not no change significantly over time. Two thirds of incident cases were diagnosed in winter/spring and point prevalence for S-JIA reached 7.15/100,000 (CI 5.29-7.45) in 2014. Disease course was monocyclic in 42% of patients with MAS findings recorded in 4.4% and no case fatality observed during median follow up of 8years. Twenty-seven patients (58%) were readmitted for S-JIA with higher rates per 100 person years in boys (58.3/100, CI 44.5-74.9 vs 14.7 CI 9.9-20.9) (p<0.001). Girls had significantly higher admissions rates for conditions not directly related to S-JIA (rate 103.3 vs 54.4, P<0.001). At last follow-up, arthroplasty had been performed in 4% of patients and there was a high rate of ED visits (72.3/100 person years) after initial s-JIA admission.

Conclusion: The epidemiology of S-JIA in Australia is not unlike other countries [2]. While s-JIA incurred no deaths in the biologic era, S-JIA admission was followed by significant morbidity and health care usage over a lengthy period suggesting the need for extended care for s-JIA patients.

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Acknowledgements: Supported by an grant from the Arthritis Foundation of Western Australia and a project grant from Arthritis Australia (2021- 5101780) to JCN. We thank the Data Custodians of the Hospital Morbidity Data Collection (HMDC), Emergency Department Data Collection (EDDC), the Western Australian Cancer Registry (WACR), the State Registry of Births, Deaths and Marriages and the staff at Data Linkage Branch at the WA Department of Health for their assistance in provision of data.

Disclosure of Interests: None Declared.

DOI: 10.1136/annrheumdis-2023-eular.1912

AB1419 IS DECREASED BODY MASS INDEX -2 Z SCORE OR LESS CORRELATING WITH AN ORGAN INVOLVEMENT PATTERN? RESULTS FROM THE JUVENILE SCLERODERMA INCEPTION COHORT

Keywords: Systemic sclerosis

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Background: Juvenile systemic sclerosis (jSSc) is an orphan disease with a prevalence of 3 in 1 000 000 children. In adult patients decreased body mass index (BMI) correlates with higher mortality. We hypothesized that jSSc patients with lower BMI at presentation have more severe organ involvement.

Objectives: To compare the organ involvement pattern at inclusion into the cohort of jSSc patients with BMI ≤ -2 z score with the patients with higher BMI.

Methods: We reviewed the clinical characteristics of patients who were recruited to the juvenile jSScC till 1st of December 2022. We compared patients with BMI ≤ -2 z score with patients (lwgroup) with higher BMI (nlwgroup). jSScC is a prospective cohort of jSSc patients, who developed the first non-Raynaud´s symptom before the age of 16 years and are under the age of 18 years at the time of inclusion.

Results: At the time of the evaluation, we had 232 patients in the cohort and 217 of them had BMI data to include in the evaluation. Thirty-three patients were in the lwgroup (15%) and 88% (n=29/33) of them diffuse subtype and in the nlwgroup 64% (113/177). The median age at onset of Raynaud phenomenon in the whole group was 10.6 years and the median age at the first non-Raynaud symptom in the whole group was 11.0 years. Median disease duration in the whole group was 2.4 years at the time of inclusion. Approximately 95% of the patients were treated with a DMARD. There were no statistically significant differences between the lwgroup compared to nlwgroup regarding antibody pattern, inflammatory marker or organ involvement pattern, except higher number of patients with Gottron papules (41% lwgroup vs 25% nlwgroup; p=0.01) and sclerodactylia (84% lwgroup vs 73 % nlwgroup; p=0.049). Regarding the patient related outcomes at inclusion in the cohort, the global disease activity by VAS 0-100 was 40 in both groups (p=0.032), but the patient global disease damage by VAS 0-100 was 50 in the lwgroup which was significantly higher compared to 30 nlwgroup (p=0.014).

