if the number of patients is small. Further studies are needed to identify a reliable predictive marker of uveitis risk in JIA patients. The finding of a significant greater prevalence of anti-DFS70 autoantibodies in healthy ANA + subjects allows to suppose that this autoantibody could represent a possible protective marker for development of AARDs in asymptomatic children with isolated ANA positivity, as for adults. To confirm this hypothesis, it would be useful to carry on the study prospectively, encompassing children with other rheumatological diseases, and prolonging the clinical and laboratory follow up.

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AB1004 JUVENILE DERMATOMYOSITIS (JDM) IN SOUTHEAST ASIA: A 20-YEAR SINGAPORE EXPERIENCE

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Background: Juvenile dermatomyositis (JDM) is a multisystem inflammatory disease of childhood with variable demographics, clinical features and outcomes. No studies have described the characteristics of JDM patients from Southeast Asia population.

Objectives: To describe the clinical characteristics and outcomes of JDM patients in Singapore over a 20-year period.

Methods: Patients diagnosed with JDM from 1999 to 2019 at KK Women's and Children's Hospital, Singapore, were recruited. Nonparametric descriptive statistics were used to described data. Kaplan-Meier analyses were used to estimate the probability of remission. Multivariate logistic and Cox regression analyses were used to determine predictors as appropriate. The significant level was set at < 0.05.

Results: 32 JDM were identified. Clinical characteristics and treatment used are shown in Table 1.

	All (n=32)	Monophasic (n=17)	Polyphasic (n=14)
Male	14 (43.8)	7 (41.2)	7 (50.0)
Age at onset (yrs)*	6.4 (4.5 – 9.8)	5.4 (4.1 – 8.5)	7.4 (5.6 – 12.3)
Lag period (mo)*	3.5 (1.0 – 12.5)	2.0 (1.0 – 16.8)	6.8 (1.2 – 15.8)
Heliotrope	16 (50)	9 (52.9)	7 (50)
Gotttron papule	23 (71.9)	12 (70.6)	11 (78.6)
Malar rash	19 (61.3)	9 (52.9)	10 (71.4)
Vasculitic rash	19 (61.3)	8 (47.1)	11 (78.6)
Arthritis	10 (31.3)	3 (17.6)	7 (50)
Nailfold changes	28 (87.5)	15 (88.2)	13 (92.9)
Calcinosis	9 (28.1)	4 (23.5)	5 (35.7)
Positive ANA	17 (53.1)	10 (58.8)	7 (50)
Positive Myositis antibodies	4 (12.5)	2 (11.8)	2 (14.3)
Laboratory at diagnosis, U/L*			
CK	324 (134 – 2229)	746 (139 – 2965)	351 (155 – 2622)
LDH	650 (450 – 943)	650 (398 – 1015)	714 (512 – 944)
ALT	28 (16 – 106)	35 (18 – 109)	38 (15 – 104)
AST	50 (30 – 108)	50 (31 – 113)	85 (36 – 215)
Aldolase	13.7 (8.1 – 28.0)	14.2 (8.4 – 26.6)	16.3 (8.6 – 38.5)
CMAS score at diagnosis	29 (21 – 43)	28 (21 – 35)	35 (11 – 43)

*median (IQR), otherwise - n (%)

Pulse methylprednisolone (pMP) was used in 53.1% of patients after diagnosis. Median time to inactive disease (ID) was 5.3 months (IQR 2.8 – 12.8). Male, older age and patients on pMP ($p = 0.003$ - 0.044) achieved ID sooner. Older patients also developed disease flare sooner after achieving ID ($p = 0.024$). No clinical features nor lab investigations predicted JDM disease course. Malay patients was associated with higher risk of calcinosis ($p = 0.017$).

Compared to adult dermatomyositis patients in Singapore¹, our cohort had more cutaneous manifestations including malar rash, vasculitic rash and nailfold changes. Table 2 shows the time for each muscle enzymes to normalise.

CK	1.49 (0.69 – 3.53)
LDH	4.53 (2.35 – 25.04)
ALT	1.71 (0.90 – 4.13)
AST	0.97 (0.53 – 3.12)
Aldolase	3.12 (2.35 – 8.30)

median in months (IQR)

Conclusion: Our cohort of JDM patients had more calcinosis compared to other Asian population². Malay population is at higher risk of this complication. It is crucial to achieve ID state in the shortest time possible to avoid significant morbidity. Our study suggests that early treatment with pMP is associated with shorter time to ID. There is no predictor identified for disease course, similar to previous studies³.

