Disclosure of Interests: None declared

Conclusion: Our study demonstrated that RTX is highly effective in children with RD, the majority with SLE, but the safety data obtained indicate the need for careful monitoring of therapy, primarily taking into account the frequency of infections. A decrease in IgG level was observed in a small proportion of pts and did not correlate with the incidence of infections. The frequency of serious infections was low.

Disclosure of Interests: None declared

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SAT0495
LONG TERM OUTCOME OF JUVENILE IDIOPATHIC ARTHRITIS IN ADULTHOOD: THE MONOCENTRIC EXPERIENCE OF 520 PATIENTS FOLLOWED FOR 20 YEARS IN A TRANSITION TERTIARY CLINIC OF PEDIATRIC RHEUMATOLOGY.

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Background: Juvenile Idiopathic Arthritis (JIA) is a chronic pediatric inflammatory disease that shows many differences compared to adult-onset arthritis. The different clinical manifestations, the assessment and the management of JIA is the reason that the transition from childhood to adulthood is an important multi-dimensional process that emphasizes a lot of aspects.

Objectives: To describe the long-term outcome of JIA.

Methods: Five-hundred and twenty patients affected by JIA and referred to a transition care rheumatology tertiary centre were considered between 1999 and 2019. The outcome assessment included remission, disease duration, medica-
tions, number of prosthetic implantation, pregnancies, mortality and social inte-
gration (employment status and educational level).

Results: A hundred and thirty-eight (26%) males and 382 (73%) females were included; 157 (30%) patients were lost to follow up. The mean age of the patients was 27 (18-57) years, with a mean age at onset of 8 years and an average disease duration of 19 years. Subtypes of JIA at disease onset included 282 (48%) oligoarthritis, 134 (26%) polyarthritis, 64 (12%) systemic arthritis, 22 (4%) psoriatic arthritis, 43 (8%) enthesitis related arthritis and 1 (0.1%) undifferentiated arthritis. Ninety-three (18%) patients suffered of uveitis. Ninety-five implant prosthesis and 16 arthrodeses were recorded. At follow up 198 (38%) patients were on remission of which 107 (20%) off medication. Among the 322 patients still on medication, 84 (16%) were under treatment for oral steroids, 226 (43%) with sDMARDs and 249 (40%) with bDMARDs. Five deaths (1%) occurred in this cohort. Two hundred and thirty-five subjects had a higher educational level, 327 had an employment. We have data of twenty-nine pregnancies. The transition age was considered after the age of 16 years old. The key word for the management of this cohort was the multi-disciplinary approach towards each patient, with the collaboration of other specialists (ophthalmologist, orthopedic, dermatologist, gastroenterologist, obstetric and psychologist).

Conclusion: In the era of biologic therapy the long-term outcome of JIA under-
went an outstanding improvement regarding a lot of variables. Two hundred and thirty-two patients were still followed, not only because of the continuation of the biological therapy, but also for a multidisciplinary care even during remission. JIA often persists over the adulthood, therefore the long term follow-up and care of these patients needs to be conducted by a rheumatologist expertized in JIA in collaboration with other specialists.

Disclosure of Interests: None declared

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SAT0497
A PILOT PROTEOMIC ANALYSIS OF PLASMA BIOMARKERS IN IGA VASCULITIS

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Background: IgA vasculitis/ Henoch Schönlein Purpura (IgAV/HSP) is the most common vasculitis of childhood, characterized by the IgA1 immune deposits in the small vessels. Although it is very common, the understanding of its molecular pathogenesis is still very limited.

Objectives: We aimed to analyse plasma proteomes of IgAV/HSP patients using nano liquid chromatography – tandem mass spectrometry (nLC-MS/MS) to investigate the disease pathogenesis.

Methods: IgAV/HSP was diagnosed according to the Ankara criteria in 2008 (1). Five active IgAV/HSP patients and two age and gender-matched healthy controls were enrolled in this pilot study. Serum samples from subjects were collected on the same day of IgAV/HSP diagnosis and before steroid or other immunosuppressive treatment initiated. Sample preparation was carried out using PreOomics IST Kit. We investigated the alteration of serum proteome using the nano LC-MS/MS approach. Bruker raw files were analyzed using the proteomics software Max Quant (1.6.7.0). The human reference proteome set from UniProt was used to identify proteins. Proteomic data were analyzed with Perseus 1.6.7.0.

