levels or due to a better response to antiTNF of those patient with higher plasma levels of 25 hydroxy-vitamin D.

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AB0969 IMPACT OF JUVENILE IDIOPATHIC ARTHRITIS ON SCHOOLING

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Background: Juvenile idiopathic arthritis (JIA) is the most common chronic rheumatic disease in children and is one of the major causes of morbidity and physical disability.Due to frequent absences,children with chronic health impairments are also often confronted with educational difficulties.
Objectives: The aims of this study were to assess the impact of JIA on children's schooling and to determine the factors that influence their school level.
Methods: This is a cross-sectional study including patients with JIA (ILAR criteria).A detailed questionnaire was completed for each participant by interviewing them or their parents as well as by information obtained from their medical records.Collected data included age,sex,subtype of JIA,disease duration,level of disability according to the Childhood Heath Assessment Questionnaire (CHAQ),visual analog scale for patient's overall assessment of disease activity,duration of morning stiffness, joint counts,erythrocyte sedimentation rate,C-Reactive Protein,Disease Activity Score (DAS28).Medications used for JIA treatment were also documented.
Data on the school performance of patients and their siblings were obtained using telephone interviews (educational level, absenteeism, school delay by repetition, drop-out).
The comparison of quantitative variables was performed with the Mann-Whitney test and the comparison of qualitative ones was performed with the Chi square test. The significance level was set at 0.05.
Results: A total of 38 patients with JIA were included, 23 female and 15 male, with a mean age of 26 years [12–51] and a mean disease duration of 237 months [5–496].The average age of the onset of the disease was 7.4 years [1.5–16].
The most common subtype was rheumatoid factor-positive polyarthritis (n=16) followed by systematic (n=7), oligoarticular (n=6), rheumatoid factor-negative polyarthritis (n=5) and Enthesitis-related arthritis (n=4). The mean DAS28 was 2.63 [0.76 - 5.55] and the median CHAQ was 0.528 [0–3].Twenty-seven of the children were receiving corticosteroid. Disease-modifying anti-rheumatic drugs were used by 34 of the 38 patients: methotrexate (n=23), sulfasalazine (n=8),leflunomide (n=7),biotherapies (n=14). Twenty patients had complications:*Hip arthritis (n=15),* growth stunting (n=12), uveitis (n=4). Joint replacement was required in 9 cases.
Four patients were illiterate,12 had dropped out of school,21 reported repeated absences due to illness.A year of schooling was repeated by 61.7% of patients.Ten out of 32 patients over the age of 20 had an university level.Almost 80% of patients were exempted of physical education.
There were no significant associations between the school-related problems,the socio-demographic characteristics and the various parameters of clinical and biological activity studied.School failure was similar among patients and their siblings (p=0.05).
Conclusions: Our study suggested that JIA negatively affects schooling of children.More studies with a larger sample are needed to identify the variables associated with school failure in order to ensure the proper management of these patients and to increase their academic performance.

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AB0970 CLINICAL AND IMMUNOLOGICAL CHARACTERISTICS IN CHILDHOOD-ONSET SYSTEMIC LUPUS ERYTHEMATOSUS PATIENTS TREATED WITH RITUXIMAB

