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 LN 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 14.1 (11.6–15.8) years and 80% female. Active renal episodes (23% total; renal BILAG A/B) had significantly increased concentration of monocytic chemotactant 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 66%, sensitivity 61%). 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?

Phil O’Neill1, Helen Lee2 and Rachel Tattersall3

1University of Sheffield Medical School, 2Sheffield Children’s Hospital and 3Sheffield Teaching Hospitals, Sheffield, UK

Correspondence to: rachel.s.tattersall@sth.nhs.uk

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 enshrined in the Department of Health You’re Welcome (YW) policy [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) 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
SARC). The audit was done by P.O’N. using standard audit procedures.

Results: Neither hospital within SARC, reached the Meets You’re Welcome (You’re Welcome) criteria. Young people 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 YFP is not all talk and no action.

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

References

O4. MINIMAL DISEASE ACTIVITY IN A CLINICAL COHORT OF CHILDREN WITH JIA: RESULTS FROM THE CHILDHOOD ARTHRITIS PROSPECTIVE STUDY
Flora McErlane, Michael Beresford, Eileen Baldam, S. E. Alice Chiang, Joyce Davidson, Helen Foster, Janet Gardner-Medwin, Mark Lunt, Lucy R. Wedderburn, Wendy Thompson and Kimme L. Hyrich
1University of Liverpool, Liverpool, 2Alder Hey Children’s NHS Foundation Trust Hospital, Liverpool, 3Royal Manchester Children’s Hospital, Manchester, 4Royal Hospital for Sick Children, Glasgow, 5Great Ormond Street Children’s Hospital, London, 6The Royal Hospital for Children, Glasgow, 7University of Liverpool, 8University College London, UK

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 in children [1], with two discriminative outcomes for oligoarticular [physician global assessment (PGA) <2.5 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) with available data were included. MDA was defined 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 $^2$ 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), 46% 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 85% 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>Group 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>65 (625)</td>
<td>65 (473)</td>
<td></td>
</tr>
<tr>
<td>Oligoarticular pattern</td>
<td>9 (637)</td>
<td>55 (620)</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>F (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

05. AUDIT OF INCIDENCE OF SCARRING FOLLOWING STEROID TENDON INJECTIONS FOR TENOSYNOVITIS IN CHILDREN WITH JIA: A SINGLE-CENTRE EXPERIENCE
Akhila Kavirayani, M. S. Thyagarajan, Jane Ellis, C. N. S. Helen Strike and A. V. Ramanan
Bristol Royal Hospital for Children, Bristol, UK
Correspondence to: akhi13n@yahoo.com

Background: Superficial skin scarring is a well-recognized complica-
tion 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.

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 2.3 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 instilled in 7/24 (29%) cases. Scarring of injection site was identified either by documentation in notes or a telephone call to the parents, with explanation of anatomical landmarks.

Conclusion: This audit is the first to focus on the incidence of tendon scarring in children with JIA, following steroid tendon injections. There is a significant incidence of tendon scarring, which could be improved by the use of topical local anaesthetic.
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
Andrea Coda1, Joyce Davidson2, Peter Fowlie3, Jo Walsh2, Tom Carlile3 and Derek Santos1
1Queen Margaret University, Edinburgh, 2Royal Hospital for Sick Children, Edinburgh and 3Ninewells Hospital, Dundee, UK

Correspondence to: acoda@qmu.ac.uk

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 patients 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 (± 3.51) and for the trial group was 10.64 (± 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 = 23) of the children 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 < 0.05); 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 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 within the multidisciplinary team in paediatric rheumatology and hopes to improve the profile of podiatrists working within paediatric hospitals and private practices.

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

2. GET A GRIP
K. W. Brimlow and S. Rangaraj
Nottingham Children’s Hospital, Nottingham, UK

Correspondence to: Kath.Brimlow@nuh.nhs.uk

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 ~20 min 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 Jamar hand hydraulic dynamometer on patients with JDM.

