 878. At this point pt was started on IV methylprednisolone 1g for three days then high dose prednisolone. Renal biopsy was done which was unfortunately insufficient however the most likely diagnosis was lupus nephritis class 3/4, other differentials were minimal change disease and renal vein thrombosis. A skin biopsy was performed which showed non specific changes. Pt was then started on cyclophosphamide. His condition generally improved, vasculitic lesion in his legs improved including the mottling of his skin which was livedo reticularis.

Learning points:

-Differential diagnosis of necrotic skin lesions

-Prevalence of vasculitis in SLE

-Manifestations of antiphospholipid syndrome

-Diagnostic criteria of antiphospholipid syndrome

-How to investigate vasculitis

Presentations will include images of legs and angiogram.
CERVICAL VASCULITIS

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A 55 year old Caucasian woman presented to a Rheumatology clinic following a referral from a Gynaecology consultant. She had been referred to Gynaecology 4 years previously due to a borderline abnormal smear test and human papillomavirus (HPV) positivity. There was no history of abnormal vaginal bleeding. The patient had reached the menopause aged 49 and was nulliparous. Colposcopy was arranged and revealed cervical stenosis with 2 small os. Monitoring with six-monthly then annual cervical smear tests was arranged.

After 2 years of monitoring, loop diathermy was recommended by the MDT due to the persistence of abnormal cells. The cervical tissue was sent for histology, which revealed florid vasculitis with surface squamous metaplasia. A referral was made to Rheumatology to rule out systemic vasculitis.

In the Rheumatology clinic, the patient denied myalgia, weight loss, fevers, sweats, headache, scalp tenderness, rash, livedo reticularis, Raynaud’s phenomenon or digital ulceration. She described occasional metacarpophalangeal joint discomfort but there was no history of swelling or stiffness. Systems review was normal and in particular there were no respiratory, cardiovascular or gastrointestinal symptoms. Examination was normal with no evidence of inflammatory arthritis, vasculitic rash, nail fold changes, focal neurology or pulmonary pathology.

Past medical history included psoriasis, mild atopy and sarcoidosis. The latter had been quiescent for 14 years, having previously been treated with low dose methotrexate. There was a family history of breast cancer and the patient was monitored with yearly mammograms. There was no significant occupational history. She was a non-smoker and drank 6 units of alcohol per week.

Bloods showed normal full blood count, U&Es, LFTs, TSH, CK, bone profile and immunoglobulins. ESR was 4 and CRP 1. ANCA, anti-CCP, rheumatoid factor and cryoglobulins were negative. ANA was positive 1:320 homogenous with normal complement and negative ds-DNA and ENA. Urine dip was clear. Chest x-ray was clear with no evidence of hilar lymphadenopathy. ACE result is pending.

As there were no features of systemic vasculitis, local treatment by the gynaecology team was recommended. She will remain under Rheumatology review to monitor for systemic features.

Single organ vasculitis (SOV) can be a separate entity or a precursor to systemic disease. Cervical vasculitis is the most common manifestation of SOV in the female genital tract. It has been linked to HPV and intraepithelial neoplasia. Histological features often resemble Polyarteritis Nodosa (PAN). More rarely Giant Cells are seen. Only 10% of cervical vasculitis is associated with systemic manifestations. Where it does, PAN and giant cell arteritis are the most common diagnoses. Treatment is with excision, or with corticosteroids or immunosuppression in systemic disease.

Learning points:
- Cervical vasculitis is most commonly an isolated single organ vasculitis
- Treatment of local disease is with excision
- 10% of patients have systemic features and investigation for systemic vasculitis is indicated
SARCOIDOSIS - AN INTERESTING CASE

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Introduction - Sarcoidosis is a granulomatous disease. The disease usually begins in the lungs, skin, or lymph nodes. Less commonly affected are the eyes, liver, heart, and brain. Any organ, however, can be affected. The signs and symptoms depend on the organ involved. Often there are no, or only mild, symptoms. Presenting here is an interesting case of sarcoidosis.

Case - 39 year old female with background history of gastroesophageal reflux disease and sickle cell trait was referred to ENT department with stuffiness of nose, hoarseness of voice. Nasoendoscopy showed friable mucosa, clots and oedematous vocal cords. CT scan of sinuses showed significant mucosal thickening. Biopsy of nasal mucosa was reported as granulomatous disorder. ANCA test was negative. ESR was 50. Chest Xray showed bihilar lymphadenopathy. CT chest showed additional mediastinal lymphadenopathy and pulmonary nodules. Pulmonary function test showed mild restrictive pattern, ACE levels were normal. She was diagnosed as sarcoidosis and started on prednisolone with good response and then changed to methotrexate.

