AB0966 THE CLINICAL OBSERVATION OF IMMUNOADSORPTION IN TREATMENT OF CHILDREN WITH REFRACTORY AUTOIMMUNE DISEASES

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Objectives: Explore the clinical efficacy and safety of the immunoadsorption assisted treatment of children with refractory autoimmune diseases.
Methods: Use HA280 type resin perfusion machine for four times of whole blood immunoadsorption treatment for one case of a 4-year-old child combined with severe dermatomyositis and pulmonary infection, one case of 9-year-old child with severe allergic purpura combined with gastrointestinal bleeding, intestinal perforation, after intestinal tract partial hepatectomy, one case of 11-year-old child with systemic juvenile idiopathic arthritis combined with head and facial cellulites and macrophage activation syndrome. Observe the improvement of its clinical manifestations, serum immunoglobulin, complement, liver and kidney function, myocardial enzymes, autoantibodies.
Results: The children's signs and symptoms improved significantly after treatment of whole blood immunoadsorption, the sensitivity of the adrenal cortex hormones increased, liver and kidney function, myocardial enzymes improved, the levels of serum IGG descended, complement (C3, C4) rose, anti-cyclic citrullinated peptide (CCP) was negative.
Conclusions: The whole blood perfusion immunity adsorption treatment is able to to reduce blood plasma IGG, improvement complement C3 and C4 level, eliminates anti-CCP immune body in vivo, reduce the cardiac muscle zymogram and the liver enzyme in a short time, increase sensitivity to the adrenal cortex hormone, alleviate immunological disease's symptom in active stage, have the affirmative function to make the baby pass the dangerous period safely. The child whole blood fills the class immunity adsorption not to present the untoward effect. No untoward effect happened.
Disclosure of Interest: None declared
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AB0967 CLINICAL, LABORATORY PROFILES AND LONG-TERM OUTCOME OF JUVENILE CUTANEOUS PAN: A SINGLE CENTER EXPERIENCE

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Background: Cutaneous polyarteritis nodosa (cPAN) is an immune complex-mediated rare disease that affects small and medium sized vessels in the dermis and subcutaneous tissue.
The clinical course is characterized by periodic exacerbations and remissions that may persist for many years. Most patients respond to NSAIDs and glucocorticoids (GC), whereas some may require DMARDs and/or immunomodulatory therapy.
Objectives: To describe the diferrent clinical patterns, laboratory findings and long term outcomes of juvenile cPAN in a tertiary care hospital.
Methods: Retrospective observational study, including all patients diagnosed with cPAN between 2002–2016. Diagnosis relied on clinical features confirmed by histological study. Recorded data included clinical features, laboratory results and long-term outcomes.
Results: 10 children were included (7 female), mean age at onset was 9.9 years (r:4.1–16.3). Delay from symptoms onset to biopsy confirmed diagnosis was 2±2.3 months; 4 patients underwent a second biopsy due to inconclusive results in the first performed.
Clinical features included cutaneous (100%) and osteomuscular involvement (50%), fever (40%), neuropathy (10%) and weight loss (10%). Reported cutaneous symptoms were 8 patients with nodules, 4 livedo, 4 purpura, 1 ulcer and 1 necrosis. Most lesions were localized in the lower limbs (8), even though it was also reported in upper limbs (3) and trunk (3). Most cases exhibited raised CRP, ESR and leukocyte count with a mean of 26.4±47.9 (mg/L), 27.7±29.7 (mm/h), and 8.6±5.3 (x 10^9/L) respectively.
As first line therapy, all patients received GC and 6 NSAIDs. 8 were given a DMARD such as HCQ or MTX. Due to persistent activity or relapse, rescue treatment with pulse-GC (20%), MMF (10%) or IVIG (20%) was instituted. Only 1 patient received penicillin prophylaxis due to relapses associated with streptococcal infection.
Mean follow-up was 3.9 years (r: 1.1–10.4). 4 patients had a monophasic disease, six suffered ≥2 relapses. At last follow-up 9 patients were on remission, even though 3 were off-therapy. No complications were reported.
Conclusions: Clinical and laboratory findings in our series was similar to previous reports. However, our patients presented a greater number of relapses and DMARDs requirement.
Disclosure of Interest: None declared
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AB0968 HYPOVITAMINOSIS D IN JUVENILE IDIOPATHIC ARTHRITIS: PREVALENCE AND RELATED FACTORS