Objectives: 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. 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 testing both CMAS and hand grip strength for all 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 PAEDIATRIC POPULATION
Caroline Grant
NHS Greater Glasgow and Clyde, Glasgow, UK

Correspondence to: caroline.grant@ggc.scot.nhs.uk

Background: The Bath Ankyllosing 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.

Objectives: This pilot study was conducted to assess whether BASMI is a usable 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 each child. Those individuals who had sequential measurements showed improvement in all areas during the assessment period. In one
individual in whom Childhood Health Assessment Questionnaire (CHAQ) scores were available at the same time points, no correlation was noted. BASMI scores appeared more closely related to patient global sway of body than others, e.g., trunk-gait showed little change over time and therefore may be a less useful measure in this age group.

Conclusion: This small pilot study demonstrated that BASMI could be used in the paediatric age group and was well tolerated by children and teenagers. With the BASMI the score is added up and divided by 5 to give a score out of 10. In this population it was found to be more sensitive to change as a score out of 50 when charting sequential measurements. Sequential BASMI measurements appear to be a useful method of determining a trend of improvement or deterioration and might prove a useful additional tool for documenting response to treatment in this patient group. This adapted BASMI should be validated in a larger cohort of children with ERA and correlated with other markers of disease activity.

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

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, Annie Hinchcliffe3, Andrew Dick4 and Athimalaipet Ramanan5
1University of Bristol, 2Bristol Royal Hospital for Children and 3Bristol Eye Hospital, Bristol, UK

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. The main outcome measures recorded were: anterior chamber cell 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 unresponsive to previous treatment with two immunosuppressive agents. Before triple therapy started, 14 of the 18 eyes had active anterior uveitis and were being treated with topical corticosteroid drops. Six months after the commencement of triple therapy, there was a clinically significant improvement in ocular inflammation in seven of these eyes and nine of 14 eyes were receiving a tapered dose of topical corticosteroids. Over the course of follow-up, 10% of patients received additional steroids for uveitis. The main outcomes recorded were: anterior chamber cell grade, ability to reduce topical corticosteroids, presence of ophthalmic complications, serious adverse events and side effects.

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/RCPoPTH SCREENING GUIDELINES, AT QUEENS MEDICAL CENTRE, NOTTINGHAM AND SHEFFIELD CHILDREN’S HOSPITAL

Diana Ekdawy1, Gupreet Nagra2, Nikki Caminsa3,4, Jenny Edgerton5, Jessy Choi5, Katy Lamb3,4, Daniel Hawley6 and Satyapal Rangaraj1
1Doncaster Royal Infirmary, Doncaster, 2Nottingham University, 3Queen’s Medical Centre, Nottingham, 4Sheffield Children’s Hospital, Sheffield and Nottingham Children’s Hospital, Nottingham, UK

Correspondence to: diamond56895@hotmail.co.uk

Background: The prevalence of uveitis in JIA is ~8–30%. This increases to 45–57% in the oligoarthritis subset. Untreated it can result in visual loss, cataracts and glaucoma, and once these complications have arisen they are often irreversible. Joint guidelines have been developed by the British Society of Paediatric and Adolescent Rheumatology (BSPAR) and the Royal College of Ophthalmology (RCOphth) to reduce the incidence of visual impairment among children diagnosed with JIA by early detection of uveitis through screening, allowing for early intervention.

Objective: To review and compare the uveitis screening services at Queen’s Medical Centre Nottingham (QMC) and Sheffield Children’s Hospital (SCH) regarding compliance with RCPoPTH/BSPAR guidelines. Through performing regional audit to elucidate areas of strength and weakness in local service provision and share best practice.

Methods: Retrospective case-note review of patients needing uveitis screening was performed. Patients at QMC referred for ophthalmology screening over a 6-month period were identified, while patients at SCH receiving an initial diagnosis of JIA over a 2-year period were identified. Data were collected using a common proforma, analysed and compared.

