O1. THE IMPACT OF MODERN MANAGEMENT ON OUTCOMES OF JIA COMPARED WITH HEALTHY CONTROLS

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Background: Development of modern therapeutic approaches over the last decade has improved short-term well-being in JIA patients. This study examines disease damage and body composition (BC).

Aims: Comparison of JIA patients’ clinical well-being with matched controls assessing BC, cross-sectional inflammatory markers and the levels of joint and uveitis-associated damage relating to their management.

Methods: 112 JIA patients (disease duration >5 years) were cross-sectionally compared with 127 age, sex, ethnicity and deprivation matched controls, measuring inflammatory markers (hsCRP, IL-6 and ICAM-1), growth and BC [height, weight, fat mass (FM/H2), free fat mass (FFM/H2), BMI and skinfold thickness]. Clinical examination recorded disease damage, supported by case-note review.

Results: Compared with controls, patients >16 years had indistinguishable values of hsCRP (P = 0.39) and ICAM-1 (P = 0.15), but >16 years had higher hsCRP, ICAM-1 and IL-6 (P = 0.001). Patients <22 years had higher Z-BMI (P = 0.019), Z-waist circumference (P = 0.001) and FM/H2 [using Tanita equations (P = 0.007), sum of four skinfolds (P = 0.008)], with 21.3% meeting obesity criteria (vs controls 5.6%) with no association with non-JIA risk factors for obesity. Compared with controls, all anthropometric equations revealed no difference in patients >22 years. Of 90 patients, Group A never required DMARDs, Group B had modern therapeutic approaches (prompt MTX (SSZ in 1) and rapid escalation to biologics) and Group C had late DMARDs/biologics intervention despite persistent inflammation. Feet, knees and temporomandibular joints were most commonly damaged. A positive correlation was found between delay in starting modern therapy and severest joint damage outcome (Table 1).

Conclusions: It is of new concern that high obesity levels in the youngest patients were significantly greater than in controls. Normal inflammatory markers, reduced frequency and severity of joint damage in the UK Juvenile-Onset Systemic Lupus Erythematosus Cohort. Arthritis Rheum 2012;64:2356–65.

Disclosure statement: The authors have declared no conflict of interest.

References

O2. NOVEL URINARY BIOMARKERS OUTPERFORM STANDARD BIOMARKERS IN JUVENILE LUPUS NEPHRITIS: A PROSPECTIVE LONGITUDINAL VALIDATION STUDY

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Background: Juvenile onset SLE (JSLE) is a severe autoimmune condition with LN seen in up to 80% of patients [1]. Current methods of monitoring renal disease activity over time rely on a variety of standard laboratory markers and the use of disease activity tools such as the British Isles Lupus Assessment Group (BILAG) index score. Improving monitoring and predicting disease activity may allow earlier intervention and improve long-term renal outcome.

Aims and Methods: This prospective longitudinal study aimed to identify whether standard and/or novel biomarkers are useful for monitoring and predicting LN disease activity. Using patients recruited to the UK JSLE study, urine and blood samples were collected during routine clinical reviews. The study had full ethical approval.

Results: The JSLE cohort (n = 64), seen at three [interquartile range (IQR) 2–5] clinical reviews over 364 (182–532) days, were aged 14 (11.6–15.8) years and 80% female. Active renal episodes (23% total; renal BILAG A/B) had significantly increased concentration of monocytic chemoattractant protein 1 (MCP1), neutrophil gelatinase-associated lipocalin (NGAL), ESR, anti-dsDNA, urine albumin:creatinine ratio (UACR), creatinine and reduced complement 3 (C3), C4 and lymphocytes. Multivariate analysis demonstrated MCP1 and C3 as independent variables (P < 0.001) for active renal disease. MCP1 was an excellent predictor of improved renal disease [area under the curve (AUC) 0.81; P = 0.013; concentration 343 pg/ml, specificity 71%, sensitivity 70%]; NGAL was a good predictor of worsened renal disease activity (AUC 0.76; P = 0.04; concentration 30 ng/ml, specificity 61%, sensitivity 60%). Urine MCP1 and NGAL changed as subsequent renal disease changed (MCP1 P = 0.015; NGAL P = 0.038). Standard markers could not predict disease activity.

Conclusion: We have demonstrated that biomarkers (MCP1, C3) perform well for monitoring renal disease in JSLE, and novel biomarkers (MCP1, NGAL) outperform standard markers for predicting change. Biomarker-led monitoring may facilitate titration of medication and allow earlier diagnosis and intervention. Collaboration with industry to develop point of care urine biomarker testing is now in progress.

Disclosure statement: The authors have declared no conflict of interest.

References

O3. ADOLESCENT FRIENDLINESS: ALL TALK, NO ACTION?

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Background: Adolescence occurs between ages 10 and 19 years [1] with complex biological and emotional developmental tasks [2, 3]. Adolescent-specific issues (e.g. risk-taking behaviour and emerging independence) should be addressed in health care consultations. Training in this area is scarce [4] and tools such as HEADSS [5] can improve professionals’ engagement in adolescent issues. Assessment of the young people friendliness of services has recently been encouraged in the Department of Health You’re Welcome (YW) measure [6, 7], which provides standards to measure whether services meet the needs of adolescents. Sheffield has a seamless adolescent rheumatology clinic across both Sheffield Children’s Hospital (SCH) and Sheffield Teaching Hospitals (STH) [8]. This project asks how young person friendly (YPF) our service is.

Objectives: To evaluate the YPF of Sheffield adolescent rheumatology clinic (SARC) and the YPF of individual adolescent consultations.

Methods: (i) Service evaluation using YW toolkit criteria to evaluate the YPF of SARC. Benchmark Meets You’re Welcome standard requires 95% of the total criteria (including nine crucial criteria) to be achieved. (ii) Audit to assess the YPF of individual consultations against nine standards from YW and HEADSS. Forty-five sets of notes were audited (selected randomly from the adolescent cohort across
O4. MINIMAL DISEASE ACTIVITY IN A CLINICAL COHORT OF CHILDREN WITH JIA: RESULTS FROM THE CHILDHOOD ARTHRITIS PROSPECTIVE STUDY

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Background: Despite recent advances in the management of children with JIA, complete clinical remission is uncommon. A state of minimal disease activity (MDA) is a useful therapeutic goal. Recently, a definition of MDA has been proposed [1], with discrete definitions for oligoarticular [physician global assessment (PGA) ≤2.5 cm and swollen joint count (SJC) = 0] and polyarticular [PGA ≤3.4 cm, parent global assessment ≤2.1 cm and SJC ≤1] disease patterns. The definition cannot be applied to enthesitis-related arthritis (ERA) but is applicable to all other ILAR subtypes.

Objective: To describe the proportion of children with JIA reaching MDA over time, during the first 2 years after diagnosis.

Methods: All children in the Childhood Arthritis Prospective Study (CAPS) included. MDA was determined for all children (except ERA) at presentation, and at 6, 12 and 24 months after diagnosis and presented for the entire cohort and within the oligoarticular and polyarticular subsets. The proportion of children in MDA with oligoarticular and polyarticular pattern disease was compared at all time points, using χ² statistics.

Results: To July 2012, 1236 children had been recruited to CAPS: median age at disease onset 6.7 years [interquartile range (IQR) 2.8–10.7], median disease duration 5.2 months (IQR 2.4–10.6), 64% female. Data to calculate MDA were available in 799, 673, 625 and 473 children at baseline and 6, 12 and 24 months, respectively. A minority of children were in MDA at presentation but this increased to 61% at 1 year and remained stable at 2 years (Table 1). Significantly more children with oligoarticular pattern disease were in MDA at all time points.

Conclusion: Although the aim of treatment in JIA is remission, only 65% of children with JIA had reached MDA after 2 years of follow-up, suggesting a significant level of ongoing disease activity. Understanding why a high proportion of children persist with higher levels of disease activity, despite advances in therapies, may help to improve targeting of therapies in the future.

Disclosure statement: The authors have declared no conflict of interest.

Table 1 JIA patients in MDA by time

<table>
<thead>
<tr>
<th>Group</th>
<th>% (total scores available)</th>
<th>Baseline</th>
<th>6 months</th>
<th>12 months</th>
<th>24 months</th>
</tr>
</thead>
<tbody>
<tr>
<td>Whole cohort</td>
<td>7 (799)</td>
<td>52 (673)</td>
<td>61 (625)</td>
<td>65 (473)</td>
<td></td>
</tr>
<tr>
<td>Oligoarticular pattern</td>
<td>9 (637)</td>
<td>55 (525)</td>
<td>65 (466)</td>
<td>71 (340)</td>
<td></td>
</tr>
<tr>
<td>Polyarticular pattern</td>
<td>0 (162)</td>
<td>41 (153)</td>
<td>51 (156)</td>
<td>50 (133)</td>
<td></td>
</tr>
<tr>
<td>P (oligo vs poly)</td>
<td>&lt;0.001</td>
<td>0.001</td>
<td>0.001</td>
<td>&lt;0.001</td>
<td></td>
</tr>
</tbody>
</table>

References


Results: Neither hospital within SARC, reached the Meets You’re Welcome benchmark, and neither met all crucial criteria. Criteria missed included robust mechanisms for ensuring confidentiality/consent and involvement of young people and non-YPF access and clinic environments. Adolescent issues were poorly documented in medical notes. Easy questions were often recorded but not more difficult questions. Young people were not documented to receive written information. P.O.N. observed that adolescent issues were often discussed, and adolescent information leaflets given, but neither process was documented. However, letters communicating outcomes between professionals and to patients were well documented.

Conclusion: The service evaluation revealed work needed at an organizational level within both hospitals in SARC, with implications beyond rheumatology. This project has resulted in a cross-hospital working group to formulate the YPF organizational strategy, improve training of professionals and ensure equality of adolescent services in the two hospitals. The mismatch between what should be discussed and what is documented in clinics is worrying. We have introduced a proforma sticker for notes to ensure that all adolescent-specific issues are documented. Patient information packs will ensure all young people receive important adolescent-specific information. A new patient steering group will formalize patient involvement. These measures should ensure that YPF is not all talk and no action.

