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.

Table1. List of differentially expressed proteins identified in IgAV compared to healthy controls

Majority protein IDs	Protein names	Gene names	p-value
O75822	Eukaryotic translation initiation factor 3 subunit J	EIF3J	0,004
P05106; H3BM21	Integrin beta-3	ITGB3	0,008
P02743	Serum amyloid P-component	APCS	0,015
A0A0J9YX35	unknown	IGKV A18;	0,015
A2NJV5;	unknown	IGKV2D-29	0,021
A0A075B6S2			
P02679	Fibrinogen gamma chain	FGG	0,022
A0A0C4DH36	unknown	IGHV3-38	0,024
P0DP03;	Ig heavy chain V-III region CAM		0,024
P01768			
O14791	Apolipoprotein L1	APOL1	0,025
P01024	Complement C3	C3	0,030
P02675	Fibrinogen beta chain; Fibrinopeptide B	FBG	0,030
P01860	Ig gamma-3 chain C region	IGHG3	0,031
A0A075B7D8	unknown	IGHV3OR15-7	0,034
P02649	Apolipoprotein E	APOE	0,035
P04406	Glyceraldehyde-3-phosphate dehydrogenase	GAPDH	0,037
A0A075B6R2;	unknown	IGHV4-4	0,039
A0A087VW49			
P21333	Filamin-A	FLNA	0,043
P04217	Alpha-1B-glycoprotein	A1BG	0,045
P02790	Hemopexin	HPX	0,046
A0A0C4DH25	unknown	IGKV3D-20	0,047
Q9H4B7	Tubulin beta-1 chain	TUBB1	0,049
P18428	Lipopolysaccharide-binding protein	LBP	0,049
P01871	Ig mu chain C region	IGHM	0,050

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

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Paediatric rheumatology

SAT0498

ENTHESITIS-RELATED ARTHRITIS (ERA) IN SOUTHEAST ASIA: A DECADE OF SINGAPORE EXPERIENCE

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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 impeding 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 describe 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 (IQR9.4-14.0) 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 enthesitis were the most common. Majority presented with pauciartthritis (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 7.6 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 (%)

Conclusion: Our ERA cohort had less uveitis and family history of HLA-B27 associated diseases, but comparable gender and age at onset as compared to reports elsewhere(2). Up to 40% of our patients presented with SIs and/or enthesitis. Majority of SIs developed within the first 5 yrs (88%) for which over one-half developed within the first year. When considering only ERA patients, interestingly that female, neg HLA-B27 and older age increased risk of SIs development. 77% of patients were treated with MTX, but 76% of the patients required aTNF later. As for SIs, concurred with adult AS data, 85% failed MTX. About one-half of non-axial disease patients failed MTX which is less response rate as compared to other JIA subtypes.

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SAT0499 PREVALENCE OF ULTRASOUND FINDINGS SUGGESTIVE OF INFLAMMATORY ARTHRITIS IN CHILDREN WITH SKIN PSORIASIS (CHILDECHOPSO)

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Background: The prevalence of psoriasis (PsO) in children is estimated between 0.5-1% (1), and can be associated with musculoskeletal involvement, although the prevalence and typology of such involvement is unknown. (2) Over the last years ultrasound associated with Doppler (PDUS) has become an important tool for evaluating joint involvement in children with Juvenile Inflammatory arthritis (JIA). Several studies have shown the high sensitivity of this technique for detecting joint involvement, as well as high acceptability, due to the lack of radiation or of sedation (3).

Objectives: To evaluate the prevalence of joints and entheses involvement in children affected by PsO.

Methods: Observational cross-sectional study aiming at evaluating 150 consecutive children (≤ 16 years) affected by skin PsO and addressed by a dermatologist. For each child a standardized clinical and ultrasound evaluation of joint and entheses is performed at the following bilateral sites: a) Entheses: quadriceps tendon, proximal and distal patellar ligament, Achilles tendon, plantar fascia and extensor elbow tendon), b) joints: metacarpophalangeal, proximal and distal interphalangeal, wrist, elbow, knee, ankle, and metatarsophalangeal. PDUS evaluation is performed by an independent examiner, blinded to the clinical assessment of each subject.

Results: 41 patients were included until now, where 9 patients presented some kind of symptom (painful joint or enthesitis), 24 patients have family history of psoriasis and none of them had family history of psoriatic arthritis. Demographic and clinical characteristics are shown in table 1, whilst PDUS findings in Table 2.