Results: In both centres oligoarthritis was the most common subtype of JIA with 29% at QMC (n = 29) and 60% at SCH (n = 25). QMC referred 100% of patients for an initial ophthalmology screening after JIA diagnosis compared with 84% at SCH. At QMC 83% of those patients had their screening appointment within the 6-week guidelines compared with 71% at SCH. 59% of patients at QM continued to have follow-up screening after the initial ophthalmology appointment compared with 86% at SCH.

Conclusion: Neither centre adhered closely enough to screening guidelines. Interesting differences between the two centres included: (i) QMC patients were more likely to receive their initial screening appointment within 6 weeks of referral; and (ii) SCH patients were more likely to receive follow-up screening adhering to guidelines. Audit findings have been presented and discussed at established regional network meetings co-organized by QMC and SCH. Shared best practice has been reflected in changes instituted at each centre informed by practice strengths and weaknesses identified through this audit. A regular programme of regional audit is now planned and collaboration with other centres will be welcome.

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

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

Mary Cruikshank1, Ethan Sen2, Clare Pain3 and Valentina Leone4
1Newcastle University, Newcastle upon Tyne, 2MACRN Arthritis Research UK CSG, 3RCPCH Specialist Advisory Committee for Paediatric Rheumatology and 4British Society of Paediatric and Adolescent Rheumatology, UK

Correspondence to: mary.cruikshank@doctors.org.uk

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 trainee’s 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: Sixteen trainee teleconferences have been held, on the third Tuesday of each month between 8 and 9pm, 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 median number of trainees attending each meeting has been 7 (range 5–13) for 4(–0) topics. There have included eosinophilic fasciitis, consensus
treatment for JDM, debate of anakinra as first-line treatment in systemic onset JIA, SLE and antiphospholipid syndrome, and SLE and transverse myelitis. Discussion feedback has been positive. Use of PowerPoint presentations has worked well via phone and generated lots of discussion. The majority of Grid trainees attended. Technical difficulties have occurred on two occasions but were quickly resolved. The meetings have provided closer links between CSAC, CSG and BSPAR trainee representatives and support for trainees across the country. Skills in teleconferencing have been gained. It is acknowledged that this method of education does not replace the advantages of face-to-face educational events. However, it can be a useful additional resource for sharing and learning from experience.

Submitted on behalf of BSPAR trainees.

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

7. THE ROLE OF VIDEOCONFERENCING AS A METHOD OF EDUCATION DELIVERY WITHIN A PAEDIATRIC RHEUMATOLOGY CLINICAL NETWORK

Mary Cruikshank1 and Jo Walsh2 on behalf of the SPARN Education Workgroup

1Newcastle University and Scottish Paediatric and Adolescent Rheumatology Network
Correspondence to: marycruikshank@doctors.org.uk

Background: Addressing and delivering educational needs for health care professionals working in a clinical network is challenging, particularly for small specialties that are spread over large geographical areas. In this is pertinent in Paediatric Rheumatology. There is increasing interest in the role that videoconferencing can play as a mechanism of increasing access to medical education within a clinical network. The Scottish Paediatric and Adolescent Rheumatology Network (SPARN) was established formally in 2009. A learning needs analysis identified an unmet need for medical education. It was proposed that a videoconference education programme be established with an aim of addressing these unmet educational needs.

Methods: The educational videoconference session was initially trialled between the two tertiary centres. Technical issues were resolved, then other network centres were invited to attend (n = 14). Topics included a range of paediatric rheumatology topics suggested by the learning needs analysis. Interdisciplinary presenters were coordinated from the two tertiary centres. The educational programme was advertised on the SPARN website and e-mail list (n = 122). Local centres were responsible for coordinating their hospital videoconference facility. A multisite bridge linked participating centres. Central technical support was provided by the Telemedicine Department at Yorkhill Hospital, Glasgow. When case presentations were used no identifiable information was presented and consent had been obtained.