Disclosure statement: The authors have declared no conflict of interest.

References


O5. AUDIT OF INCIDENCE OF SCARRING FOLLOWING STEROID TENDON INJECTIONS FOR TENOSYNOVITIS IN CHILDREN WITH JIA: A SINGLE-CENTRE EXPERIENCE

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Background: Superficial skin scarring is a well-recognized complication of steroid joint injections in JIA with an approximate incidence of ≤2%. With increasing recognition of tenosynovitis, more steroid tendon injections are being performed. There are no published studies on the incidence of scarring secondary to steroid tendon injections in children with JIA.

Objective: The aim of this study was to evaluate the incidence of scarring following steroid tendon injections done by an interventional paediatric radiologist in children with JIA.

Methods: All children with JIA who underwent tendon injections in Bristol Children’s Hospital were identified from the Paediatric Rheumatology database. A proforma was devised and relevant information collected from medical notes, including number of episodes of tendon injection and specific tendons injected. Scarring was identified either by documentation in notes or a telephone call to the parents, with explanation of anatomical landmarks.

Results: 17 patients with JIA had steroid tendon injections over a period of 2.5 years. The median age was 9 years (range 2.5–16). There were 24 episodes of tendon injection, with a total of 36 tendons injected, all around the ankle. Tenosynovitis was diagnosed by US in 83% of episodes and MRI scan in 17%. 58% had arthritis of the adjacent joint. The median interval between detection of tenosynovitis and injection of tendons was 7 weeks. All injections were done using triamcinolone hexacetonide and performed under general anaesthetic with US guidance by an interventional paediatric radiologist. Lidocaine was used for local anaesthetic. Scarring of injection site was identified in 9/24 (37%) episodes and 6/17 (35%) patients. Deep scars occurred in three patients. Five patients had repeat tendon injections, of whom four had scarring (80%). There was no clear pattern of association of scarring with dose of steroid or administration of lidocaine.
Conclusion: In our retrospective series of 17 JIA patients who had steroid tendon injections, scarring was observed in 35%. In patients in whom tendon injections were repeated, the incidence of scarring was 80%. Our data seem to suggest that steroid tendon injections for tenosynovitis in JIA patients might be associated with a higher incidence of scarring as compared with joint injections. This information might be useful when obtaining informed consent for the procedure. Prospective studies recruiting a larger cohort of patients are needed to substantiate these findings.

Disclosure statement: The authors have declared no conflict of interest.

Abstracts

1. FOOT ORTHOSES IN JIA: RESULTS FROM A RANDOMIZED CONTROLLED TRIAL
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Background: Currently there is limited evidence supporting podiatric treatment of children with JIA. This study aimed to investigate whether pre-formed, cost-effective foot orthoses (FOs) impacted on pain, quality of life (primary outcomes) and/or gait parameters (secondary outcomes) in children affected by JIA.

Methods: Intervention was blinded to the patients. The trial group was supplied with FOs, with the addition of chairside corrections and the control insole instead was made without corrections. Both insoles had the same black EVA top cover. Primary outcome measures were recorded at each of three data recording appointments over a 6-month period, using validated questionnaires such as VAS, Childhood Health Assessment Questionnaire (CHAQ) and PedsQL. Tekscan equipment (F-Scan and HR Walkway) measured in-shoe pressure and force data with and without orthotic intervention, using same type of sensors of equal resolution. Multiple foot strikes and repetitive gait patterns were compared pre and post-treatment. The HR Walkway captures multiple sequential footsteps during barefoot walking; allowing barefoot and in-shoe measurements to be compared.

Results: Sixty children were recruited: 48.3% (n=29) control and 51.7% (n=31) active-treatment group. Within the control group 20.7% (n=6) of subjects were male. Within the active-treatment group, 29% (n=9) of subjects were male. Age range was 5–18 years; mean age for the control group was 11.17 (s.d. 3.51) and for the trial group was 10.64 (s.d. 3.84). In order to attribute any effect solely to the FO intervention, details of changes of medication and/or new joint injections were recorded during the trial: 65.5% (n=19) of the control group were on stable medication; 74.2% (n=19) of the control group were on stable medication; 74.2% (n=19) of the control group were on stable medication. Overall, 99.4% (n=31) active-treatment group. Within the control group 20.7% (n=6) of patients receiving active treatment were on stable medication. Overall, 99.4% (n=179/180) of appointments were completed and contributed to this preliminary data analysis. Significant improvement was identified in the primary outcomes favouring active treatment with regard to pain and quality of life measures: VAS (P <0.05); CHAQ (P <0.05); PedsQL paediatric-generic (P >0.05); PedsQL paediatric rheumatology (P =0.03); PedsQL parent generic (P >0.05); PedsQL parent rheumatology (P <0.05). Significant differences were also identified between the groups for gait time, stance time, total plantar surface, heel contact, midfoot, fifth metatarsal head and distal phalanx.

Conclusions: The results show that FOs are effective in improving pain, quality of life and most gait parameters in JIA children. This trial also provides new evidence for the role of podiatrists working within paediatric hospitals and private practices.

Disclosure statement: The authors have declared no conflict of interest.

2. GET A GRIP
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Background: The Childhood Myositis Assessment Score (CMAS) is a validated assessment tool for JDM. The CMAS 14 correlates highly with the Childhood Health Assessment Questionnaire (CHAQ) score and manual muscle testing. It correlates moderately with the physician-assessed global disease score [1]. It takes ~20min to complete and tests endurance and stamina. Grip strength is important as a measure of general health and is often used in screening for normal motor function. It is one of the most reliable clinical measurements for estimation of strength and is used extensively on adults [2]. We routinely test hand grip strength using the Clifton NJ Marjar hand hydraulic dynamometer on patients with JDM.

Objective: To analyse hand grip strength in JDM patients and compare this with CMAS scores.

Methodology: This was a retrospective audit. CMAS and Jamar grip strength scores were recorded from the medical notes for 12 patients. Grip strength in JDM patients was recorded during the trial: 65.5% (n=19) of the control group were on stable medication; 74.2% (n=19) of the control group were on stable medication. Overall, 99.4% (n=31) active-treatment group. Within the control group 20.7% (n=6) of the control group 20.7% (n=6) of the control group 20.7% (n=6) of the control group were on stable medication. Overall, 99.4% (n=31) active-treatment group. Within the control group 20.7% (n=6) of the control group 20.7% (n=6) of the control group 20.7% (n=6) of the control group were on stable medication. The Jamar is generally thought to be durable, cost effective and has good inter-observer and intra-observer reliability. It measures isometric grip force from 0 to 90 kg (0–200 lbs). It has five grip settings and we always use the second setting. We used the Guidelines in the American OT Journal 1986 for using the Jamar [3]. The best score of three was taken. The American OT Journal published average scores for normal children aged 6–19 (adults to 75 years).

Results: Some trends were noted: normal grip strength scores were low compared with the scores for similar aged normal children. Due to the low numbers the results were not statistically significant to conclude whether hand grip strength in JDM patients correlates with CMAS score.

Conclusion: There is evidence that grip strength is a predictor of total muscle strength in healthy children and adolescents [4]. However, there is also evidence that grip strength does not correlate with lower limb strength in patients with pathology [5]. Therefore we recommend both CMAS and hand strength are tested for JDM patients. The evidence is not clear in this area and therefore further research is required.

Disclosure statement: The authors have declared no conflict of interest.

References

3. BATH ANKYLOSING SPONDYLITIS METROLOGY INDEX IN THE PAEDIATIC POPULATION
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Background: The Bath Ankylosing Spondylitis Metrology Index (BASMI) is an objective measure of axial involvement in AS and is validated for use in adults. There are no published data relating to its use in children.

Conclusions: This pilot study was conducted to assess whether BASMI is a useful tool in children and young people with enthesitis-related arthritis (ERA).

Method: BASMI is a standardized assessment tool using five measurements (tragus–wall, lumbar flexion, intermalleolar distance, lumbar side flexion and cervical rotation). Over a 3-year period 14 children with a diagnosis of ERA were identified as suitable for assessment.

Results: Of the 14 children assessed, 12 were HLA B27+ and all had a diagnosis of ERA confirmed by a paediatric rheumatology consultant. There were 11 males and 3 females; the average age at time of initial assessment was 13.4 years; 11 of the individuals had only a single BASMI assessment completed; 3 had sequential measurements done over a 1- to 3-year time period. In the age range assessed (11–16 years), the BASMI was carried out as per the standardized adult format. It proved easy to use and took less than 5 min to complete with all participants. Those individuals who had sequential measurements showed improvement in all areas during the assessment period. In one
4. THE SAFETY AND EFFICACY OF TRIPLE IMMUNOSUPPRESSIVE THERAPY IN THE TREATMENT OF REFRACTORY CHRONIC NON-INFECTIONOUS UVEITIS IN CHILDHOOD

Jessica Little1, C. N. S. Helen Strike2, Anni Hinchingclif2, Andrew Dick3 and Ahtimalaipet Ramanan2 5. JOINT AUDIT OF UVEITIS SCREENING SERVICES FOR CHILDREN WITH JIA, ACCORDING TO BSPAR/RCOphth SCREENING GUIDELINES, AT QUEENS MEDICAL CENTRE, NOTTINGHAM AND SHEFFIELD CHILDREN’S HOSPITAL

Objectives:

To assess the safety and efficacy of triple immunosuppressive therapy in the treatment of refractory chronic non-infectious childhood uveitis.

Methods:

A retrospective study was performed on all patients diagnosed with chronic, non-infectious uveitis under the age of 16 who had at some stage been treated with triple immunosuppressive therapy for at least 6 months at the combined paediatric rheumatology–ophthalmology clinic. Triple therapy was defined as using any three immunosuppressive agents simultaneously (MTX, MMF/tacrolimus and an anti-TNF agent is the most likely combination). The main outcome measures recorded were: anterior chamber grade, ability to reduce topical corticosteroids, presence of ophthalmic complications, serious adverse events and side effects.

