AB1103 SEVERE PULMONARY ARTERIAL HYPERTENSION AS THE INITIAL MANIFESTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS IN A 7-YEAR-OLD MALE PATIENT: A CASE REPORT

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Background: Pleural pulmonary manifestations in patients with systemic lupus erythematosus are reported in approximately 5% of cases. Presenting as pleural effusion, alveolar haemorrhage, diffuse interstitial lung disease, pulmonary infections and pulmonary arterial hypertension, among others, they are a manifestation difficult to diagnosis. Pulmonary arterial hypertension is a rare condition, usually occurring 3 to 5 years after the diagnosis of systemic lupus erythematosus.
Objectives: We report the case of a 7-year-old male patient who presented with pulmonary arterial hypertension as the initial manifestation of systemic lupus erythematosus.
Methods: Case report
Results: A 7-year-old male patient who was admitted to the Pneumology Department at Children's Hospital of Mexico Federico Gómez due to respiratory distress. In emergency assessment, pulmonary arterial hypertension of 66 mmHg was identified, of unknown cause. The patient did not have significant medical background, having enjoyed of good health up to 6 months prior to his admission. He presented with a history of non-quantified fever, as well as episodes of fatigue and dyspnea. Two months before admission, chest pain was added, exacerbated with inspiration. On admission, transthoracic echocardiography revealed severe dilatation of right cavities, moderate tricuspid insufficiency, with left ventricular ejection fraction of 56% and arterial pulmonary pressure of 66 mmHg. The diagnostic approach is initiated. Due to a history of pulmonary tuberculosis in the patient grandmother, the patient was studied with BAAR and cervical lymph node biopsy, ruling out the diagnosis. Infectious process causing the manifestations was also ruled out. The patient was discharged with medical treatment, requiring readmission in for 7 days, with facial oedema and in lower extremities, generalised pallor, asthenia, adynamia and 4 days before a decrease in urinary volumes and frequency. On admission, right heart failure, secondary to increase of pulmonary hypertension for discontinuation of diuretic administration. A renal biopsy was performed, which was reported as class IV lupus nephropathy, with an index of activity and chronicity of 0. The diagnosis of systemic lupus erythematosus is integrated based on the ACR criteria. Induction of remission of lupus nephropathy based on the CARRA protocol. As treatment was administered the patient showed important clinical improvement.
Conclusions: Pulmonary arterial hypertension is a rare condition, usually occurring 3 to 5 years after the diagnosis of systemic lupus erythematosus. In paediatric population, it is reported as a lupus complication in 5% to 14% of patients, and less than 1% as an initial manifestation. It is a clinical complication that gives the patient a high risk of morbidity and mortality. It is important to acknowledge that pulmonary arterial hypertension can be the initial manifestation of lupus in paediatric population. A prompt identification assures a prompt treatment and a better prognosis.

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AB1104 MALIGNANCY-RELATED ARTHROPATHIES IN PAEDIATRIC PATIENTS SEEN IN A TERTIARY HOSPITAL