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Background: Systemic lupus erythematosus (SLE) is an autoimmune disease that is more severe in pediatric population than in adults. Biological therapy with anti-CD20 (rituximab) is an option in patient that do not respond to conventional therapy.
Objectives: The aim of this study is to determine the clinical and immunological response in 9 patients with childhood-onset systemic lupus erythematosus (cSLE) that received treatment with rituximab in a third level hospital
Methods: This is a retrospective observational study. 9 patients treated with Rituximab between November 2007 and October 2016 were included and their medical records were reviewed. The response to treatment at 6 months and one year after the first infusion of Rituximab were assessed. Patients with overlap syndromes were excluded. All patients fulfilled four or more of the 1982 revised American College of Rheumatology criteria for the diagnosis of SLE (<16 years).
Results: Nine pediatric patients with SLE treated with rituximab were included, all of them were female. The age at diagnosis of SLE was a mean of 15,22 years. The mean time duration of disease was 87,55months (5–255m). 7 patients were caucasians. Rituximab was indicated in 6 patients with class IV of lupus nephritis (LN) 1/9 with class III LN, 1/9 with severe cutaneous lupus, and with severe hematological manifestations in 1 case (haemolytic anemia). In addition, 6/9 patients had mucocutaneous and articular manifestations. The disease activity of all patients was assessed using SELENA-SLEDAI index pre rituximab infusion, the mean was
17,11 (8–33). All patients had low level of complement C3 and C4 and 8/9 increased anti-DNA. In 8/9 patients Rituximab was used as a rescue treatment and in a single case as a first line.
3/6 patients with renal involvement were previously treated with cyclophosphamide (CF) iv and mycophenolate, 2/6 CF. In case of cutaneous involvement the previous treatment was methotrexate, azathioprine (AZA) and dapsone and in case of hemolytic anemia was AZA.
The treatment protocol was 1 gram x 2 (1 cycle) in 7/9 patients, 375mg/m^2 x 4 in 1/9 cases and 600mg monthly for 5 months in the case of hemolytic anemia. Five patients received more than 1 cycle. After the administration of Rituximab, the SELENA-SLEDAI activity index was 4.5 points. At 6 months a complete response was obtained in the case of hematological and cutaneous manifestations, in 2 cases of lupus nephritis (proteinuria <0.5 g/day) and partial response was obtained in 2 cases. Data were not analyzed in 2 patients (death and less than 6 months of the first dose of rituximab). Patients with partial response and lack of response achieved complete response at 12 months. 2/9 patients had side effects (Rituximab pneumonitis in 1 case and infections in 2 cases). Mortality was 11.11% (1/9 patients, per infection and lupus activity, SLEDAI pre rituximab =33)
Conclusions: In our study, although it consisted of few patients, it was objected that Rituximab therapy in patients with cSLE is effective, reduces lupus activity index, especially in cases of renal, cutaneous and hematologic involvement, that don't respond to conventional therapy. It may be consider in the future as an effective alternative treatment at first line treatment.

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AB0971 CHARACTERISATION OF THE PATIENTS AT THE TIME OF THE TRANSITION INTO THE ADULT RHEUMATOLOGY. 63% OF THE PATIENTS ARE IN REMISSION AT THE TIME OF THE TRANSITION

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Background: The issue of transition from pediatric to adult rheumatology service is an emerging important topic. In 2012 a transition clinic was established according to the "Berliner Transitionsprogramm" [1] in cooperation of the Hamburger Zentrum für Kinder-und Jugendrheumatologie and the Rheumatology Unit of the Marien Hospital in Hamburg. The Berliner programm suggests three visits of the patient in transitionprocess, in the presence of the pedriatic and an adult rheumatologist together, and the fourth visit conducted by the adult rheumatologist alone. We present the characteristics of the patients at the time of the 4th visit.
Objectives: To characterize the patient population at the time of enrollment into the adult service in the frame of our transition programm.
Methods: We collected patient data starting 8/2012 to 11/2016. We summarized the patient population, who succesfully transitioned from pediatric to adult rheumatologic service, concerning diagnosis, sex, age at the time of diagnosis, disease duration at the time of transition, JADAS, HAQ, VAS globular assessment, VAS pain, medication and disease activity.
Results: 73 patients were transitioned. 65% of them female. Mean age at diagnosis of the patients was 12.5 years. Mean disease duration at time of transition ws 10.8 years. The mean JADAS Score was 3.18 and the mean HAQ Score was 0.136. The patients global activity score was, on a VAS of 0 to 100, 14.03 and the global pain score, on a VAS of 0 to 100, 12.33. 39.7% of the patients received synthetic DMARDS and 34% biologic DMARDS: Only 1 patient received steroids. 24.6% of the patients were off medication. 63% of the patients were in remission, 61% of them on medication and 39% off medication in remission.
Conclusions: In this monocenter cohort 63% of patients were in remission, and with the mean JADAS Score of 3.18 most of them have low disease activity under the current treatment. The mean HAQ Score with 0.136 reflects a score, which is expected in healthy controls. But 75.4% of the patients needed medication to

achieve this good condition. The amount of patients on medication emphasize the need of the structured transition.