Results:

Nine patients were included in the analysis; all had been using any three immunosuppressive agents simultaneously (MTX, MMF/tacrolimus and an anti-TNF agent) for at least 6 months at the combined paediatric rheumatology–ophthalmology clinic.

Before triple therapy started, 14 of the 18 eyes had active anterior uveitis and were being treated with topical corticosteroid drops. Six months after commencement of triple therapy, there was a clinically significant improvement in ocular inflammation in seven of these eyes and nine were receiving a tapered dose of topical corticosteroids. Over the course of follow-up, ocular inflammation fluctuated and six of the nine patients required additional steroids for uveitis. During the follow-up period, there were three serious adverse events recorded, which were: one episode of chicken pox and two episodes of pneumonia that required admission to hospital.

Conclusion:

In terms of efficacy, initial results show triple therapy did improve control of uveitis in certain patients. However, in terms of safety, results suggest triple therapy could potentially be associated with serious adverse events.

Disclosure statement: The authors have declared no conflict of interest.

5. JOINT AUDIT OF UVEITIS SCREENING SERVICES FOR CHILDREN WITH JIA, ACCORDING TO BSPAR/RCOphth SCREENING GUIDELINES, AT QUEENS MEDICAL CENTRE, NOTTINGHAM AND SHEFFIELD CHILDREN’S HOSPITAL

Diana Ekdawy1, Gupreet Nagra2, Nikki Camina3 and Athimalaipet Ramanan2

Background: Childhood chronic non-infectious uveitis is a rare disease that is difficult to treat. In spite of current treatment regimens with two immunosuppressive agents, there is still a high rate of ocular complications in children with uveitis (~40%). This can result in significant visual impairment.

Objective: To assess the safety and efficacy of triple immunosuppressive therapy in the treatment of refractory chronic non-infectious childhood uveitis.

Methods: A retrospective study was performed on all patients diagnosed with chronic, non-infectious uveitis under the age of 16 who had at some stage been treated with triple immunosuppressive therapy for at least 6 months at the combined paediatric rheumatology–ophthalmology clinic. Triple therapy was defined as using any three immunosuppressive agents simultaneously (MTX, MMF/tacrolimus and an anti-TNF agent is the most likely combination). The main outcome measures recorded were: anterior chamber grade, ability to reduce topical corticosteroids, presence of ophthalmic complications, serious adverse events and side effects.

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Disclosure statement: The authors have declared no conflict of interest.

6. THE ROLE OF TELECONFERENCING AS AN EDUCATIONAL RESOURCE FOR PAEDIATRIC RHEUMATOLOGY TRAINEES

Mary Cruikshank2; 3; 4, Ethan Sen4, Clare Pain5 and Valentina Leone6

Background: There is increasing interest in the role that tele and video conferencing can play in access to postgraduate medical education, particularly for small specialties that are spread over large geographical areas. During the BSPAR Trainees’ Meeting in January 2012, there was request and enthusiasm for the development of further postgraduate educational resources. It was proposed that a trainees’ educational teleconference meeting be piloted for 6 months with the aim of aiding postgraduate training.

Methods: Monthly invitations were sent to all trainees on the BSPAR trainee mailing list (total n = 55, Paediatric Rheumatology Grid trainees n = 12), asking them to email the Chair if they wished to participate. Trainees were invited to present a case with literature review. PowerPoint presentations were emailed to the Chair then circulated only to those attending the meeting. Presentations did not include any patient- or hospital-identifiable information and consent had been obtained. The teleconference was financed by a combination of residual funds from the BSPAR Trainees’ Meeting and self-funding of the cost of a local telephone call.

Results: Six trainee teleconferences have been held, on the third Tuesday of each month between 8 and 9 pm, chaired by the Royal College of Paediatric and Child Health Specialist Advisory Committee (CSAC) for Higher Specialist Training in Paediatric Rheumatology trainee representative. The format of the meeting consisted of an initial discussion of CSAC/ Clinical Studies Group (CSG)/BSPAR trainee issues followed by the educational session. The mean number of trainees attending each meeting has been 7.8 (range 5–13) for 4–10 topics. Topics have included eosinophilic fasciitis, consensus...
Background: In paediatric and adolescent rheumatology (PAR), professionals lack confidence in joint examination [1], routine documentation of pGALS examination is poor [2] and yet clinical care training in busy hospital environments is challenging. Experience with patients-as-educators [3], in the context of evidence regarding simulation-based skills training, has led to an adult rheumatology virtual ward round (VWR) being established for Sheffield medical students. This has been positively evaluated [4] but this teaching method is not previously described in PAR. Local concern was expressed that children and young people’s (CYP) participation in such an innovation would be problematic. We present a multi-professional PAR education day comprising a VWR of patients from our service, problem-based learning sessions and lectures.

Objectives: To improve VWR participants’ clinical joint examination skills and pilot the extension of VWR methods to encompass CYP as patient-educators.

Methods: Forty health care professionals from paediatric and adult backgrounds (doctors, nurses, therapists, medical students) attended. The course was free with refreshments funded by Pfizer and participant/educator expenses from a charitable donation. We recruited by personal invitation 16 CYP (4–25 years) and 2 adults (30, 41 years) for the VWR. The morning session was the VWR preceded by a quiz to establish baseline knowledge. Participants in groups of five rotated through eight stations (20 min) with two/three patients and one educator. Educators were paediatric rheumatologists from the Yorkshire Paediatric Rheumatology Network Group (YPRNG) and Sheffield nurse specialists/therapists. The afternoon session comprised lectures and problem-based learning facilitated by national experts. Finally, the quiz was repeated to assess learning. Quiz results and feedback from patients, participants and educators were collected.

Results: 80% of VWR participants reported only some confidence in examining joints pre-course whereas 70% felt confident in most aspects of pGALS afterwards. Similar improvements were demonstrated in knowledge of JIA with positive participant feedback. Consistent themes from patients’/families’ feedback included being ‘keen to give something back’ and ‘wanting to help others’ by participating in the VWR. Even the youngest tolerated it well and demonstrated in knowledge of JIA with positive participant feedback. Consistent themes from patients’/families’ feedback included being ‘keen to give something back’ and ‘wanting to help others’ by participating in the VWR. Even the youngest tolerated it well and

Discussion: Feedback has been positive. Use of videoconferencing has generally worked well and technical issues have improved since its initiation. The educational sessions have provided closer links between network centres. Although geographical barriers have been overcome, finding a suitable and convenient time remains difficult. Pitching education at suitable levels for all members of the multidisciplinary team remains challenging. It is acknowledged that this method of education does not replace the advantages of face-to-face educational events but can be a useful additional resource for sharing and learning from experience.

Disclosure statement: The authors have declared no conflict of interest.

References
5. Background: In 2010 BSpar alongside the Arthritis and Musculoskeletal Alliance (ARMA) released 43 standards of care relating to the management of children with JIA [1]. The standards are aimed at achieving optimal care but recent audits [2, 3] have shown that UK paediatric rheumatology falls short of the standards, even in tertiary centres. Ideally all standards should be met, but implementation will take time and resources. It is critical that the views of young people and their families are sought when prioritizing standard implementation.

References
5. Background: In 2010 BSpar alongside the Arthritis and Musculoskeletal Alliance (ARMA) released 43 standards of care relating to the management of children with JIA [1]. The standards are aimed at achieving optimal care but recent audits [2, 3] have shown that UK paediatric rheumatology falls short of the standards, even in tertiary centres. Ideally all standards should be met, but implementation will take time and resources. It is critical that the views of young people and their families are sought when prioritizing standard implementation.
Objective: As part of the development of a regional paediatric rheumatology network in the east of England we sought the views of children and families on the standards of care and on their willingness to travel for tertiary centre care.

Method: A questionnaire was sent out to patients and parents from six centres across the region from 15 May 2012 to 15 August 2012. These included a small district general hospital (DGH), four medium sized DGHs and one teaching hospital offering tertiary level paediatric rheumatology. The questionnaire was adapted from one used previously in the Oxford region [4]. It asked patients/parents to rate each standard as either vital, very important, important or useful. They were then asked to go back and rank their top 10 standards. A secondary section asked their views on frequency of attendance and acceptable travel time to a regional tertiary centre.

Results: 33 questionnaires were returned in the time frame of the study. Five did not rank a top 10 and three miss-ranked their response. Standard 4 was ranked as number 1 by 8/33 responders. 23 of the 43 standards were ranked vital more than any other category, 40% of responders felt they should travel for longer than 1 h and 37.5% felt they should be seen at least three times per year in a tertiary centre.

Conclusion: Children and parents feel that the majority of standards are vital and should be part of the standard care they receive. They ranked the use and regulation of drugs most highly, but skill in recognizing the diagnosis, as well as early referral to specialist care, was also prioritized. A minimum of annual review in a tertiary setting was vital, however, the majority want to be seen more often, with this specialist care close at hand.

Disclosure statement: The authors have declared no conflict of interest.