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Background: Along with constitutional symptoms such as fever, fatigue, and weight loss, childhood malignant diseases often also present with various musculoskeletal symptoms and findings, prompting consult or referral to a paediatric rheumatology clinic.
Objectives: The study described the characteristics of patients seen at the paediatric rheumatology clinic who were eventually diagnosed with malignancy.
Methods: Medical records of 35 children diagnosed with hematologic or solid malignancy who were seen at the paediatric rheumatology clinic from August 2013 to December 2017 because of with joint pains or difficulty in ambulation were reviewed.
Results: Twenty of the 35 patients (57.1%) were males and 15 (42.9%) were females. The mean age at diagnosis was 7.3 years old ±4.1 (range 2.5–17.0). The time of diagnosis from the onset of symptoms has a mean of 4.5 months±7.4. Fever was the most common constitutional symptom (82.8%), followed by pallor (45.7%) and weight loss (37.1%), while the most common musculoskeletal symptom or finding was joint pains (100%), followed by difficulty in ambulation (40%), joint swelling (37.1%) and nocturnal pains (31.4%). Mean haemoglobin level was 93.9 g/L±18.9. Mean white cell count was 11.9 × 10^9/L±7.1, mean segmenters was 41.6%±21.7, and mean lymphocytes was 53.0%±24.2. Mean platelet count was 294.0 × 10^9/L±220.5. Mean ESR was 99.6 m/h±33.7 at baseline. Radiographic evaluation done were all normal. Peripheral blood smears and bone marrow aspiration study were done in all patients. Thirty-three patients (94.2%) were diagnosed with acute leukaemia, 18 of which was acute lymphocytic leukaemia, 14 acute lymphocytic leukaemia pre-B, and 1 acute myelogenous. One patient had Non-Hodgkins Lymphoma and 1 patient had hemoblastoma.
Conclusions: Children with joint pains associated with findings atypical of a rheumatologic disorder presentation should be further investigated for the possibility of malignancy.

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AB1105 INFLUENCE OF JUVENILE IDIOPATHIC ARTHRITIS ON THE QUALITY OF LIFE OF YOUNG ADULTS IN THE TRANSITION PERIOD TO ADULT RHEUMATOLOGIC CARE IN UKRAINE

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Background: Young adults with juvenile idiopathic arthritis (JIA) often have an active disease with significant functional impairment in adulthood that can affect their physical and mental functions.
Objectives: The aim of the study was to determine the impact of JIA on quality of life, mental and physical health in young adults with JIA during the transition healthcare in Ukraine.
Methods: The cross-sectional study of 89 young adults aged 16 to 22 years with a history of JIA and 25 age- and sex-matched controls (without rheumatic disease) were included. The study was performed at the Aleksandrivsky Central Clinical Hospital, Kyiv, Ukraine in the period between April 2015 and February 2017. There was performed an evaluation of the disease activity by JADAS, received therapy, quality of life by the SF36, the functional activity (HAQ), long-term JIA damage indices JADI-A and JADI-E,[1] PHQ-9, Beck depression scale.
Results: Patients with JIA had worse ($p<0.001$) physical health in comparison with the control group. Physical ($p<0.001$) and role ($p<0.05$) functioning and bodily pain ($p<0.001$) in JIA patients were decreased, compared with the controls. However, the indicators responsible for psychological function in patients with JIA did not differ from the controls. The analysis of functional activity revealed a strong negative effect of the disease on physical role functioning ($p<0.001$), bodily pain ($p<0.001$), general health ($p<0.001$), vitality ($p<0.001$), social role functioning ($p<0.001$), and mental health ($p<0.001$), which are included in physical ($p<0.001$) and mental ($p<0.05$) health. HAQ had strong negative effects on physical functioning ($r=-0.56$, $p<0.001$), role function ($r=-0.33$, $p<0.001$), bodily pain ($r=-0,60$, $p<0.001$), general health ($r=-0,40$, $p=0.01$), vitality ($r=-0,46$, $p<0.001$), social functioning ($r=-0.48$, $p<0.001$) and mental health ($r=-0.42$, $p<0.001$). Articular long-term damages (JADI-A) have a predominantly negative effect on the patient's physical health ($r=-0,27$, $p<0,05$) and on the indices associated with it: physical functioning ($r=-0,24$, $p<0.05$), bodily pain ($r=-0.24$, $p<0.05$), general health ($r=-0.24$, $p<0.05$), vitality ($r=-0.19$, $p<0.05$), social functioning ($r=-0.27$,$p<0.05$), mental health ($r=-0.22$, $p<0.05$). While the extra-articular long-term damages – JADI-E also have a predominantly negative effect on the patient's physical health ($r=-0.22$, $p<0.05$) and on the indexes associated with it: physical functioning ($r=-0,28$,$p<0,05$), bodily pain ($r=-0,20$, $p<0,05$), general health ($r=-0,23$, $p<0,05$), mental health ($r=-0.23$, $p<0.05$), as well as a positive association with Beck depression scale ($r=0,28$, $p<0.05$) and PHQ-9 ($r=0.28$, $p<0.05$).
Conclusions: In our transitional cohort of Ukrainian patients at the era of biological therapies, juvenile idiopathic arthritis had a larger effect on the physical than mental SF-36 subscale. Pain was the main factor influencing quality of life. Extra-articular long-term JIA damages have impact on physical and mental health of young adults. Additional evaluation of mental health by PHQ-9 and Beck depression scale is recommended for evaluation signs of depression in Ukrainian young adults in transition period.