Results: 18 videoconference teaching sessions, each lasting 2 h, have now been held, on a monthly basis since October 2010. The number of centres attending has increased (n = 14) and included representatives of all specialties of the network. Topics have included: generic teaching on JIA, paediatric CTDs, JDM, infection and rheumatology, non-organic pain syndromes, orthopaedic problems in rheumatology and medications used in paediatric rheumatology. A joint session on vasculitis was hosted between SPARN and Scottish Paediatric Renal Network (SPRIN) in 2012.

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 supporting education within a clinical network.

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

8. THE ROD AMOS RHEUMATOLOGY EDUCATION (RARE) DAY: PART OF THE SOLUTION TO CLINICAL SKILLS TRAINING IN PAEDIATRIC AND ADOLESCENT RHEUMATOLOGY?

Rachel Tattersall1, Dan Hawley2, Lisa Dunkley1, Helen Lee1 and Anne-Marie McMahon2

1Sheffield Teaching Hospitals and 2Sheffield Children’s Hospital, Sheffield, UK
Correspondence to: rachel.s.tattersall@sth.nhs.uk

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 skills 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 reported 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 participant’s 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 concerns regarding missing school/ work were overridden as being ‘just like another appointment’.

Conclusions: The VWR is applicable to CYP and was positively evaluated by clinicians, participants and patients. Joint PAR training and simulated clinical scenarios are effective and focus efforts in times of scarce resources. Patients As Rheumatology Teachers are a beneficial PART of improving training. We recommend the RARE day as a practical and applicable model for innovating education in PAR.

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

References


Peter Bale1 and Kate Armon2
1Ipswich Hospital NHS Trust, Ipswich and 2Norfolk and Norwich University Hospital, Norwich, UK
Correspondence to: Pete.Bale@nyhoo.co.uk

Background: In 2010 B Spar 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] has 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.

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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 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 respondents, 23 of the 43 standards were ranked vital more than other category. 40% of respondents felt they shouldn’t travel for longer than 1h 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>
<thead>
<tr>
<th>Table 1</th>
<th>Demonstrates the 10 standards ranked vital the most times and the 10 standards, which were ranked in the top 10 the most times</th>
</tr>
</thead>
<tbody>
<tr>
<td>Standard</td>
<td>The following standards were ranked vital most frequently</td>
</tr>
<tr>
<td>22</td>
<td>Drugs used for the treatment of JIA will be prescribed and monitored in accordance with BSPAR/NICE guidelines</td>
</tr>
<tr>
<td>5</td>
<td>Health care practitioners should refer all children with suspected JIA to a paediatric rheumatology team within 6 weeks of symptom onset</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>
</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>
</tr>
<tr>
<td>8</td>
<td>Children with JIA and active disease should have a full assessment of their disease, health, psycho-social and pain management and educational needs</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>
</tr>
<tr>
<td>7</td>
<td>Members of the paediatric rheumatology team will have appropriate training and experience as defined by professional bodies</td>
</tr>
<tr>
<td>18</td>
<td>Those children with JIA and active disease should have regular specialist review</td>
</tr>
<tr>
<td>29</td>
<td>Specialist surgery should be performed by appropriately trained surgeons with experience in the management of JIA</td>
</tr>
<tr>
<td>4</td>
<td>All health care professionals likely to come into contact with a child with JIA should acquire the skills to recognize the condition</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

4. Wilkinson N. Questionnaire developed for Oxford Deaneary parental survey 2011, provided to Kate Armon for use in this survey.

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

Tania Amin1 and Mark Wood2

1Leeds Teaching Hospital Trust and 2Leeds Children’s Hospital, Leeds, UK

Correspondence to: taniaamin@hotmail.co.uk

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-α agent was a second anti-TNF-α in ~80% of respondents and for 92%, 100% and 100% of respondents respectively within the first three choices.
(ii) In RF-positive patients rituximbab (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-TNFα 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

Rebecca Davies1, Tauton R. Southwood2, Lianne Kearsley-Fleet3 and Kimme L. Hyrich1 on behalf of the BSPAR Etanercept Register.