Table 1

<table>
<thead>
<tr>
<th>Standard</th>
<th>The following standards were ranked vital most frequently</th>
<th>% Vital</th>
<th>% In top 10</th>
</tr>
</thead>
<tbody>
<tr>
<td>22</td>
<td>Drugs used for the treatment of JIA will be prescribed and monitored in accordance with BSPAR/NICE guidelines</td>
<td>84.8</td>
<td>34.6</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>Health care practitioners should refer all children with suspected JIA to a paediatric rheumatology team within six weeks of symptom onset</td>
<td>69.7</td>
<td>46.1</td>
</tr>
<tr>
<td>11</td>
<td>All children and young people with JIA should be reviewed at least annually by a designated regional paediatric service</td>
<td>66.7</td>
<td>23.1</td>
</tr>
<tr>
<td>23</td>
<td>Children and parents should be fully informed about the benefits and risks of taking both licensed and unlicensed drugs</td>
<td>66.7</td>
<td>34.6</td>
</tr>
<tr>
<td>8</td>
<td>An early diagnosis and treatment with JIA should have a full assessment of their disease, health, psycho-social and pain management and educational needs</td>
<td>66.7</td>
<td>50.0</td>
</tr>
<tr>
<td>27</td>
<td>Children should be screened by an ophthalmologist with training and experience in paediatric uveitis and be part of a regional network</td>
<td>63.6</td>
<td>42.3</td>
</tr>
<tr>
<td>7</td>
<td>Members of the paediatric rheumatology team will have appropriate training and experience as defined by professional bodies</td>
<td>63.6</td>
<td>26.9</td>
</tr>
<tr>
<td>18</td>
<td>Those children with JIA and active disease should have regular specialist review</td>
<td>60.6</td>
<td>19.2</td>
</tr>
<tr>
<td>29</td>
<td>Specialist surgery should be performed by appropriately trained surgeons with experience in the management of JIA</td>
<td>60.6</td>
<td>23.1</td>
</tr>
<tr>
<td>4</td>
<td>All health care professionals should be able to contact a child with JIA who should acquire the skills to recognize the condition</td>
<td>57.6</td>
<td>46.1</td>
</tr>
</tbody>
</table>

Table 2 Demonstrates the standards ranked useful most times

<table>
<thead>
<tr>
<th>Standard</th>
<th>Standards ranked useful the most times</th>
<th>% Useful</th>
</tr>
</thead>
<tbody>
<tr>
<td>35</td>
<td>The paediatric rheumatology team should encourage and facilitate age appropriate participation in interests, e.g. sports</td>
<td>24.2</td>
</tr>
<tr>
<td>33</td>
<td>Children with JIA should be given skills to disclose their arthritis to others, should they choose to</td>
<td>24.2</td>
</tr>
<tr>
<td>32</td>
<td>Children with JIA should be provided with safe and positive opportunities to meet others with the same condition</td>
<td>21.2</td>
</tr>
</tbody>
</table>

References
3. Bale P, Armon K. Service provision for children with juvenile idiopathic arthritis (JIA) in the east of England (EoE); a comparison with national centres. Abstract accepted to RCPCH conference May 2012.
4. Wilkinson N. Questionnaire developed for Oxford Deanery parental survey 2011, provided to Kate Armon for use in this survey.

10. SURVEY OF STRATEGIES USED TO MANAGE RESISTANT JIA IN THE UK

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Background: In the Yorkshire and Humberside Paediatric Rheumatology Network every child with JIA requiring non-tariff medications that are not NICE approved, requires an individual funding application to the relevant Primary Care Trust. With commissioners for our network we are trying to develop agreed guidelines to simplify this process. Towards this a description of the current practice of all UK centres would be helpful.

Objective: We aimed to collect and describe this information for this process and to share with all UK centres. We have focused on the situations where the use of non-tariff treatments are sought therefore have concentrated on JIA that is resistant to the first-line treatments that we use within our network.

Methods: An online survey was designed, posing questions around specific scenarios for patients with different subtypes of JIA and uveitis. Participants were invited to rank treatment options in the order that they would use them, following failure of described therapies. All consultants on the BSPAR mailing list were invited to take part. A follow-up targeted email will be sent to consultants in the centres not represented with additional data anticipated.

Results: 13 consultants covering 10 regions completed the survey. Some results of note are:

(i) In RF-positive polyarthritis, RF-negative polyarthritis and PsA the first choice of drug following failure of the first anti-TNF agent was a second anti-TNF in ~80% of respondents and for 92%, 100% and 100% of respondents respectively within the first three choices.
(ii) In RF-positive patients rituximab (8/13 respondents) and abatacept (8/13 respondents) were in the top three choices.
(iii) In RF-negative patients choices were more variable between abatacept (7/13), tocilizumab (6/13) and DMARDs (5/13) for the top three choices.
(iv) In systemic JIA, patients with arthritis-predominant ongoing disease process were more likely to be prescribed anti-TNF agents or tocilizumab following MTX failure, whereas there was a 2:1 split between tocilizumab and anakinra for those with ongoing systemic symptoms.
(v) In JIA-associated uveitis adalimumab was most often prescribed after MTX, whereas in idiopathic uveitis MMF or adalimumab was used with almost equal frequency.

More detailed results can be provided.

Conclusion: We hope that this description of current UK practice will help inform discussions with commissioners and possibly act as a starting point for future UK guidelines.

Disclosure statement: The authors have declared no conflict of interest.

11. BASELINE CO-MORBIDITIES IN PATIENTS WITH JIA STARTING ETANERCEPT OR METHOTREXATE: RESULTS FROM THE BSPAR ETANERCEPT REGISTER

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Background and objectives: Understanding the short- and long-term safety of etanercept (ETN) use in children with JIA is of paramount importance to patients, families and the rheumatology community. However, when interpreting adverse events, it is important to consider the burden of co-morbidity among children starting new therapies for JIA, which may increase the risk of certain adverse events. Therefore, the aim of this analysis was to describe and quantify the baseline medical and health conditions among children newly starting either ETN or MTX for JIA.
Methods: The national BSPAR ETN register was established in 2004 to monitor the safety and effectiveness of ETN in children with JIA. A comparison cohort of children with JIA who are biologic naive starting MTX or NOZA was recruited. To 31 December 2011, 876 patients were enrolled (ETN 679, MTX 197). Rheumatology centres were asked to provide a list of all co-existing medical conditions and whether they were active at the time of drug start. These co-morbidities were medDRA coded and grouped into one of 16 categories. Comparisons between the cohorts were made using non-parametric descriptive statistics.

Results: ETN patients were older with a mean age of 11 years compared with 8 years in the MTX cohort. They also had longer median disease duration (ETN 4 years, MTX 1 year). Within both cohorts, 44% patients had at least one co-morbidity (ETN 46%, MTX 37%), with a higher proportion of children starting ETN having multiple co-morbidities (ETN 22%, MTX 11%). The most frequent co-morbidities presented were atopic conditions, chronic anterior uveitis and congenital/genetic/developmental conditions. There were some differences in co-morbidities between the groups, for example ETN-treated patients were more likely to have osteoporosis or osteoarthritis (ETN 6%, MTX 0%) and growth/developmental abnormalities (ETN 9%, MTX 3%), which may be explained by the longer disease duration in this cohort. ETN patients also presented with more eye (ETN 6%, MTX 1%) and renal conditions (ETN 6%, MTX 2%).

Conclusion: Co-morbidity is common among children with severe JIA, with a similar distribution between treatment groups. ETN-treated patients had more co-morbidities overall and so should be considered when interpreting long-term outcomes amongst these patients.

Disclosure statement: The authors have declared no conflict of interest.

12. USE OF NON-ETANERCEPT BIOLOGICS IN CHILDREN WITH JIA: RESULTS FROM THE BIOLOGICS FOR CHILDREN WITH RHEUMATIC DISEASES STUDY

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Background: The introduction of biologic therapies has revolutionised the management of JIA. Previously, the only choice of licensed therapy in the UK was etanercept (ETN), with adalimumab (age ≥4 years), abatacept (age ≥6 years) and tocilizumab (systemic arthritis) more recently included.

Objective: The objective of this analysis was to describe non-ETN biologic pattern of use in children with JIA.

Methods: The Biologics for Children with Rheumatic Diseases (BOCR) study is an ongoing prospective observational cohort study that has been collecting detailed information on children <18 years of age starting a non-ETN biologic therapy for JIA since 2010. Detailed demographic and disease information, including past biologic therapies, has been collected at baseline. Using non-parametric descriptive statistics the use of non-ETN therapy as a first-line or subsequent biologic therapy was compared, including patterns of prescription, use under licensed indications, ILAR subtypes and disease activity/ severity.

Results: To 08/16/2012, 148 children across the UK were recruited: median age 10 years, 64% female. The most common ILAR subtypes were systemic arthritis (30%) and RF-negative polyarthritis (26%). Seventy-one patients (48%) were starting a non-ETN biologic as first-line biologic therapy, of which 35 (49%) were prescribed off-licence, largely accounted for by infliximab and anakinra. First-line biologic users were a younger cohort compared with subsequent biologic users (P = 0.0077). All patients on anakinra had systemic arthritis, whereas only 73% of tocilizumab patients used it for this licensed indication. Forty-five per cent of first-line users vs 25% of subsequent users had a history of chronic anterior uveitis (P = 0.031). Of those registered at the point of starting a second-line biologic, 78% had received prior ETN. The majority of all patients receiving previous biologic treatment had received only one prior biologic (71%) although 18 children had received two prior biologics, 3 children had received three therapies (RF negative) and 2 children (RF negative) had received five previous biologics. Subsequent biologic users had a higher limited joint count (P = 0.0026).

Conclusions: In the UK, many children not receiving non-ETN biologics, although almost half of these are being prescribed off-licence. Continual follow-up in children with JIA will help to address questions of the best choice of biologic therapy, as both first-line and subsequent treatment, as well as determine the safety of these drugs in children, for which limited clinical experience exists.

Disclosure statement: The authors have declared no conflict of interest.

13. SAFETY AND EFFICACY OF US-GUIDED HIP INJECTIONS IN JIA

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Aim: To assess the safety and efficacy of US-guided injections in hip joints of children with JIA.

Methods: All children who received US-guided hip injections from March 2009 to June 2012 were included in the assessment. Safety and efficacy of the injections were assessed clinically.

Results: 23 children received US-guided hip injections under general anaesthesia. The procedure was done by a single paediatric rheumatologist in all cases. Triamcinolone hexacetonide was used to inject the hip joint for weight: ≤10 kg, 10 mg; 10–20 kg, 20 mg; 20–40 kg, 30 mg; and >40 kg, 40 mg. 14 girls and 9 boys formed the study sample. Median age was 13.7 years (range 1.3–18 years), Radiological evidence of active arthritis was demonstrated prior to injections by US in 65%, MRI in 22% and both US and MRI in 13%. A total of 44 hips were injected during this period. 31 hips were injected for the first time, 9 hips were injected for a second time and 4 hips needed a third episode of injection. Efficacy was assessed on a follow-up visit at least 3 months after the joint injection. Duration of effect was calculated up to the last follow-up visit. Efficacy was 71% for the first episode and 89% for the second episode. Duration of response was for a mean period of 7 months (range 1–30 months). Imaging on follow-up was done if there was clinical suspicion of disease activity. Of 31 hips injected, on follow-up, activity was suspected in 18 hips (58%), of which normal imaging was found in 10 hips (55%), evidence of ongoing disease activity in the form of joint effusion/synovial thickening was found in 9 (50%) and disease-related damage in 1 (5%). Of the 9 hips injected for a second time, the effect lasted for a mean period of 8.9 months (range 4–15 months). There was one episode of cutaneous bruising (2.3%) and no evidence of avascular necrosis was found in any hip on imaging at the last follow-up visit.