2 years later she presented with subacute headache. CT head was normal. Lumbar puncture showed raised proteins. MRI head showed bifrontal haziness suggestive of encephalopathy. She was diagnosed as neurosarcoidosis and prednisolone 40mg was started. Headache improved and steroid was tapered and stopped.

She then developed diarrhoea and infective causes have been ruled out. She is awaiting colonoscopy.

Conclusion - It is very important to include sarcoidosis in differential diagnoses for many systemic conditions and patients are likely to be seen and managed by multiple specialties.
TWO CASES OF PL-7 POSITIVE ANTISYNTHETASE SYNDROME PRESENTING WITH INTERSTITIAL LUNG DISEASE - DELAYED AND EARLY DIAGNOSIS

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Background: Interstitial lung disease (ILD) can dominate the clinical picture in antisynthetase syndrome (ASS). Especially in the presence of non-Jo-1 aminoacyl-tRNA synthetase (ARS) antibodies, it can precede myositis for many years and patients presenting with ILD may be mistakenly diagnosed with idiopathic interstitial pneumonia. Anti PL-7 is one of the eight recognised ARS autoantibodies, it is associated with ILD and is detected in 2-5% of patients with polymyositis. Over the past few years laboratory tests for non Jo-1 ARS antibodies, including anti-PL-7, have become commercially available allowing for an earlier diagnosis and treatment of patients with ASS.

Objective: We present two cases of patients with PL-7 positive ASS presenting initially to respiratory medicine with interstitial lung disease.

Cases description: A 44 year old Caucasian man was diagnosed with ILD-NSIP by the respiratory physicians in 2011. He had then presented with dyspnoea; pulmonary function tests revealed restrictive pattern, HRCT of the chest showed cryptogenic organising pneumonia and lung biopsy suggested NSIP. Autoimmune screen was negative. He was treated mainly with moderate to low doses of steroids and he remained relatively stable for 4 years. In December 2015 he developed dysphagia, notable myalgia with raised CK (3944), Raynaud’s phenomenon and some deterioration in lung disease. He was then referred to rheumatology department. In retrospect it seemed that he had steroid responsive myalgia at onset and a transiently high ALT since 2011. MRI revealed symmetrical oedema of the thigh muscles bilaterally. He had repeat Hep2 immunofluorescence and cytoplasmic staining was seen. The myositis specific antibodies screen revealed anti PL-7 antibodies positive. Anti- PL-7 ASS was diagnosed and he was treated with steroids and immunosuppressives including cyclophosphamide and cyclosporin.

The second case is about a 64 year old woman presenting to the respiratory clinic with cough, shortness of breath and Raynaud’s phenomenon. The chest HRCT showed mixed pattern of NSIP and bronchiectasis with bronchiolitis. Autoimmune screen was negative for ANA but equivocal for ENAs. Myositis specific antibodies screen was performed and revealed positive anti-PL-7 antibodies. The patient was then reviewed in Rheumatology clinic and it emerged that she was feeling progressively weaker although objectively muscle strength was preserved. CK was checked and was raised 1700. Anti-PL-7 ASS was diagnosed and the patient has been started on Prednisolone and Azathioprine.

Conclusions: The above two cases underscore the importance of searching for anti-PL-7 and other ARS antibodies in patients with ILD. The early detection of an underlying ASS can influence the treatment and prognosis for these patients. In the first case the syndrome remained occult for five years and there was a delay in the initiation of appropriate immunosuppression. In the second case, early identification of the underlying connective tissue disease influences the management of the interstitial lung disease and we anticipate that it will improve the overall prognosis. Clinical multidisciplinary follow-up of these patients, with a high level of alertness to specific clinical signs is of high importance and we have now available the appropriate tools for the early detection of an ASS with extra-muscular manifestations and rare specificities of ARS.
We present a rare case of primary Sjögren’s syndrome (pSS) and granulomatosis with polyangiitis (GPA) in a 57 year old lady. She presented in October 2015 with pain in her left wrist and shoulder, cervical spine, knees and ankles. She reported problems with low back pain since her early twenties, for which she’d had facet joint injections. She had a six year history of dry eyes and mouth. She was troubled by fatigue and poor sleep.

She was known to have depression, fibromyalgia, hypertension, well controlled asthma, a raised body mass index and a fatty liver. She’d also developed pancreatitis post cholecystectomy. There was a family history of arthritis, but the patient was unsure of the type.