1Manchester Academic Health Science, Manchester, UK
2University of Manchester & Birmingham Children’s Hospital and 3Manchester Academic Health Science, Manchester, UK

Correspondence to: rebecca.davies@manchester.ac.uk

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.

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Methods: The national BSR/PAS/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 was recently recruited. To 31 December 2011, 876 patients with JIA 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 osteopenia or osteoporosis (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 3%) and skeletal 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 comparing 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
Lianne Kearsley-Fleet1, Eileen Baldam2, Michael Beresford2, Rebecca Davies3, Helen E. Foster4, Katy Mowbray1, Tauton R. Southwood4, Wendy Thomson3 and Kimme L. Hyrich4
1Manchester Academic Health Science Centre, 2Alder Hey Children’s Hospital, Liverpool, 3University of Manchester, 4Newcastle University and 5University of Birmingham & Birmingham Children’s Hospital, UK
Correspondence: lianne.fleet@manchester.ac.uk

Background: The introduction of biologic therapies has revolutionized 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 (BCRD) study is a long 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 therapy, is collected at baseline. Utilising 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 = 0.007). 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 prior biologics and 1 child (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 on 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
Manjari Agarwal1, Akhila Kavirayani3, A. V. Ramanan1 and Jane Ellis1
1Bristol Royal Hospital for Children, Bristol, UK
Correspondence to: akhilamk@yahoo.com

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.

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
Eve Smith1, William Gray2, David Taylor-Robinson3, Helen E. Foster4 and Michael W. Beresford3
1Great North Children’s Hospital, 2Northumbria Healthcare NHS Foundation Trust, University of Liverpool, 3Newcastle University and 4Alder Hey Children’s NHS Foundation Trust, Liverpool, UK
Correspondence to: evemsmith@yahoo.co.uk

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 rheumatology, 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 rheumatology 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 containing with time to diagnosis included: being British (odds ratio [OR] coefficient $= 0.268$, $P = 0.007$), Caribbean or African ($P = 0.159$, $P = 0.011$), 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 ($-0.125$, $P = 0.046$) 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 ($P = 0.047$) or having nephritis ($P = 0.045$) 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, distance from nearest tertiary paediatric rheumatology service, socioeconomic status and family history of autoimmune disease were not found to be significant predictors of access to care.

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

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

Thomas Morgan1, Louise Watson2 and Michael W. Beresford3
1Alder Hey Institute for Child Health, University of Liverpool and 2Alder Hey Children’s NHS Foundation Trust Hospital, Liverpool, UK
3Alder Hey Children’s NHS Foundation Trust Hospital, Liverpool, UK
Correspondence to: thomas.morgan@doctors.org.uk

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 equally accurate in both populations. Children have a more acute, aggressive disease and greater therapeutic burden. Female preponderance is less pronounced in JSLE: female: 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 presented as median and interquartile range (IQR).

Results: Within the cohort of 240 patients, 23% ($n = 56$) were aged under 10 when JSLE was diagnosed [79% female ($n = 44$), 21% male ($n = 12$); ratio 3.7:1]. The median number of ACR criteria, 5.4 (range 4–8) was consistent between males (5.7) and females (5.4). Median ages at diagnosis within this group were 8.2 years for females (IQR 6.5–9.0) and 5.2 years for males (IQR 3.8–7.7). Males were significantly younger than at diagnosis ($P = 0.03$). The median disease duration was 4 years (IQR 1.7–7.0 years). There was a trend towards discoid rashes occurring more frequently in males, though not reaching statistical significance (male 33%, female 9%; $P = 0.06$). There was no difference in the preponderance of arthritis in males (67%) vs females (57%) in those aged <10 years. There were no observed differences between the sexes in any of the 10 other ACR criteria either at diagnosis or latest follow-up.

Conclusions: These results suggest that the clinical phenotype of JSLE in the youngest patients does not greatly differ between male and female patients. This contrasts with comparisons of adolescent and adult SLE patients in whom a greater difference in the clinical phenotype exists between the sexes.