Conclusion: US-guided hip injections carried out by a paediatric rheumatologist are safe and efficacious. They could be a potential alternative to fluoroscopically guided joint injections, avoiding radiation in children.

Disclosure statement: The authors have declared no conflict of interest.

14. PREDICTORS OF ACCESS TO CARE IN JSLE: EVIDENCE FROM THE UK JSLE COHORT STUDY

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Background: Delays in access to specialist care for children with paediatric rheumatic disease are likely to be associated with adverse outcomes [1, 2]. Factors influencing access to care for juvenile onset SLE (JSLE) remain mainly unknown.

Objectives: We aimed to investigate factors that may influence the interval between symptom onset and JSLE diagnosis within a UK cohort.

Methods: Data from 257 patients recruited to the UK JSLE Cohort study [3] were analysed. Potential predictors of access to care explored included: socioeconomic status, ethnicity, gender, age at presentation, presenting features, ACR criteria, origin of referral to paediatric rheumatologist, distance from nearest tertiary paediatric rheumatology service and family history of autoimmune disease. Correlation tests were employed to identify variables associated with a log of time between symptom onset and diagnosis (P < 0.1). Linear regression was then used to identify independent predictors of access to care.

Results: 257 children from across the UK with JSLE were included in the analysis (216 female, 41 male, ratio 5:3:1). Median time from symptom onset to diagnosis was 0.37 years [range 0–14.07 years, interquartile range (IQR) 0.17–1.38]. Median distance to a tertiary paediatric rheumatologist centre was 20.5 miles (IQR 9.2–41.5). The
JSLE cohort population Index of Multiple Deprivation scores were found to be higher than for the English population as a whole and not significantly associated with time to diagnosis ($P = 0.12$). Variables correlating with time to diagnosis included: being British (odds ratio 0.29, coefficient $-0.268$, $P = 0.007$), Caribbean or African ($-0.159$, $P = 0.011$), presenting to a specific tertiary referral centre ($0.139$, $P = 0.026$), being referred by a paediatrician ($-0.130$, $P = 0.037$) and having nephritis ($-0.138$, $P = 0.027$), immunological disorder ($-0.125$, $P = 0.039$) or haemato-logical disorder ($-0.164$, $P = 0.09$) at presentation. A linear regression model identified being Caribbean/African ($P = 0.006$) or Asian ($P = 0.045$) ethnicity, being referred by a paediatrician ($P = 0.049$) or nephritis ($P = 0.045$) at presentation as independent predictors of shorter time to diagnosis.

Conclusion: Within this cohort, ethnic origin and the initial source of referral were strong predictors of interval in establishing diagnosis of JSLE. LN at presentation is a significant independent predictor of shorter time to diagnosis. Gender, age at presentation, ACR score, referral were strong predictors of interval in establishing diagnosis of JSLE.LN at presentation is a significant independent predictor of shorter time to diagnosis. Gender, age at presentation, ACR score.

Disclosure statement: The authors have declared no conflict of interest.

15. GENDER DIFFERENCES IN THE ACR CLASSIFICATION CRITERIA PATIENTS DIAGNOSED WITH JUVENILE ONSET SLE YOUNGER THAN 10 YEARS

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Background: In 15–20% of SLE patients, disease onset occurs during childhood/adolescence, termed juvenile onset SLE (JSLE). The main aspects of JSLE resemble adult onset SLE and the ACR criteria are thought to be equally accurate in both populations. Children have a more acute, aggressive disease and greater therapeutic burden. Female preponderance is less pronounced in JSLE: male 5:1. The UK JSLE Cohort Study, a large national inception cohort, offers the opportunity to effectively describe JSLE. Previously, data from this study highlighted differences in the clinical presentation between the sexes with arthritis commoner in females and discoid rashes commoner in males. In pre-pubertal patients, younger than 10, sex hormones are less likely to influence the pathogenesis of SLE.

Objectives: To investigate the influence gender may have on the disease features in JSLE patients less than 10 years old.

Methods: Patients within the UK JSLE cohort study aged <10 at diagnosis, with four or more ACR criteria were included. ACR criteria were used to compare disease features at diagnosis and latest follow-up. Ethical approval was in place. Statistical analyses used SPSS with results reported as median and interquartile range (IQR).

Results: Within the cohort of 240 patients, 23% ($n = 56$) were aged 15 years or less at the time of diagnosis ($P = 0.001$) presenting to a specific tertiary referral centre ($0.139$, $P = 0.026$), being referred by a paediatrician ($0.130$, $P = 0.037$) or having nephritis ($0.138$, $P = 0.027$), immunological disorder ($0.125$, $P = 0.039$) or haematological disorder ($0.164$, $P = 0.09$) at presentation. A linear regression model identified being Caribbean/African ($P = 0.006$) or Asian ($P = 0.045$) ethnicity, being referred by a paediatrician ($0.049$) or nephritis ($0.045$) at presentation as independent predictors of shorter time to diagnosis.

Conclusion: Within this cohort, ethnic origin and the initial source of referral were strong predictors of interval in establishing diagnosis of JSLE. LN at presentation is a significant independent predictor of shorter time to diagnosis. Gender, age at presentation, ACR score, referral were strong predictors of interval in establishing diagnosis of JSLE. LN at presentation is a significant independent predictor of shorter time to diagnosis.

Disclosure statement: The authors have declared no conflict of interest.

16. PREVALENCE AND SIGNIFICANCE OF THROMBOCYTOPENIA IN JUVENILE ONSET SLE AT PRESENTATION AND 1 YEAR FOLLOW-UP

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Background: Thrombocytopenia (TCP) is a common haematological manifestation in both adult onset SLE and juvenile onset SLE (JSLE) and has been associated with increased global disease activity, specific organ involvement, deranged laboratory markers and increased damage accrual and mortality. However, studies in JSLE are few and small scale.

Objective: To establish (i) the prevalence of TCP in JSLE patients at time of diagnosis (T0) and two-twelve-month follow-up (T12) and (ii) associations of TCP with disease activity, laboratory markers and disease-related damage.

Methods: Patients recruited between 2004 and 2011 to the UK JSLE inception cohort study and registry were included, and at least four ACR criteria for diagnosis at T0. Demographics, disease activity, laboratory results and damage accrual data were analysed at T0 and T12 (range 9–15 months). TCP was defined as $<150000$ platelets/mm$^3$, as per the British Isles Lupus Assessment Group (BILAG)-2004 definitions. Results are presented as median (range).

Results: 155 patients [83% female; age at diagnosis: 12.6 (1.8–17.9) years] were analysed. 27 (17%) had TCP at T0 (98 x 10$^9$/l, 6–147) and 6 (6%) at T12 (94 x 10$^9$/l, 31–142). Two patients were thrombocytopenic at both T0 and T12. Five patients at T0 and one at T12 had platelets $<50000$/mm$^3$. Thrombocytopenic patients had statistically significantly higher total BILAG-2004 score at both T0 (BILAG-2004: 16.9 (2–53) vs $12.7$ (0–53); $P = 0.002$) and T12 [5.5 (2–15) vs $3.3$ (0;16–$P = 0.04$)]. TCP was statistically significantly associated with active haematological involvement at T0 (BILAG-2004 category A or B, $n = 14/27$) compared with patients without TCP ($n = 35/128$, $P = 0.03$). Of the laboratory markers of disease activity measured, only C3 in TCP and non-TCP patients, respectively, statistically significantly differed at T0 [0.6 mg/l (0.2–1.3) vs $0.8$ mg/l (0.2–2.0); $P = 0.006$]. Disease-related damage accrual, measured by the SLE International Collaborative Clinic (SLICC) score, showed no statistically significant difference in those with or without TCP [SLICC score $0.3$ (0–4) vs $0.14$ (0–4); $P = 0.90$] after 3.8 (0.9–13.2) years of follow-up.

Conclusions: 17% of JSLE patients in this cohort had TCP at T0, though most resolved by T12. Thrombocytopenic patients at T0 had lower C3 levels and significantly higher disease activity scores at both diagnosis and follow-up, but had not accrued more disease-related damage to date. Ongoing follow-up is taking place.

Disclosure statement: The authors have declared no conflict of interest.

17. PROFILING ANTIBODIES TO SERUM PROTEASES IN PATIENTS WITH JUVENILE ONSET SYSTEMIC LUPUS ERYTHEMATOSUS

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Background: We have recently shown that adult patients with SLE have increased anti-Fxa IgG antibodies as compared with healthy and inflammatory arthritis disease controls. To our knowledge, however, the presence of these and other antibodies directed against serine proteases (SPs) in juvenile onset SLE (JSLE) has not been profiled. It is generally accepted that clinical manifestations of JSLE are different from adult onset SLE in their severity, accumulation of disease-related damage and renal manifestations with less clear differences in laboratory markers of disease. Therefore, we undertook an analysis of the prevalence of antibodies to Factor Xa and other SPs in patients with JSLE to see whether these antibodies were elevated in this disease group compared with adult patients with SLE and JIA disease controls.

Methods: Serum was obtained from patients of University College Hospital with JSLE ($n = 35$), JIA ($n = 14$), adult SLE ($n = 106$) and from 39 adult healthy controls. Serum was tested for the presence of IgG directed against thrombin (Thr), Factor Viila (FVIIa) and Factor Xa (Fxa) by ELISA. Results were expressed as a percentage of binding compared with a positive control where the positive value was defined as being $>250$, above the mean of adult healthy controls.