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AB1106 DEPRESSION AND ANXIETY IN PAEDIATRICSYSTEMIC LUPUS ERYTHEMATOSUS. A SYSTEMATIC REVIEW

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Background: Depression and anxiety are common and treatable childhood mental health disorders that have been reported to impact outcomes for individuals with childhood-onset Systemic Lupus Erythematosus (cSLE)[1]. Research into the prevalence of depression/anxiety and cSLE comorbidity has reported conflicting results, and to our knowledge, no previous review of these data has been undertaken.
Objectives: To synthesise current knowledge regarding the association of depression and anxiety disorders with cSLE among paediatric patients.
Methods: Studies were identified through a comprehensive search of MEDLINE, EMBASE, PsychINFO, LILACS and Web Of Science (from database inception – July 2017) using MESH headings and Keywords for 'lupus erythematosus', and 'depression' or 'anxiety'. Included studies measured depression and/or anxiety symptoms prospectively among children and youth 8 to 21 years of age with a diagnosis cSLE. Data were extracted by two independent coders and where discrepancies occurred, agreement was reached by consensus.
Results: Sixty-two studies met criteria for full text review, and of these, 13 studies were included in the final analysis. The majority (80%) of studies were of cross-sectional design, with sample sizes ranging from 14 to 100 (mean=47) participants. The mean age of participants was 15.6 years and participants were predominantly female. Prevalence rates for depression ranged from 6.7% to 54%. Anxiety symptom prevalence was 20% to 34%. All studies employed self-report instruments to assess depression and anxiety; none of the studies utilised semi-structured diagnostic interview to make psychiatric diagnoses. Significant heterogeneity precluded meta-analysis of the data.
Conclusions: Depression and anxiety may be common comorbidities of cSLE however current research is limited by a paucity of studies, small sample sizes and an inability to confirm psychiatric diagnoses. Future research addressing these limitations is needed.

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AB1107 TRANSITION FROM PAEDIATRIC TO ADULT RHEUMATOLOGY SERVICES IN NEWCASTLE TRUST HOSPITALS