Results: The data file includes peptide and protein identification, accession numbers, protein and gene names, sequence coverage and label free quantification (LFQ) values of each sample. 345 proteins were reported per sample. Identiﬁcations from the reverse decoy database, identiﬁed by site only and known contaminants were excluded. Data were log transformed. Two sample T-test was performed between groups. We identiﬁed 23 signiﬁcantly different expressed proteins (Table 1). Mainly the differentially expressed proteins were in the Ig and complement pathway, innate immune inflammatory,
and were among the structural cytoskeletal filaments. The levels of Complement C3, Apolipoprotein E, Glyceraldehyde-3-phosphate dehydrogenase, Filamin-A, Alpha-1B-glycoprotein, Tubulin beta-1 chain, Lipopolysaccharide-binding protein, Ig mu chain C region were significantly higher in IGAV patients.

Table 1. List of differentially expressed proteins identified in IgAV compared to healthy controls

<table>
<thead>
<tr>
<th>Majority protein IDs</th>
<th>Protein names</th>
<th>Gene names</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>O75822</td>
<td>Eukaryotic translation initiation factor 3 subunit J</td>
<td>EIF3J</td>
<td>0.004</td>
</tr>
<tr>
<td>P05106; H3BM2; P027847; A0A0J9YX35; A2JN7V5; A0A0J6BGWS2; P02679; A0A0C4D3H36; P01D03; P01768; O14791; P01024; P02675; P02180; A0A0J5SD7B; P02649; P04406; A0A075B6R2; A0A0B7W6W49; P21333; P04217; P02790; A0A0C4D2H25; G1H4B7; P18428; P01871</td>
<td></td>
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</tr>
</tbody>
</table>

Conclusion: This pilot proteomic study may provide us a perspective in the pathogenesis of IgAV (HSP).

References:

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Background: ERA is the most common Juvenile Idiopathic Arthritis (JIA) subtype in Singapore (1), but less common in the West. Clinical characteristics and treatment of ERA in the region is not well-described thus impede the diagnosis and management plan which could lead to poorer outcomes.

Objectives: To describe the clinical characteristics, joint manifestation and treatment of ERA in a large monocentric cohort in Singapore over 10-year period.

Methods: Children diagnosed with ERA according to ILAR criteria with a minimum follow-up of 3-month duration were recruited from our registry, from 2009 to 2019, at KK Women’s and Children’s Hospital, Singapore. Nonparametric descriptive statistics including median (IQR) were used to described data. Kaplan-Meier survival analyses were used to estimate the probability of ever sacroiliitis development. Multivariate logistic and Cox regression analyses were used to determine predictors as appropriate. The significant level was set at < 0.05.

Results: A cohort of 147 ERA out of 439 JIA patients (male 88%; Chinese 80%) were included. Median age at onset was 11.9 yrs (IQR: 9.4-14.0) and disease duration was 6.0 yrs (3.1-8.9). Median lag period was 2.9 mo (1.2-7.4). Family history of HLA-B27 related diseases was positive at 8%. Acute uveitis occurred only 3%. Joint distribution at diagnosis and cumulative involvement were shown in Fig 1. Hip, sacroiliac and knee were the three most common joints involved. 24% presented with enthesitis and Achilles tendon enthesis were the most common. Majority presented with pauciarticular (84%), while 12% of patients had no peripheral joint involvement. 40% of patients presented with sacroiliitis (SIs) with 59% had bilateral involvement. Median duration to develop SIs was 76 mo (IQR 2.0-26.9). Probability of SIs development was 36%, 55% and 70% at 1, 5 and 10 yrs after onset, respectively. Interestingly, neg HLA-B27, female and older age at onset predicted SIs (p=0.001-0.044). Hip arthritis increased (p=0.043) but tarsitis decreased (p=0.031) the risk of SIs. Again, female, hip arthritis at diagnosis and neg HLA-B27 had a shorter time to SIs (p=0.004-0.007). Fig 2 showed medication used in our ERA cohort. Methotrexate (MTX) remained the most common DMARD used. However, 76% required anti-TNF therapy (aTNF) due to MTX failure. For SIs patients, 86% were on MTX but 85% of these, as compared to patients without axial disease, 60%, failed MTX. Only 10% of patients had aTNF without MTX.

Fig 1. Proportion of joint involvement at onset and cumulative involvement during the course of disease (%)

Fig 2. Proportion of medications used in ERA cohort during the course of disease (%)