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

Faekah Gohar1, Louise Watson2 and Michael W. Beresford3
1Institute of Women’s and Child Health, University of Liverpool, 2Alder Hey Institute for Child Health, University of Liverpool and 3Alder Hey Children’s NHS Foundation Trust Hospital, Liverpool, UK
Correspondence to: faekah@liverpool.ac.uk

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 twelve-month follow-up (T12); and (ii) associations of TCP with disease activity, laboratory markers and disease-related damage.

Method: Patients recruited between 2004 and 2011 to the UK JSLE inception cohort study and repository 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 $\leq 150$ 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 (90 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 $\leq 50,000$/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); $P = 0.005$]. Disease-related damage accrual, measured by the SLICC 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

Bahar Artin-Esen1, Ania Radziszewska1, Charis Pericleous2, Aniria Rahman1, Ian Giles1 and Yiannis Ioannou1
1University College London, UK
2Institute of Women’s and Child Health, University of Liverpool, UK
Correspondence to: ania.radziszewska@ucl.ac.uk

Background: We have recently shown that adult patients with SLE have increased anti-Fxα 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 Xα 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 VIIa (FVIIa) and Factor Xα (Fxa) by ELISA. Results were expressed as a percentage of binding compared with a positive control where the positive value was defined as being $\geq 2.5\%$, above the mean of adult healthy controls.

Results: Only IgG anti-Fxα antibodies were found to be found more frequently in adult patients with SLE ($n = 52/106$, 49.1%) compared with JSLE ($n = 5/35$, 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

Darryl Jashek1, Ellen Mosley2 and Satyapal Rangaraj2
1University of Nottingham and 2Nottingham Children’s Hospital, Nottingham University Hospitals NHS Trust, Nottingham, UK

Correspondence to: mzydd@nottingham.ac.uk

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 studies, 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.

Author Patient Study type Outcome

Guenika A et al., 2011 [1], Belgium 53 JSLE patients, DXA scanning performed Cross-sectional study No direct correlation was observed between the analysed parameters and densitometry findings according to the Z-score

Comperry, Lacassagne S, 2007 [2], Canada 64 JSLE patients, DXA scanning performed Cross-sectional study 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.

Wright TB et al., 2009 [3], USA 38 JSLE patients, 207 healthy controls Cross-sectional study Severe vitamin D deficiency (25(OH)D <10 ng/ml) was observed in a significantly higher proportion of subjects with SLE (P = 0.001).

Casella CB et al., 2012 [4], Brazil 57 JSLE patients, 37 healthy controls Cross-sectional study 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 (>400 IU).

References

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

Elena Moraitis1, Katie Arnold2 and Clarissa Pilkington1
1Great Ormond Street Hospital, London and 2University College London, UK

Correspondence to: elenamoraitis@gmail.com

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 26/46 (56%) had no rash, 32/46 (69%) 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 OSTEOMYELITIS: A CASE SERIES ANALYSIS

Nicki-Jayne Russell1, Marion Roderick2 and Athimalaipet Ramanan2
1University of Bristol and 2Bristol Royal Hospital for Children, Bristol, UK

Correspondence to: nr8035@bristol.ac.uk

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.
Methods: Case-notes of 38 patients undergoing treatment for CRMO from 2003 to 2012 at Bristol Children’s Hospital were analysed and details entered into a custom-made spreadsheet containing 82 parameters. In order to establish the criteria that had been used for diagnosis, plain films, CT, MRI, bone scans and biopsy results were recorded.

Results: The most common presenting location of bony swelling was the clavicle (29%), followed by tibia (26%), femur (24%) and spine (21%). The preceding differential diagnoses considered were infectious osteomyelitis (26%), Langerhans cell histiocytosis (13%) and malignancy (11%). The median age of the cohort is 9 years (3–14), with a female: male ratio of 29:9. All children had plain radiographs, many of them substantial in number. Fifty-three per cent of children received prolonged courses of antibiotics prior to diagnosis. MRI STIR was performed in 71% of patients, all showing characteristic signal enhancement. Biopsies were taken in 71% of cases and bone scans in 42%. Thirty-three (87%) were treated with NSAIDs with 16 (48%) suffering from breakthrough pain. These 16 patients were treated with bisphosphonate 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.