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Background: 25hydroxy-vitamin D not only plays a key role in calcium homeostasis, but also has antiinflammatory and immunomodulatory properties. Hypovitaminosis D prevalence in children suffering from Juvenile Idiopathic Arthritis (JIA) ranges from 6% to 30% according to different publications.
Objectives: Evaluate hypovitaminosis D prevalence in JIA pediatric patients in Spain and assess involved factors.
Methods: Observational cross-sectional study in JIA Spanish patients from 4 to 15 years, monitored by a Pediatric Rheumatology Unit. Monoarticular forms and patients with other chronic diseases or receiving different treatments from those indicated for JIA were excluded.
Anthropometric, clinical and treatment data were recorded. Bone metabolism parameters and validated diet (KIDMED) and exercise (PAQ-C/PAQ-A) questionnaires were obtained.
Hypovitaminosis D was defined as 25hydroxy-vitamin D plasma levels lower than 30 ng/ml
Results: 76 children participated. Their characteristics are included in table 1.
The population's prevalence estimation of hypovytaminosis D in children with JIA was 16 - 35% (CI 95%).
We found no relationship between 25 hydroxy-vitamin D levels and sex, JIA subtype neither duration or dose of systemic glucocorticoids.
In bivariate analysis we found direct association between hypovitaminosis D and Body Mass Index percentile (BMIp) (p=0,05), received dose of prednisone (p=0,03) and clinical activity duration (p=0,04); and an inverse relationship with physical activity level (p=0,04).
In multivariate analysis, relationship between hypovitaminosis D and BMIp (B 0,024; p 0,016) and with disease activity (B 0,015; p 0,01) were maintained. Moreover, we found an inverse association with biological disease-modifiying antirheumatic drugs (B-4,69; p 0,048), specifically with anti-tumoral necrosis factor α (antiTNFα) (B -4,7; p 0,042)

PATIENTS' CHARACTERITICS (N=76)		
Gender (Male), n (%)		23 (30,3)
Age (years), median (IR)		10,83 (8,52-13,54)
25OH-Vitamin D (ng/mL), mean (+/-SD)		34,04 ng/mL (8,90ng/mL)
DISEASE CHARACTERISTICS (N=76)		
JIA subtype, n (%)	Systemic	9 (11,8)
	Persistent oligoarticular	35 (46,1)
	Extended oligoarticular	12 (15,8)
	Positive RF polyarticular	1 (1,3)
	Negative RF Polyarticular	19 (25,0)
Duration disease (years), median (IR)		6,55 (3,29-9,45)
Inflammatory activity duration (days), median (IR)		385,0(246,75-761,25)
RECEIVED TREATMENTS (N=76)*		
Systemic GC, n (%)		61 (80,2)
GC treatment duration (days), median (IR) *(n=84)		125,00 (66,00-179,00)
Mean GC dose (mg/kg/day), median (IR)		0,276 (0,169-0,457)
Synthetic DMARDs treatment, n (%)		39 (51,3)
Biological DMARDs treatment,, n (%)		21(27,6)
Biological DMARDs treatment subtype, n(%)	Anti-TNFα	15 (19,7)
	Anti-IL1	4 (5,2)
	Anti-IL6	2 (2,6)

JIA: Juvenile idiopathic arthritis; RF: Rheumatic factor; GC: glucocorticoids; DMARD: Disease-modifying antirheumatic drug; Anti- TNFα: anti- tumoral necrosis factor α;Anti-IL1: anti-interleukin 1; Anti-IL6: anti-interleukin 6

Conclusions: Hypovitaminosis D prevalence in our population is similar to previously described.
JIA patients with higher BMIp have more hypovitaminosis D, as it has been reported in other inflammatory diseases.
A direct relationship exists between inflammatory activity and vitamin D, but we need more studies to asses if one is cause or consecuence of the other.
Patients treated with antiTNF have better plasma levels of 25 hydroxy-vitamin D, this can be explained because these drugs may increase 25 hydroxy-vitamin D

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