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AB1005 BULLOUS LUPUS (BSLE) AS THE FIRST MANIFESTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS IN THE PEDIATRIC POPULATION (PSLE): A DIAGNOSTIC CHALLENGE IN DAILY PRACTICE

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Background: Cutaneous manifestations are observed in 59–85% of patients with SLE but less than 5% developed BSLE. In the GLADEL cohort, the prevalence is 0.41%. BSLE literature in children is scarce

Fig 1. Histology and DIF of BSLE H-E a and b: subepidermal blisters c and d: abundant neutrophils are observed e: DIF full house linear pattern

Fig 2. skin lesions of BSLE a: tense serous blisters, scars and erosions b: purpuric macules in palms c: blisters in groin d-g: blisters and hypopigmented scars

Objectives: to describe the clinical characteristics of the patients with BSLE

Methods: series of cases between 2010-2019 of two reference centers. The cases met Camisa and Grimwood criteria for BSLE

Results: 5 cases had bullous lesions that resolved with residual hypopigmentation. One case had focal seizure and other patient had arthritis with leukopenia and thrombocytopenia. 2 patients had proteinuria <500 mg/24 hours. There were no cases of lupus nephritis. The median SLEDAI-2K score was 12 (IR: 8-17). All had ANAs in titers greater than 1:160 and four had anti-DNA (+). 5 patients had anti-RNP and 4 had anti-Sm. One case had anti-Ro/anti-La. All presented low C3 and 80% had low C4. 80% had ESR $\geq$ 20 mm/hour and CRP greater than 0.5 mg/dl in 60%. All presented clinical response with glucocorticoids and dapsone; one patient had methemoglobinemia that improved. At 3 months, the blisters did not recur in 4 patients except one case that presented relapse due to inadherence.

Histologically, the most common finding was subepidermal blisters with neutrophils in the papillar dermis. DIF showed linear deposits of Igs and complement in 4 cases and granular deposits in one case; IgG/IgM were in 5 of the samples. IgA was positive in 60% and C3 in 80%

Conclusion: In this series, BSLE was associated with neuropsychiatric, joint and haematological involvement in 40% of patients, without lupus nephritis. Such abnormalities had a parallel course to skin involvement, without recurrences. BSLE tends to have a single-phase behavior and in children unlike adults, severe renal involvement is uncommon

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Table 1. Overview of patients with BSLE. BMZ: basal membrane zone MTP: methylprednisolone DDS: dapsone

Age	SLEDAI-2K	Distribution	Mucosal lesions	Systemic findings	Laboratory	DIF	Treatment
14 M	8	Face Trunk Extremities Genitals	Ulcers on mouth and tongue Nasal erosions	No	ANA 1:2560 speckled pattern Anti-DNA 1:40 UI ENAS: anti-Sm + anti-RNP + C3: 86 mg/dl C4: 3.9 mg/dl	Igs deposition in the dermal side Linear pattern IgG, IgA, IgM and C3	MTP Pulses PDN 50mg/day CQ AZA DDS
12 F	12	Trunk Upper and lower extremities	Ulcers on mouth and tongue	Proteinuria non significant (<500 mg/24 hrs)	ANA 1:2560 homogeneous Anti-DNA >200 IU ENAS: anti-Sm + anti-RNP + C3: 36.3 mg/dl C4: 2.10 mg/dl	Igs deposition along the BMZ Linear pattern IgG, IgM, C3 and C1q	PDN 35 mg/day CQ AZA DDS
15 F	10	Face Trunk Extremities	Ulcers on mouth, hard palate and tongue	Proteinuria non significant	ANA 1:640 speckled and cytoplasmic Anti-DNA – ENAS: anti-RNP + anti-Sm + anti-Ro + Anti-La + C3: 56 mg/dl C4: 8.1 mg/dl	Igs deposition along the BMZ Linear pattern IgG, IgA, IgM and C3	PDN 50mg/day CQ DDS Mycophenolate IVlg

Table 1. Cont

10 F	17	Blister lesions: face, upper and lower extremities Purpura: both legs	No	Hematologic: leukopenia, lymphopenia and thrombocytopenia Arthritis	ANA: 1:640 speckled Anti-DNA 1:40 UI ENAS: anti-RNP + anti-Sm - C3: 51 mg/dl C4: 4.4 mg/dl LA: + IgG aCL: 92 GPL IgM aCL: -	Igs deposition along the BMZ Granular pattern IgM. IgG: + IgA, C3: -	PDN 30 mg/day DDS
9 F	16	Face Trunk Extremities	Ulcers on mouth and tongue	Focal convulsion	ANA 1:160 homogeneous Anti-DNA 1:10 UI ENAS: anti-RNP + anti-Sm + C3: 73 mg/dl C4: 12.8 mg/dl	Igs deposition in the dermal side Linear pattern IgG, IgA, IgM and C3	MTP Pulses PDN 25mg/day CQ DDS