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AB0972 CHILDHOOD ONSET STROKE AND VASCULITIS ASSOCIATED WITH DEFICIENCY OF ADENOSINE DEAMINASE 2 (DADA2)

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Background: The deficiency of Adenosine Deaminase 2 (DADA2) is a rare autosomal recessive condition resulting from mutations in CECR1 (Cat Eye Syndrome Chromosome Region 1) gene, mapped to chromosome 22q11.1. It is a type of autoinflammatory disease, mainly characterised by early-onset polyarteritis, haemorrhagic, ischemic strokes and hypogammaglobulinemia (1). We report a case of 7-year boy presenting with haemorrhagic stroke and vasculitis responding to immunosuppression with Anti-TNF drug.

Objectives: A 7 year boy presented to the emergency department with reduced consciousness, headaches and nose bleeds. Initial imaging showed an intraparenchymal haemorrhage requiring frontal craniotomy and evacuation of the haematoma. This acute presentation was preceded by a history of recurrent fevers, weight loss, testicular pain, erythema nodosum and tender lymph nodes.

Methods: The laboratory findings revealed anaemia, ESR of up to 61mm/h, C-reactive protein 100, normal immunoglobulins, positive Anti nuclear antibody (ANA), mildly raised antibodies to double stranded DNA (dsDNA) and Proteinase 3 antibody. Skin biopsy confirmed panniculitis. CT imaging and angiography of the head at the time of acute presentation showed intraparenchymal haemorrhage and aneurysm of the left middle cerebral artery. Further CT angiography of the whole body revealed renal and liver microaneurysms. A provisional diagnosis of Polyarteris nodosa was made and started on steroids and cyclophosphamide. He had further genetic testing, showing mutation in CECR1, leading to Adenosine deaminase 2 deficiency. Patient responded to cyclophosphamide induction regime, which was followed by Etanercept.

Results: We report a case of ADA2 deficiency presenting initially with features of an autoinflammatory disorder, complicated by acute stroke secondary to haemorrhage. Our patient exhibited most of the clinical symptoms previously reported in ADA2 deficiency, including its association with polyarteritis nodosa (2). Although, he did not exhibit hypogammaglobulinaemia (3) which has been reported, interestingly, he was positive for markers of autoimmunity (ANA, ANCA) (4). It has been reported that treatment with anti-TNF and IL-6 (5) could lead to improvement, and our patients initial response to cyclophosphamide was excellent, followed by continued treatment with Etanercept.

Conclusions: Screening for adenosine deaminase 2 deficiency should be considered in all children presenting with neurological symptoms and features of vasculitis.

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AB0973 ANTIPHOSPHOLIPID ANTIBODIES IN CHILDREN WITH SYSTEMIC LUPUS ERYTHMATOSUS AND JUVENILE IDIOPATHIC ARTHRITIS

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Background: Antiphospholipid antibodies (aPL) are a family of autoantibodies that present in a small percentage of the population but occur more commonly in patients with Antiphospholipid syndrome (APS) and Systemic lupus erythematosus (SLE).

Objectives: We aimed to determine of the level of anti-β2-glycoprotein I (anti-β2GPI) (IgG and IgM) isotypes and anticardiolipin antibodies (aCL) as inflammatory markers in children with SLE and Juvenile Idiopathic Arthritis (JIA) and figure out their relation to the clinical manifestations and activity of the disease.

Methods: This prospective study included one hundred twenty children, sixty five having SLE and fifty five having JIA, their ages range between 4.5 – 16 years (37 males and 83 females).In addition, twenty apparently healthy children of comparable age, sex and nutritional status were used as a control group. All patients and normal controls were subjected to full clinical and laboratory investigations included aCL and anti-β2GPI level (IgG and IgM) measured by a standardized ELISA.