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Background: Transitional care is defined as "the purposeful, planned movement of adolescents and young adults with chronic physical and medical conditions from child-centred to adult-oriented healthcare systems."[1]
In 2016, EULAR produced recommendations on how this transition process should occur within rheumatology.[2]
Objectives: To audit the Newcastle-Upon-Tyne Hospitals rheumatology transitional care service against EULAR recommendations.
Methods: EULAR recommendations were adapted into a questionnaire, which was reviewed and edited by several Young Adults (YAs) with Juvenile Idiopathic Arthritis (JIA) attending clinics.
Patients attending specialist YAJIA clinics between November 2017 and January 2018 were asked to complete the questionnaire; responses were audited against EULAR recommendations.
Results: 28 YAs with JIA completed the audit questionnaire (10 males and 18 females, age range 16–25). Not all questions were answered by all YAs.
Of the 17 patients with 'childhood-onset disease' (onset ≤age 10) 12% (2/17) had their transition started by age 14 ('essential' recommendation) and none by age 11 (the 'ideal' recommendation). Of the 11 patients with 'adolescent-onset disease' (onset >age 10) 18% (2/11) had their transition started at the time of diagnosis (EULAR recommendation 2).
63% (17/27) of patients had ≥1 'direct' contacts with adult and paediatric rheumatology via a joint appointment (EULAR recommendation 3).
11% (3/28) of patients were aware of a documented individual transition plan (EULAR recommendation 4).
40% (6/15) were able to list ≥3 multidisciplinary team (MDT) members that had positively impacted their care. 54% (15/28) were able to provide the name of a transition coordinator (EULAR recommendation 6).
81% (21/26) of respondents 'agreed or strongly agreed' that they had been signposted to information on their condition, 48% (12/25) to peer support groups, mentoring schemes and charities, and 25% (6/24) to information sources on careers and finance. 64% (of patients were consulted on how they would like their parent/carer to be involved in their care during and after their transition (EULAR recommendation 7).
82% (23/28) of respondents reported having copies of letters concerning their care and transition (EULAR recommendation 8).
Conclusions: Newcastle-upon-Tyne hospitals transition services are in line EULAR recommendations in terms of MDT involvement in the transition process and addressing the medical needs of patients during transition, whilst signposting them to other agencies and ensuring they have copies of communication.
The audit also identified areas for improvement including: the need for a single named coordinator for all patients; ensuring discussions with patients about the transition process begin at an earlier age, and making sure patients are aware of and able to contribute to their documented and individulised transition plan.

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AB1108 THE EFFICACY AND SAFETY OF TREATMENT OF 152 NON-SYSTEMIC JUVENILE IDIOPATHIC ARTHRITISPATIENTS WITH ETANERCEPT: FACTORS, ASSOCIATED WITH ACHIEVEMENT REMISSION AND RISK OF FLARE

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Background: Juvenile idiopathic arthritis (JIA) is the most frequent chronic inflammatory joint disease in childhood, required biologics administration if the previous therapy falls. Etanercept is the most worldwide biologic using in JIA patients.
Objectives: The aim of our study was to evaluate the efficacy and safety of etanercept in children with non-systemic categories of JIA and determine predictors of achievement of the remission and risk of flares.
Methods: In the retrospective observational study were included 152 children with non-systemic categories of JIA, treated with etanercept. Standard JIA measures and outcomes were utilised, remission was based on C. Wallace criteria (2004). We used descriptive statistics, χ^2-test, Fisher's exact test, Mac-Nemar test, Mann-Whitny, Wilcoxon, Friedmann and log-rank tests, AUC-ROC analysis, odds ratio and relative risk calculation with Cox regression models.
Results: The cumulative remission was achieved in 58.8% patients during the trial. The maximum remission rates (80%) were in children with treatment duration near the 5 years and increased from year to year. Patient who achieved remission had less JIA onset age (p=0.015), age of inclusion in the study (p=0.004) and age of etanercept administration (p=0.00007). The main predictors of achievement remission were JIA onset age ≤7.8 years (OR=4,3 (95%CI: 1.9–9.8), p=0.0003), age of inclusion in the study ≤14.0 years (OR=2.85 (95%CI: 1.4–5.9), p=0.00007), age of etanercept administration ≤10.0 years (OR=3.5 (95%CI: 1.7–7.2), p=0.0007), time before etanercept administration ≤2.4 years (OR=2.7 (95%CI: 1.3; 5.5), p=0.0007). In Cox regression model (p=0.007) HLA B27 positivity (RR=2.15 (95%CI: 0.98; 4.75), p=0.06) and time before etanercept administration ≤2.4 years (RR=2.4 (95%CI: 1.4; 4.4), p=0.003) were main predictors of remission achievement. Polyarticular JIA increased the risk of flare compare to oligoarticular (RR=2.7 (95%CI: 0.9; 8.2), p=0.08), then concomitant methotrexate decresed the risk of flare (RR=0.32 (0.1; 1.15), p=0.05) in Cox regression model. During the