Table 1. Demographic and clinical characteristics:

	Total (n=41)	Asymptomatic (n=32)	Symptomatic (n=9)	p
Males	22 (53.7%)	18 (56.3%)	4 (44.4%)	NS
Age	9.4 $\pm$ 3.9	8.91 $\pm$ 3.5	11.3 $\pm$ 4.6	NS
PsO duration (years)	3.5 $\pm$ 3.4	2.9 $\pm$ 2.9	5.6 $\pm$ 4.5	<0.05
PASI	5 $\pm$ 4.3	4.8 $\pm$ 4.5	5.7 $\pm$ 3.56	NS
BSA	4.3 $\pm$ 3.9	4.47 $\pm$ 4.3	4.1 $\pm$ 2.26	NS
Nail involvement	25 (61%)	19 (59.4%)	6 (66.7%)	NS
Plaques psoriasis	20 (48.8%)	14 (43.8%)	6 (66.7%)	NS
TJC ≥ 1 , n/tot (%)	9 (22%)	0 (0)	9 (100%)	<0.001
SJC ≥ 1 , n/tot (%)	2 (4.9%)	0 (0)	2 (22.2%)	<0.05
Enthesal pain, n/tot (%)	6 (14.6%)	0 (0)	6 (66.7%)	<0.001
Dactylitis	0	0	0	NA

PASI: Psoriasis area severity index. BSA: Body surface area. TJC: Tender Joint Count. SJC: Swollen Joint Count.

Conclusion: This study shows that the presence of ultrasound abnormalities was higher in the symptomatic group and considering the fact that the most frequent inflammatory ultrasound finding was enthesitis, we might consider the enthesitis as a possible landmark for developing juvenile psoriatic arthritis.

References:

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Disclosure of Interests: None declared

Table 2 PDUS Findings:

Patients	Total (n=41)	Asymptomatic (n=32)	Symptomatic (n=9)	p
≥ 1 ultrasound abnormality, n/tot (%)	19 (46.3%)	13 (40.6%)	6 (66.7%)	NS
≥ 1 joint effusion, n/tot (%)	11 (26.8%)	9 (28.1%)	2 (22.2%)	NS
≥ 1 synovitis, n/tot (%)	3 (7.3%)	1 (3.1%)	2 (22.2%)	NS
≥ 1 enthesitis, n/tot (%)	7 (17.1%)	3 (9.4%)	4 (44.4%)	<0.05
≥ 1 tenosynovitis, n/tot (%)	0 (0)	0 (0)	0 (0)	NA
≥ 1 nail with modified structure, n/tot (%)	22 (53.7%)	19 (59.4%)	3 (33.3%)	NS

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SAT0500 HOW THE ADULT CRISS WORKS IN PEDIATRIC JSSC PATIENTS - RESULTS FROM THE JUVENILE SCLERODERMA INCEPTION COHORT

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Background: The Composite Response Index in Systemic Sclerosis (CRISS) was developed by Dinesh Khanna as a response measure in patients with adult systemic sclerosis. CRISS aims to capture the complexity of systemic sclerosis and to provide a sensitive measure for change in disease activity. The CRISS score is based on a two-step approach. First, significant disease worsening or new-onset organ damage is defined as non-responsiveness. In patients who did not fulfill the criteria of part one, a probability of improvement is calculated for each patient based the Rodnan Skin Score (mRSS), percent predicted forced vital capacity (FVC%), patient and physician global assessments (PGA), and the Health Assessment Questionnaire Disability Index (HAQ-DI). A probability of 0.6 or higher indicates improvement.

Objectives: The objective of this study was to validate the CRISS in a prospectively followed cohort of patients with juvenile systemic sclerosis (jSSc).

Methods: Data from the prospective international inception cohort for jSSc was used to validate the CRISS. Patients with an available 12-months follow-up were included in the analyses. Clinically improvement was defined by the anchor question about improvement (much better or little better versus almost the same, little worse or much worse) in patients overall health due to scleroderma since the last visit provided by the treating physician.

Results: Forty seven jSSc patients were included in the analysis. 74.2% had diffuse subtype. The physician rated the disease as improved in 34 patients (72.3%) since the last visit. No patient had a renal crisis or new onset of left ventricular failure during the 12-months follow-up. Three patients (3.4%) each had a new onset or worsening of lung fibrosis and new onset of pulmonary arterial hypertension. In total, 6 patients resulted in a rating of not improved based on the CRISS in part I. The mRSS, FVC%, CHAQ and PGA significantly improved during the 12-months follow-up in patients who were rated as improved. The predicted probability based on the CRISS algorithm resulted in an area under curve of 0.77 predicting the anchor question of improvement. In summary, 33 (70.0%) patients were correctly classified by the adult CRISS score resulting in an overall area under curve of 0.7.

Conclusion: The CRISS score was evaluated in a pediatric jSSc cohort for the first time. It showed a good performance. However, it seems that the formula of part II of the CRISS score needs a calibration to pediatric jSSc patients.

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