Results: IgG isotype of anti-β2GPI was found to be positive in 27.7% and 14.5% for SLE and JIA groups respectively. However IgM isotype of anti-β2GPI was found to be positive in 24.6% and 7.25% for SLE and JIA groups respectively. The mean levels of both IgG and IgM isotypes of anti-β2GPI were found to be significantly increased in comparison to controls ($P<0.001$) in both SLE and JIA groups. IgG isotype of aCL was found to be positive in 23.1% and 18.2% for SLE and JIA groups respectively. However IgM isotype of aCL was found to be positive in 18.5% and 18.2% for SLE and JIA groups respectively. A significant positive correlation was found between IgM and IgG isotypes of anti-β2GPI and with their corresponding class of aCL in both SLE and JIA groups. A significant positive correlation was found between the elevation of anti-β2GPI (IgG) and thrombocytopenia together with neuropsychiatric disease in SLE. While in JIA elevation of anti-β2GPI was found to be correlated only with elevation of aCL irrespective to clinical or laboratory data.

Conclusions: The study reported a higher prevalence of aPL in children with SLE and JIA. Elevated levels of anti-β2GPI (IgG) correlated with thrombocytopenia together with neuropsychiatric disease in SLE.

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AB0974 ETANERCEPT TREATMENT FOR A PATIENT WITH REFRACTORY MACROPHAGE ACTIVATION SYNDROME IN JUVENILE SYSTEMIC LUPUS ERYTHEMATOUS

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Background: Macrophage activation syndrome (MAS) is a serious, potentially fatal complication of childhood systemic inflammatory disorders, and it is most frequent in Systemic Juvenile Idiopthic Arthritis, for instance, it is increasingly reported in other pediatric rheumatic diseases as lupus erythematosus and Kawasaki disease.

Objectives: To describe clinical case report of a 16 year old girl with Juvenil Systemic Lupus Erythematosus and Macrophage Activation Syndrome refractory.

Methods: A 16-year-old woman with recent diagnosis of Systemic Lupus Erythematosus in November 2015. At admission with continuos fever lasting 2 months, with initial laboratory studies with Triglycerides 395 mg/dl, Ferritin 3.300 ug/l, soluble receptor IL-2 2.838 U/ml, fibrinogen 166 mg/dl, Hemophagocytos in bone marrow, presence of persistent cytopenias. Initial management with methylprednisolone 30 mg/kg/day (3 days) without clinical response was initiated. Management is added to Cyclosporin A (10 mg/kg/day), reporting subtherapeutic serum levels despite high doses without clinical response and improvement of laboratory controls. During the hospital stay complete 13 weeks of treatment with Etposide (180 mg/dose), with ferritin levels in 4,180 ug/l, triglycerides 438, Fibrinogen 455 mg/dl, WBC 5,000 ul^{-1} (3,400 neutrophils, lymphocytes 1,050), platelet count 78 x 103 ml^{-1}, hemoglobin 7.2 g/dl. After three months of treatment, she was given with Etanercept 0.4mg/kg/dose, 2 times a day. Currently in week 3 of treatment with WBC 6,500 ml^{-1}, hemoglobin 9.7 g/dl, platelet count 140 x $103ml^{-1}$ and ferritin 4,270 ng/ml. The patient remains disease with costicosteroid, cyclosporine, and etanercept, without adverse events.

Results: Patient treated with Etanercept during 5 weeks, presenting clinical response for MAS.

Conclusions: A general therapeutic protocol for MAS is not available: first line treatment is usually represented by parenteral administration of high dose corticosteroids. Mild forms are reported to respond to steroids alone in association with supportive medicaments. Steroid-resistant cases or the most severe forms of MAS require the addition of cyclosporine A, other therapeutic regimens have been studied such as high-dose intravenous immunoglobulins, antithymocyte globulins, etanercept, etoposide and plasmapheresis

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