21. PAMIDRONATE THERAPY FOR CHRONIC RECURRENT MULTIFOCAL OSTEOMYELITIS: A CASE SERIES ANALYSIS

Marion Roderick1, Nicki Russell1 and A. V. Ramanan1
1Bristol Royal Hospital for Children, Bristol, UK

Correspondence to: avramanan@hotmail.com

Background: There is increasing evidence supporting the use of bisphosphonates, particularly pamidronate, for patients with chronic recurrent multifocal osteomyelitis (CRMO) who have breakthrough pain on NSAIDs. Pamidronate is known to inhibit osteoclasts and may reduce CRMO lesion expansion by this action; however, it also has anti-cytokine properties and this may also be what makes it an effective treatment for CRMO.

Objective: To establish the safety and efficacy of pamidronate in the reduction of clinical symptoms and radiological features of CRMO in a cohort of 18 patients at Bristol Children’s Hospital.

Methods: Case-notes of 38 patients undergoing treatment for CRMO from 2003 to 2012 at Bristol Children’s Hospital were analysed. The children undergoing pamidronate treatment were identified and their radiology and clinical findings examined in relation to their treatment. Symptoms were assessed using a questionnaire regarding functionality and bone pain. MRI scans were performed in all patients following therapy.

Results: Thirty-three of 38 patients (87%) had been treated with NSAIDs and 16 of 33 (42%) were suffering from persistent symptoms. A total of 18 patients were treated with i.v. pamidronate (1 mg/kg/day, 3-day regimen, 3-month intervals for up to 1 year) and received an average of four doses. If symptoms persisted, they received a further infusion (providing MRI results were favourable). Of the 18 children treated with pamidronate, at the time of this review, 11 had completed the dosage regime. Of these 11 completing treatment, 7 (84%) 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 at the time of this review and had no post-treatment MRI results.

Conclusion: In children with CRMO resistant to NSAIDs, pamidronate may induce remission in a significant number of patients as demonstrated by both clinical symptoms and radiological assessment. However, as CRMO is a relapsing remitting condition a randomized controlled trial would provide the best evidence but the rarity of the condition makes this very difficult.

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>Maneuver tested</th>
<th>All MPS (n = 15)</th>
<th>MPS I (p = 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>
</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>
</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>
</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>
</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>
</tr>
<tr>
<td>MCP, DIP, PIP extension</td>
<td>Hands and wrists together</td>
<td>68.9</td>
<td>50</td>
<td>100</td>
<td>91.7</td>
</tr>
<tr>
<td>Forward flexion of spine</td>
<td>Bend forwards. Observe curvature of spine from all sides</td>
<td>67.2</td>
<td>66.7</td>
<td>100</td>
<td>66.7</td>
</tr>
<tr>
<td>Spinal deformity</td>
<td></td>
<td>66.7</td>
<td>70.8</td>
<td>100</td>
<td>55.6</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>
</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>
</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>0</td>
</tr>
<tr>
<td>Neck extension</td>
<td>Look up to the ceiling</td>
<td>57.8</td>
<td>45.8</td>
<td>100</td>
<td>75</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>
</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>200</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>
</tr>
<tr>
<td>Forearm supination</td>
<td>Turn hand over</td>
<td>54.5</td>
<td>35.3</td>
<td>50</td>
<td>81.8</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>
</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>
</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>
</tr>
<tr>
<td>Knee flexion</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>
</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>
</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>
</tr>
<tr>
<td>Ankle planter flexion</td>
<td>Walk on tip-toes</td>
<td>8.3</td>
<td>4.2</td>
<td>N/D</td>
<td>22.2</td>
</tr>
</tbody>
</table>

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

and inter-observer κ = 0.62 (range 0.51–0.77). Hip manoeuvres 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 further 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?