On initial examination there was no evidence of rash, synovitis or joint deformity. There were multiple tender trigger points. A Schirmer’s test was positive. Routine bloods were normal. Anti-nuclear antibodies and anti-Ro antibodies were positive. Unstimulated sialometry showed significant dryness (0.1ml produced in 15 minutes), and an ultrasound scan showed subtle changes in the submandibular glands consistent with Sjögren’s. The patient was prescribed topical agents for symptom control.

Almost a year later in September 2016, the patient presented feeling generally unwell with one stone of weight loss over the previous six weeks. She had developed a new rash, joint pain and swelling, and severe eye pain, which was diagnosed as anterior uveitis. On examination there was a vasculitic rash on the limbs, and synovitis in the right metacarpophalangeal joints and both wrists. Shoulder movements were reduced on the right. There were 2 pluses of blood and 1 plus of protein in the urine. Routine bloods were normal except for a raised alkaline phosphatase and gamma-glutamyl transpeptidase. The C-reactive protein was 240. Cytoplasmic-ANCA (c-ANCA) against proteinase 3 was positive. A chest radiograph was normal. A skin biopsy showed a leucocytoclastic vasculitis.

GPA was diagnosed and the patient was started on 30mg prednisolone daily. Shortly afterwards, mycophenolate mofetil was commenced. Initially the patient showed a good response. However, when the prednisolone was reduced to 15mg daily, she began to feel unwell again with recurrence of the rash, hand and ankle swelling and oral ulcers. The prednisolone was increased back to 20mg daily and the mycophenolate was titrated to 3g daily in divided doses. The patient continues to be under regular review.

Granulomatosis with polyangiitis (GPA) is an anti-neutrophil cytoplasmic antibody (ANCA) associated vasculitis affecting small vessels. Primary Sjögren’s syndrome (pSS) is a systemic autoimmune disease characterised by lacrimal and salivary gland dysfunction. A rare association between the two conditions has been reported.

Karargyris et al describe a 61 year old lady presenting with fever and myalgia. She had a positive Schirmer’s test, anti-nuclear antibody (ANA), anti-Ro antibodies and c-ANCA. A salivary gland biopsy revealed lymphoplasmacytic infiltrates, and a biopsy of the pulmonary opacities noted on chest radiograph showed granulomatous lesions with vasculitis of medium-sized vessels typical of GPA.

Yazisiz et al describe a 60 year old lady presenting with dyspnoea. She had a positive Schirmer’s test and scintigraphy. ANA, anti-Ro antibodies and C-ANCA against proteinase 3 were positive. A chest radiograph showed bilateral reticulonodular shadowing and a biopsy of the lesions revealed granulomatous inflammation with giant cells.

Sato et al report a similar case of Sjögren’s syndrome with GPA-like involvement of the lungs. They describe a 69 year old lady who presented with fever and cough and was noted to have sicca symptoms. A chest radiograph showed bilateral areas of consolidation. A Schirmer’s test was...
positive, as were anti-Ro and anti-La antibodies, and ANCA against myeloperoxidase. Sialoscintigraphy was positive and a lung biopsy revealed infiltration of the alveoli by lymphocytes with scattered neutrophils and microscopic vasculitis of the small arteries.

These three cases report a relationship between pSS and a limited pulmonary form of GPA. Our patient did not have lung involvement but did have a more widespread vasculitis affecting her skin, joints, eyes and kidneys. The more generalised nature of the vasculitis makes the case more akin to those described by Guellec et al. They describe a 58 year old lady with a two year history of pSS, with joint and peripheral nervous system involvement, presenting with worsening paraesthesia. During admission she developed haemorrhagic shock, found to be secondary to abdominal aneurysms. She had a positive c-ANCA with antibodies against proteinase 3, and a biopsy of the colon showed necrotising vasculitis.

The second patient, a 58 year old lady, had a two year history of sicca symptoms and widespread chronic urticaria. She presented with a new rash, lymphadenopathy and ear chondritis. She was ANA, anti-Ro and c-ANCA positive. She had antibodies against proteinase 3. A Schirmer’s test was positive. A salivary gland biopsy showed non-specific interstitial sialadenitis and a skin biopsy showed a leucocytoclastic vasculitis.

These cases, along with our own, show that GPA can occur with pSS. Thus far, GPA has been diagnosed simultaneously with or subsequently to a diagnosis of pSS; it has not yet been reported prior to a diagnosis of pSS. Guellec et al suggest that GPA could be considered as an uncommon but possible complication of pSS. They also suggest that the onset of GPA correlates with the presence of extra-glandular manifestations of pSS. This was not found to be the case in our patient who only had sicca symptoms.

The association between GPA and pSS is rare. The exact nature of the relationship remains unclear. However, these cases show that patients with pSS and a positive ANCA can develop a systemic vasculitis, and should therefore be carefully monitored.

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