Results: Only IgG anti-Fxa antibodies were found more frequently in adult patients with SLE ($n = 52/106$, 49.1%) compared with JSLE ($n = 5/33$, 15.2%, $P = 0.001$). There was no statistically significant difference in anti-FVIIa or anti-Thr between these two groups. In contrast, only anti-Thr levels were elevated ($n = 17/35$, 48.6%) in patients with JSLE compared with JIA ($n = 1/14$, 7.1%, $P < 0.01$) and
there was no difference between anti-FXa and anti-FVIIa levels, which were low for both groups.

**Conclusion:** Of the anti-SP antibodies tested, levels of anti-FXa antibodies were useful in distinguishing between patients with adult and juvenile onset SLE whilst levels of anti-Thr antibodies distinguished JSLE from JIA controls but not from adult onset SLE. Further studies are now underway to correlate these findings with the clinical and serological phenotype of juvenile and adult onset disease.

**Disclosure statement:** The authors have declared no conflict of interest.

### 18. VITAMIN D SUPPLEMENTS IN PATIENTS WITH JUVENILE SLE

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**Background:** Vitamin D deficiency is rising in children. Vitamin D is an essential nutrient. Low levels of vitamin D are associated with low bone mass density, secondary hyperparathyroidism and subsequent fracture risk. The Department of Health advise supplementation for those <4 years of age, >65 years of age, pregnant, lactating or at risk of low sun exposure. Vitamin D deficiency is common in children. The risk of vitamin D deficiency is increased when patients are dark skinned, photosensitive and adhere to sun protection advice, such as in juvenile SLE (JSLE).

**Objective:** To review evidence available on JSLE, bone density and vitamin D to understand whether patients would benefit from vitamin D supplements.

**Method:** Electronic search of PubMed and the Cochrane Library up to August 2012.

**Results:** Eight citations found, four cross-sectional, three cohort studies and one review.

**Conclusion:** Vitamin D deficiency in juvenile systemic lupus erythematosus is associated with low bone mineral density. Studies suggest that reduced bone mineral density is related to cumulative steroid dosage, and levels of vitamin D are associated with increased disease duration and severity in JSLE. Vitamin D deficiency is commonly found at diagnosis and may need higher than standard dosage supplementation. Further research is required for optimal level of supplementation in varying disease status.

**Disclosure statement:** The authors have declared no conflict of interest.

<table>
<thead>
<tr>
<th>Author</th>
<th>Patient</th>
<th>Study type</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Guenka A et al. 2011 [1], Belgium</td>
<td>53 JSLE patients, DXA scanning performed</td>
<td>Cross-sectional study</td>
<td>No direct correlation was observed between the analysed parameters and densitometry findings according to the Z-score.</td>
</tr>
<tr>
<td>Compeyrot-Lacassagne S, 2007 [2], Canada</td>
<td>64 JSLE patients, DXA scanning performed</td>
<td>Cross-sectional study</td>
<td>Lumbar spine osteopenia was seen in 37.5% and osteoporosis in 20.3%. Decreased hip BMD was present in 18.8%. Osteopenia and osteoporosis are more common in JSLE and are associated more closely with longer disease duration than cumulative steroid dose.</td>
</tr>
<tr>
<td>Wright TB et al., 2009 [3], USA</td>
<td>38 JSLE patients, 207 healthy controls</td>
<td>Cross-sectional study</td>
<td>Severe vitamin D deficiency (25 (OH)D &lt;10 ng/ml) was observed in a significantly higher proportion of subjects with SLE (P = 0.001).</td>
</tr>
<tr>
<td>Casella CB et al., 2012 [4], Brazil</td>
<td>57 JSLE patients, 37 healthy controls</td>
<td>Cross-sectional study</td>
<td>25 (OH)D levels were similar in patients and controls (21.44 vs 22.54 ng/ml, P = 0.519), regardless of supplementation (65% of patients and none in controls). Higher doses of vitamin D may be needed (&gt;400 IU).</td>
</tr>
</tbody>
</table>

### 19. EFFECTIVENESS OF INTRAVENOUS CYCLOPHOSPHAMIDE IN SEVERE OR REFRACTORY JUVENILE DERMATOMYOSITIS

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**Background:** JDM is a rare autoimmune vasculopathy affecting primarily the muscle and skin. Early and aggressive treatment improves outcome and prevents complications. Cyclophosphamide has been used as a second-line agent in the treatment of severe or refractory JDM. The published literature on the effectiveness of cyclophosphamide in JDM is limited to small case series and case reports.

**Objective:** To describe the response to cyclophosphamide in patients with severe or refractory JDM.

**Methods:** 56 patients treated with cyclophosphamide between 2000 and 2011 were identified in the JDM National (UK and Ireland) Cohort Biomarker Study and Repository for Idiopathic Inflammatory Myopathies. 8 patients were excluded due to incomplete data or too short follow-up. For the 48 patients included, demographics, myositis core outcome variables, skin measures, laboratory measures, steroid dose and other treatments were recorded at baseline, and at 6-, 12-, 18-, 24-month and last follow-up after commencement of the drug.

**Results:** Indications for starting cyclophosphamide were ulcerative or severe skin disease, profound muscle weakness, lung disease, gastrointestinal vasculopathy or refractory disease. All patients starting with muscle weakness (n = 44) significantly improved at 12 months, and the gains were maintained at follow-up. Physician VAS was available for 32 patients and these all improved by 12 and 24 months, and for 31 remained stable at follow-up. At the last follow-up 26/48 (56%) had no rash, 32/48 (67%) had normal nailfolds, 37/45 (82%) had no Gottron’s and calcinosis improved in 9/14 (64%).

**Conclusions:** This study, the largest to date, demonstrated significant improvement in both muscle and skin domains in patients with JDM treated with i.v. cyclophosphamide. Cyclophosphamide appears to be effective in the treatment of severe or refractory JDM.

**Disclosure statement:** The authors have declared no conflict of interest.

### 20. CHRONIC RECURRENT MULTIFOCAL OSTEOSEPTITIS: A CASE SERIES ANALYSIS

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**Background:** Chronic recurrent multifocal osteomyelitis (CRMO) is an inflammatory bone disease occurring primarily in children and adolescents. Currently, due to the lack of awareness of the disease and the absence of specific diagnostic tests, children undergo many investigations and treatments before a diagnosis is finally reached. In our description of the findings from a large cohort of children with CRMO we outline common features enabling earlier recognition and diagnosis.

**Objective:** The aim of the study was to describe the clinical and radiological features, the response to different therapies and the outcomes in a cohort of patients with CRMO. This was done by completing a retrospective case-note and imaging review of children undergoing assessment and treatment at Bristol Children’s Hospital.

**References**


22. DIFFERENTIATING BETWEEN JIA AND SEPTIC ARTHRITIS: IDENTIFICATION OF IMPORTANT FEATURES IN HISTORY, EXAMINATION AND LABORATORY INVESTIGATION

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Background: The presentation of a single swollen knee joint in a child can pose a diagnostic dilemma that requires a timely and correct diagnosis to avoid complications and unnecessary investigations. This study concentrates on two of these differentials: septic arthritis (SA), which needs emergency management, and JIA, where aggressive management such as synovial biopsy can be avoided. The aim is to identify clinical findings and routine laboratory investigations that could aid in the early and appropriate diagnosis, with referral to the correct specialist.

Methods and materials: A retrospective review was performed using the case-notes of 44 children presenting with a single swollen knee joint between 2000 and 2010 with a definitive diagnosis of either SA or JIA. Comparisons of features potentially predictive of the final diagnosis were compared using univariate analysis and multivariate analysis.

Results: Three of the five factors identified as significantly suggesting SA over JIA are identified via history and examination; these are fever, being unwell and duration of onset. Three routinely investigated factors, WBC, CRP and ESR, were also identified as significantly different though logistic regression. Two sample t-test and Fisher’s exact test also identified a raised neutrophil count, non-weight bearing, lack of signs of chronic joint damage and a hot joint as important factors. Stepwise regression found a combination of CRP > 7 mmol/l, onset -4 days and the child being generally unwell as significantly indicative of SA.

Conclusion: Routinely performed investigations and a detailed history of onset are useful in differentiating between SA and JIA. Identification of these features will potentially allow for a more timely and accurate diagnosis of a single swollen knee.

Disclosure statement: The authors have declared no conflict of interest.

23. ASSESSMENT OF MUSCuloskeletal ABNORMALITIES IN CHILDREN WITH MUCopolYSaccharidoses USING A SIMPLE MUSCuloskeletal EXAMINATION (Paediatric Gait, Arms, Legs and Spine)

Ethan Sen1, Mercedes Chan2, Elizabeth Hardy3, Tim Rapley4, Pauline Herrmann5, James E. Wraith5 and Helen Foster1

Objective: To establish the safety and efficacy of pamidronate in the treatment of children with CRMO.

Methods: A total of 18 patients were treated with i.v. pamidronate (1 mg/kg/day, every 4 weeks) for a median duration of 12 months (4–24 months). Of these, 11 completed the dosage regime. Of these 11 completing treatment, an average of four doses. If symptoms persisted, they received a further infusion (providing MRI results were favourable). Of the 18 patients, 7 (64%) had a reduction in high signal on MRI and became clinically asymptomatic. In the remaining four (36%) bone pain persisted and MRI was unchanged. Seven patients were still undergoing pamidronate therapy. Other treatments included DMARDs (21%) and steroids (13%). These results were similar to those found in published reviews.

Conclusion: This cohort is one of the largest series in the literature. It is important to increase awareness of CRMO as a diagnostic differential when a child presents with insidious onset bone pain. MRI STIR provides important evidence in the diagnosis of CRMO and may prevent the use of unnecessary radiation and treatment by aiding earlier diagnosis. A central database would facilitate a greater understanding of the diagnostic criteria and treatment options.