Julia Clarkson1, Janet Gardner-Medwin2 and Vincent Choudhery2

1Glasgow University and 2Royal Hospital for Sick Children, Glasgow, UK

**Correspondence to:** 1103540c@student.gla.ac.uk

**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 staff (24 consultants, 38 A+E trainees, 7 GP trainees and 5 paediatric trainees). Only 20 RLs included a diagnosis that matched the PR diagnosis, 34 RLs were not clearly demonstrated in the videos.

25. ANCA-POSITIVE VASCULITIS ASSOCIATED WITH LEVAMISOLE

Catriona McCvitty1, Joyce Davidson1, D. H. Hughes1 and Neil Martin1

1Royal Hospital for Sick Children, Glasgow, UK

**Correspondence to:** cmccvitty@gmail.com

**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. P-ANCA was strongly positive with both MPO and PR3 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, whilst it is perhaps more important 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 Shamilla Jandial2

1Northern Deanery and 2Newcastle upon Tyne Hospitals, UK

Correspondence to: kishowarrier@doctors.org.uk

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. 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) with haemolysis but no dysproteinaemia. Pediatr Nephrol 1999;13:602–3.

27. ACUTE CHOREA AS A PRESENTING FEATURE OF JUVENILE SLE

Daire O’Leary1, David Staunton1, Cloidagh Lowry2 and Niamh McSweeney1

1Cork University Hospital and 2Our Lady’s Children’s Hospital, Cork, Ireland

Correspondence to: daireo1@yahoo.com

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 ESFR 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 catherer cerebral angiogram were normal with periventricular changes on T2. There was no evidence of active inflammatory myopathy and very weak proximally (CMAS 11).

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

1MCRN/Arthritis Research UK CSG, UK, 2Newcastle upon Tyne Hospitals and 3Great North Children’s Hospital, Newcastle upon Tyne, UK

Correspondence to: ethan.sen@doctors.org.uk

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 flue-like illness but no fevers. He had been using ibuprofen with little benefit. Examination showed a pale child, but no rashes or clubbing, with general weakness of his thighs but normal muscle strength at distal (ipsilateral). MRI of both thighs showed intense oedema encircling the femoral shaft on the right and along the medial aspect of the femur on the left. The features were in keeping with a myoperiostitis. The patient was discussed with Paediatric Haematology, Immunology and Orthopaedic Oncology teams. Subsequently a skeletal survey showed peristemal new bone formation along the shaft of the distal radius and ulna, and upper tibia. Bone marrow aspirate showed no evidence of malignancy. Trephine biopsy revealed reactive, mildly hypercellular marrow with scattered reactive plasma cells. At the bony surface was an area of inflammatory tissue with expanded stroma and secondary fibrosis. The patient was managed mostly as an outpatient with brief admissions for investigations. He was treated with regular oral naproxen. At follow-up, 5 weeks after presentation, he remained systemically well with some improvement in his thigh pain. There was almost complete normalisation of inflammatory markers with ESR 15 and CRP 5. A search of the literature revealed similarity between our patient and those with Goldblloom syndrome, of which there are fewer than 15 reported cases. The condition was originally described by Goldblloom and others in 1986 in two school-age children characterized by idiopathic periosteal hyperostosis with dysproteinaemia. Both had bone pain and recovery with widespread periosteal new bone formation on radiographs. Dysproteinaemia was characterized by elevated γ-globulin levels 2–3 times the upper limit of normal. Unlike our patient, fever was a prominent feature in these original cases and presentation was after weeks of symptom onset. In all reported patients, symptoms improved.
spontaneously over several months. We recommend that in children with limb pain and periosteal hyperostosis, Goldblum syndrome is included in the differential diagnosis.

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

Adnan Raja
Department of Paediatrics, Hull Royal Infirmary, Hull, UK
Correspondence to: adnan.raja@nhs.net

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.