Disclosure statement: The authors have declared no conflict of interest.
### Table 1: Musculoskeletal abnormalities detected by pGALS (figures stated as percentage abnormal)

<table>
<thead>
<tr>
<th>Manoeuvre tested</th>
<th>pGALS instruction</th>
<th>All MPS (n = 15)</th>
<th>MPS I (n = 8)</th>
<th>MPS I-HS (n = 2)</th>
<th>MPS II (n = 4)</th>
<th>Mannosidosis (n = 1)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Shoulder flexion</td>
<td>Reach arms up</td>
<td>88.9</td>
<td>91.7</td>
<td>100</td>
<td>100</td>
<td>0</td>
</tr>
<tr>
<td>Shoulder abduction, external rotation</td>
<td>Hands behind neck</td>
<td>88.9</td>
<td>83.3</td>
<td>100</td>
<td>100</td>
<td>66.7</td>
</tr>
<tr>
<td>Elbow flexion</td>
<td>Hands out in front</td>
<td>80</td>
<td>62.5</td>
<td>100</td>
<td>100</td>
<td>100</td>
</tr>
<tr>
<td>Wrist flexion</td>
<td>Hands and wrists together</td>
<td>77.8</td>
<td>66.7</td>
<td>83.3</td>
<td>100</td>
<td>66.7</td>
</tr>
<tr>
<td>TMJ excursion</td>
<td>Open mouth wide and try to put three fingers inside</td>
<td>75.6</td>
<td>70.8</td>
<td>83.3</td>
<td>33.3</td>
<td></td>
</tr>
<tr>
<td>MCP, DIP, PIP flexion</td>
<td>Hands and wrists together 12</td>
<td>68.9</td>
<td>50</td>
<td>100</td>
<td>91.7</td>
<td>66.7</td>
</tr>
<tr>
<td>Forward flexion of spine</td>
<td>Bend forwards. Observe curvature of spine from all sides</td>
<td>67.5</td>
<td>66.7</td>
<td>100</td>
<td>66.7</td>
<td></td>
</tr>
<tr>
<td>Spinal deformity</td>
<td></td>
<td>66.7</td>
<td>70.8</td>
<td>100</td>
<td>55.6</td>
<td>50</td>
</tr>
<tr>
<td>Wrist extension</td>
<td>Hands back to back</td>
<td>65.9</td>
<td>50</td>
<td>100</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>Gait</td>
<td>Observe patient walking</td>
<td>62.2</td>
<td>62.5</td>
<td>N/D</td>
<td>70</td>
<td>33.3</td>
</tr>
<tr>
<td>MCP/DIP/PIP extension</td>
<td>Hands out in front</td>
<td>57.8</td>
<td>33.3</td>
<td>100</td>
<td>100</td>
<td>0</td>
</tr>
<tr>
<td>Neck extension</td>
<td>Lock up to the ceiling</td>
<td>57.8</td>
<td>45.8</td>
<td>100</td>
<td>75</td>
<td>0</td>
</tr>
<tr>
<td>Elbow extension</td>
<td>Reach up</td>
<td>57.1</td>
<td>33.3</td>
<td>100</td>
<td>81.8</td>
<td>50</td>
</tr>
<tr>
<td>Cervical spine lateral flexion</td>
<td>Touch ear to shoulder</td>
<td>56.8</td>
<td>33.3</td>
<td>100</td>
<td>33.3</td>
<td></td>
</tr>
<tr>
<td>Ankle dorsiflexion</td>
<td>Walk on heels</td>
<td>55.6</td>
<td>45.8</td>
<td>N/D</td>
<td>77.8</td>
<td>66.7</td>
</tr>
<tr>
<td>Forearm supination</td>
<td>Turn hand over</td>
<td>54.5</td>
<td>35.3</td>
<td>50</td>
<td>81.9</td>
<td>100</td>
</tr>
<tr>
<td>Opposition of thumb and third to fifth fingers</td>
<td>Touch tips of fingers with thumb</td>
<td>44.4</td>
<td>20.8</td>
<td>100</td>
<td>66.7</td>
<td>33.3</td>
</tr>
<tr>
<td>MCP/DIP/PIP flexion</td>
<td>Make a fist</td>
<td>37.8</td>
<td>4.2</td>
<td>100</td>
<td>83.3</td>
<td>0</td>
</tr>
<tr>
<td>Knee extension</td>
<td>Bring ankle up to bottom</td>
<td>28.9</td>
<td>26.1</td>
<td>N/D</td>
<td>33.3</td>
<td>33.3</td>
</tr>
<tr>
<td>Knee flexion</td>
<td></td>
<td>23.1</td>
<td>8.3</td>
<td>N/D</td>
<td>58.3</td>
<td>0</td>
</tr>
<tr>
<td>Opposition of thumb and index finger</td>
<td>Touch tip of finger with thumb</td>
<td>22.2</td>
<td>0</td>
<td>83.3</td>
<td>41.7</td>
<td>0</td>
</tr>
<tr>
<td>Elbow flexion</td>
<td>Put hands behind neck</td>
<td>22.2</td>
<td>4.2</td>
<td>50</td>
<td>41.7</td>
<td>33.3</td>
</tr>
<tr>
<td>Elbow flexion</td>
<td>Put hands and wrists together</td>
<td>13.6</td>
<td>4.2</td>
<td>50</td>
<td>18.2</td>
<td>0</td>
</tr>
<tr>
<td>Ankle plantar flexion</td>
<td>Walk on tip-toes</td>
<td>8.3</td>
<td>4.2</td>
<td>N/D</td>
<td>22.2</td>
<td>0</td>
</tr>
</tbody>
</table>

N/D: not demonstrated; TMJ: temporomandibular joint; MCP: metacarpophalangeal; DIP: distal interphalangeal; PIP: proximal interphalangeal.

and inter-observer $\kappa = 0.62$ (range 0.51–0.77). Hip manœuvres within pGALS were not clearly demonstrated in the videos.

**Conclusion:** In this observational study, pGALS identifies MSK abnormalities in children with MPS. Restricted joint movement (and especially upper limb) was a consistent finding. We acknowledge that further work is needed to include pGALS assessment of the hip and also to test pGALS in an additional population of children with MPS; notably in younger children with MPS I-HS as this subtype often has MSK abnormalities as the only feature. The use of pGALS and awareness of patterns of joint involvement may be a useful adjunct to facilitate earlier recognition of these rare conditions and facilitate access to specialist care.

**Disclosure statement:** The authors have declared no conflict of interest.

### 24. HOW GOOD IS THE QUALITY OF REFERRAL LETTERS TO PEDIATRIC RHEUMATOLOGY FROM THE ACCIDENT AND EMERGENCY DEPARTMENT?

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**Background:** Referral letters (RLs) to Paediatric Rheumatology (PR) often lack key information to facilitate correct triage. Strategies to enhance the quality of referrals include providing referral guidelines and targeted PR teaching.

**Objective:** The aim of this study was to assess the quality of RL sent from the Accident and Emergency (A&E) department to PR, and any change in quality after PR teaching.

**Methods:** An audit was conducted of the quality of A&E RL to PR written between 2007 and 2012. The quality of RL was graded and comparison of RL written before or after targeted PR teaching was made. Information from the Canadian Rheumatology Association was used to identify the requirements of a RL to rheumatology. Each RL was assigned a grade from 0 (weakest) to 3 (excellent).

**Results:** 74 RLs were reviewed from A&E RL to Paediatric Rheumatology. 38 RLs were recorded as either grade 2 or 3, with the A&E trainees sending the most grade 3 letters (5/11). The number of high-quality RLs was unchanged after teaching. The number of grade 0 letters fell from 9 to 3 after teaching.

**Conclusion:** The overall quality of RLs was better than previously published. A pre-referral checklist could prove beneficial. PR teaching improves doctors’ understanding of paediatric musculoskeletal disorders but could be targeted to improve confidence in managing benign conditions, avoiding unnecessary referral, and confidence in diagnosing hip presentations and JIA.

**Disclosure statement:** The authors have declared no conflict of interest.

### 25. ANCA-POSITIVE VASCULITIS ASSOCIATED WITH LEVAMISOLE

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**Background:** Levamisole is an antihelminthic agent with immunomodulating properties that is used to reduce the risk of relapse in children with frequently relapsing steroid-dependent nephrotic syndrome. It has been reported to be associated with circulating autoantibody formation and vasculitis in both adults and children [1-3].

**Case report:** We report the case of an 8-year-old girl who developed ANCA-positive vasculitis following a prolonged course of levamisole. She was diagnosed with steroid-dependent nephrotic syndrome aged 4 and commenced on levamisole in addition to low dose alternate day prednisolone the following year due to frequent relapses. She had been taking levamisole for 42 months when she presented with fever, lethargy, polyarticular arthritis and a blanching erythematous rash. Abdominal US revealed splenomegaly, ESR and CRP were raised and blood film was normal. pANCA positivity persisted at follow-up 2 months later with negative PR3 and PRB antibodies detected. She was treated initially with ibuprofen and cessation of levamisole. Six weeks after stopping levamisole she was readmitted with ongoing arthritis, lethargy, splenomegaly and a vasculitic rash on both thenar eminences. ESR and CRP remained high. She was treated with four doses of i.v. methylprednisolone and a weaning course of oral prednisolone over 6 weeks. Her symptoms resolved and inflammatory markers normalized with this treatment but ANCA positivity persisted at follow-up 2 months later with negative PR3 and a reduced titre of MPO. In summary we report a case of levamisole-associated ANCA-positive vasculitis that occurred after 42 months of drug exposure and responded promptly to treatment with i.v. methylprednisolone. This is in contrast to previous paediatric reports in which symptoms resolved following withdrawal of
levamisole and specific treatment was not required [1–3]. It is important for both paediatric rheumatologists and nephrologists to be aware of this significant potential adverse effect of levamisole, which tends to occur following prolonged exposure and may require treatment for persisting symptoms despite withdrawal of the drug.

Disclosure statement: The authors have declared no conflict of interest.

References

26. PROFOUND MUSCLE WEAKNESS WITH NEUROLOGICAL AND RENAL INVOLVEMENT: JDM OR NOT?
Kishore Warrier1, Ethan Sen1, Mario Abinun2 and Sharmila Jandial2

AND RENAL INVOLVEMENT: JDM OR NOT?

Case report:
A previously well 10-year-old boy presented to local hospital with a 3-week history of vomiting, abdominal pain, allergic rash and worsening lethargy. He had hypertension and proteinuria and was referred to the paediatric nephrology unit. On arrival, he was oedematous with malar rash, Gottron’s papules (elbows and knuckles), dilated nailfold capillaries; and profoundly weak with oedematous and tender muscles. He had normal FBC with ESR 21 mm/h but normal CRP. His creatinine kinase was 18364; lactate dehydrogenase 1188 and ALT 252 U/l; but the rest of the biochemistry was normal. Autoantibody screen revealed weakly positive ANA (20) with negative dsDNA, ENA and subacute myositis antibody screen.

He had significant proteinuria and microscopic haematuria. With provisional diagnosis of JDM, he was started on i.v. methylprednisolone (MP). After the first dose, he developed two generalized convulsions needing intubation and ventilation and transfer to the paediatric intensive care unit. His MRI and magnetic resonance angiography of the brain were normal with no vasculopathy and EEG did not show seizure activity but he was hyponatraemic (sodium 122 mmol/l). MRI of muscles and muscle biopsy were suggestive of JDM. He became anuric (peak creatinine 105 mmol/l) requiring CVVH (Birmingham vasculitis protocol) and is due to start MTX.

Investigations to find overlap with other CTDs and underlying infection, malignancy and metabolic disorders were negative. Within a week, he was off respiratory support, eating normally and walking unsupported although weak proximally (CMAS 11).

Discussion: JDM is a systemic vasculopathy affecting mainly skin and muscles. Renal and neurological involvements are rare in JDM; in the absence of overlap with other CTDs or a primary vasculitis in this case, acute renal injury possibly resulted from myoglobinuria although that is very rare in JDM, unlike in acute viral myositis. The seizures were presumably related to hyponatraemia. His presentation is atypical of JDM, requiring exclusion of malignancy and other causes and warrants treatment with potent immunosuppression.

Disclosure statement: The authors have declared no conflict of interest.

27. ACUTE CHOREA AS A PRESENTING FEATURE OF JUVENILE SLE
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Background: Neuropsychiatric manifestations of juvenile SLE (JSLE) (i.e. chorea) are rare, being present in <3% of cases at diagnosis. Such cases pose significant diagnostic and management challenges.

Here we describe the case of a patient with JSLE who presented with florid chorea and discuss the specific management issues encountered and review the relevant literature.

Case report: A 12-year-old Caucasian girl, with a background history of mild learning difficulties, attention deficit hyperactivity disorder and ASD, presented with a 1-week history of progressive, generalized chorea, emotional lability and intermittent confusion. The history of chorea was preceded by a 2-week history of fever, urinary frequency and dysuria. There was a widespread erythematous rash involving her trunk, a purpuric rash on her buttocks with necrotic lesions on her knees, with evidence of peripheral joint swelling and mouth ulceration. Investigations at presentation demonstrated lymphopenia, normochromic, normocytic anaemia and mild thrombocytopenia. Creatinine kinase was elevated at 799 with an ESR of 23, normal range CRP and renal function. Anti-dsDNA antibodies were present at a titre of >380 with positive ANA, anti-Ro and anti-La antibodies. Complement C3 and C4 were both reduced at 0.6 and 0.08, respectively. Importantly, anti- phospholipid antibodies were not detected and cerebrospinal fluid analysis showed no evidence of malignant infiltration or infection. Magnetic resonance angiogram brain and cathereter cerebral angiogram was normal, except for cervicoscal artery stenosis due to hypoventilation requiring non-invasive ventilation. He had been using ibuprofen with little benefit. Examination showed a pale child, but no rashes or clubbing, with normal aortic and diastolic feeds and no evidence of heart failure.

Conclusion: We describe a patient with JSLE presenting with chorea who had a dramatic response to immunosuppressive therapy and is now stable on sodium valproate and MMF. As neuropsychiatric complications of JSLE may lead to significant morbidity, prompt recognition and early treatment is vital to ensure a successful outcome.

Disclosure statement: The authors have declared no conflict of interest.

28. PAINS IN THE LEGS: A CASE OF GOLDBLOOM SYNDROME?
Ethan Sen1, Mario Abinun2, Mark Friswell3 and Helen Foster3

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Case report: A previously well, 10-year-old Caucasian boy was referred with a 2-month history of significant, bilateral upper thigh pain and difficulty climbing stairs. He had no pains elsewhere and was able to continue all activities except karate. There was a preceding flu-like illness but no fevers. He had been using ibuprofen with little benefit. Examination showed a pale child, but no rashes or clubbing, with normal aortic and diastolic feeds and no evidence of heart failure.

Conclusion: We describe a patient with JSLE presenting with chorea who had a dramatic response to immunosuppressive therapy and is now stable on sodium valproate and MMF. As neuropsychiatric complications of JSLE may lead to significant morbidity, prompt recognition and early treatment is vital to ensure a successful outcome.

Disclosure statement: The authors have declared no conflict of interest.

Background: Neuropsychiatric manifestations of juvenile SLE (JSLE) (i.e. chorea) are rare, being present in <3% of cases at diagnosis. Such cases pose significant diagnostic and management challenges. In all reported patients, symptoms improved 2–3 weeks of symptom onset. In all reported patients, symptoms improved
Case report: We present the case of an 8-year-old girl who initially presented with pyrexia, lethargy, anorexia, weight loss, arthralgia and a dry cough. She had a vasculitic rash on her palms and feet, lymphopenia and raised amylase. A skin biopsy revealed IgA deposits. She also developed acute tubular necrosis. Her working diagnosis at this stage was systemic vasculitis of undetermined cause and she was treated with i.v. methylprednisolone and plasma exchange with good response. She re-presented a month later with acute encephalopathy. She developed secondary haemophagocytic lymphohistiocytosis and was treated with dexamethasone, IVIG and ciclosporin. An MRI brain revealed a brain stem infarct and she developed cardiac dysfunction. She required intubation and ventilation and was also treated with inotropic support and haemodiafiltration. Two weeks after her admission she developed acute tubulointerstitial nephritis and was treated with i.v. cyclophosphamide. Aspergillus in her pleural fluid and cytomegalovirus in her sputum, Aspergillus in her pleural fluid and cytomegalovirus in her blood. She required intubation and ventilation, high-dose inotropic support and haemodialfiltration. Two weeks after her admission she deteriorated further and her blood pressure became increasingly labile. A CT brain showed multiple infarcts and communicating hydrocephalus. Her brain began to cone and despite maximum supportive therapy she was pronounced brainstem dead. Support was withdrawn at this stage and she passed away several hours later. This case demonstrates both the variability in the presentation of SLE and the devastating consequences the disease can have.

Disclosure statement: The authors have declared no conflict of interest.

29. AN UNUSUAL CASE OF PAEDIATRIC SLE

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Case report: We present the case of an 8-year-old girl who initially presented with pyrexia, lethargy, anorexia, weight loss, arthralgia and a dry cough. She had a vasculitic rash on her palms and feet, lymphopenia and raised amylase. A skin biopsy revealed IgA deposits. She also developed acute tubular necrosis. Her working diagnosis at this stage was systemic vasculitis of undetermined cause and she was treated with i.v. methylprednisolone and plasma exchange with good response. She re-presented a month later with acute encephalopathy. She developed secondary haemophagocytic lymphohistiocytosis and was treated with dexamethasone, IVIG and ciclosporin. An MRI brain revealed a brain stem infarct and she developed cardiac dysfunction. She required intubation and ventilation and was also treated with inotropic support and haemodiafiltration. Two weeks after her admission she developed acute tubulointerstitial nephritis and was treated with i.v. cyclophosphamide. Aspergillus in her pleural fluid and cytomegalovirus in her sputum, Aspergillus in her pleural fluid and cytomegalovirus in her blood. She required intubation and ventilation, high-dose inotropic support and haemodialfiltration. Two weeks after her admission she deteriorated further and her blood pressure became increasingly labile. A CT brain showed multiple infarcts and communicating hydrocephalus. Her brain began to cone and despite maximum supportive therapy she was pronounced brainstem dead. Support was withdrawn at this stage and she passed away several hours later. This case demonstrates both the variability in the presentation of SLE and the devastating consequences the disease can have.

Disclosure statement: The authors have declared no conflict of interest.

30. WHAT COMES FIRST, THE INVESTIGATIVE RESULTS OR THE ACQUIRED HISTORY? A CASE IN WHICH WE FOLLOWED THE PATHOGEN, NOT THE PATIENT

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Introduction: The art of history taking is most challenging to the paediatrician. The basic human responses we rely on as physicians are usually inhibited, distorted or exaggerated. A delay in the correct diagnosis can lead to further complications, as seen with numerous childhood diseases. This case highlights some of the essential learning points to consider when approaching the unwell child with seemingly normal clinical parameters.

Background: A 4-year-old girl presented with non-specific abdominal pain and a raised temperature. Studies have shown that fewer than 1 in 10 children febrile on admission have an underlying significant bacterial infection. Surgical review and investigations ruled out the diagnosis of appendicitis and she was discharged later that day. Forty-eight hours later the patient was readmitted after blood cultures taken on the previous admission grew Staphylococcus aureus. A rise in inflammatory markers indicated an MRI scan of the hips, revealing a focal 2 cm abscess formation in the left adductor brevis muscle, with similar changes on the right. The paediatric orthopaedic team deemed drainage of the collection unnecessary. Clinical improvement followed a 2-week course of i.v. flucloxacillin and the patient was discharged with a 16-day course of oral flucloxacillin. A review of the case illustrated a possible cause, a history of impetigo followed by prolonged exposure to horse riding.

Discussion: The repeated activity of horse riding has been shown to apply strain to the adductor brevis muscle, which may have caused an injury and contributed towards the abscess formation.

Conclusion: In an age where high-resolution imaging and advanced laboratory techniques are changing the way in which we diagnose patients, the fundamental principles of clinical practice must not be forgotten. We still possess the most powerful weapon in combating illness, a thorough and targeted history followed by a detailed and relevant examination.

Disclosure statement: The authors have declared no conflict of interest.