Table 1. Comparison of patients with different BMI z-scores at time of inclusion in the cohort

	Z-Score ≤ -2 N=33	Z-Score > -2 to < 2 N=177	P value
Gottron Papules	41% (13/32)	25% (44/175)	0.001
Puffy Fingers	13% (4/30)	38% (61/159)	0.049
Sclerodactylie	84% (27/32)	73% (121/166)	0.049
Patient global disease damage	50 (30 – 75) n=27	30 (10 – 55) n=133	0.014

Conclusion: In our jSSc cohort, currently the largest of the world, we could not find any differences regarding major internal organ involvement in patients with lower BMI at time of inclusion in the cohort. Nevertheless, there is a significant difference in patient related outcomes regarding global organ damage between the two groups. The long-term prognosis of these patients should be addressed in future studies.

REFERENCES: NIL.

Acknowledgements: NIL.

Disclosure of Interests: None Declared.

DOI: 10.1136/annrheumdis-2023-eular.2090

AB1420 PAEDIATRIC PRESENTATION OF ANTIPHOSPHOLIPID SYNDROME: A REVIEW OF RECENT LITERATURE WITH ESTIMATION OF LOCAL PREVALENCE

Keywords: Anti-phospholipid syndrome, Systemic lupus erythematosus, Epidemiology

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Background: No evidence exists on any study investigating the prevalence of paediatric Antiphospholipid Syndrome (APS) in the general population (1) and most of the available recommendations on how to manage these patients come from observational studies and physicians' experience.

Objectives: We aimed to investigate the epidemiology, the clinical and laboratory characteristics of the paediatric involvement of Antiphospholipid syndrome (APS), by performing a systematic review of the current evidence and reviewing local experience in the Northwest Italy.

Methods: We performed a detailed literature search to identify articles describing clinical and laboratory characteristics of paediatric APS patients. In concomitance, we conducted a registry-study collecting data from the Piedmont and Aosta Valley Rare Disease Registry including paediatric patients diagnosed with APS in the last ten years.

Results: The systematic review included nine articles with a total of 386 paediatric patients [65% females, 50% with Systemic Lupus Erythematosus (SLE) as concomitant diagnosis]. Rate of venous and arterial thrombosis were 57% and 35% respectively. "Extra-criteria manifestations" of APS included mostly hematological and neurological involvement (91 thrombocytopenia, 32 haemolyticanaemia, 19 chorea, 8 multiple sclerosis-like lesions, 9 migraines and one transverse myelitis). Almost one quarter of patients (19%) reported recurrent events and 13% manifested as CAPS. Further details of the included studies are detailed in Table 1. A total of 17 paediatric patients (mean age 15.1 ±2.8, 76% female) developed APS in the Northwest of Italy. In 29% of cases SLE was a concomitant diagnosis. Deep vein thrombosis was the most frequent manifestation (28%) followed by Catastrophic APS (CAPS) (6%). Taking into account that the paediatric population of the Piedmont and Aosta Valley Region is around 680,000 persons, the calculated estimated prevalence of paediatric APS in Piedmont and Aosta Valley Region is 2.5 cases per 100,000 people. When analyzing the data of the register in an eleven year period (from January 2011 to December 2021), the estimated mean annual incidence was 0.2 cases per 100,000 inhabitants.

Conclusion: Clinical manifestations of paediatric APS seem to be more severe and with a high prevalence of non-criteria manifestations. International efforts are needed to better characterize this condition and to develop new specific diagnostic criteria to avoid missed/delayed diagnosis in children with APS.

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Table 1. Main characteristics of the studies included in the Systematic Review

First Author, year of publication	Design	Setting	Focus	Number of paediatric APS	Age (mean, ±SD; median, range)	PAPS (n)	SAPS (n)
M. Gattorno, 2003	Prospective	Multicenter	pPAPS prospective cohort	14	9 (3-13)	12	2
Y. Berkun, 2006	Prospective	Multicenter	pPAPS prospective cohort	28	12.9 ±3.7	23	5
T. Avcin, 2008	Registry	Multicenter	Descriptive cohort	121	10.7(1–17)	61	60
H. Berman, 2014	Registry	Multi center	CAPS	45	11,5 ±4,6	31	14
A.Nageswara Rao, 2016	Retrospective cohort	Single center	Descriptive cohort	17	15,3 (1-19)	9	8
J. Ma, 2017	Retrospective cohort	Single center	Descriptive cohort	58	14 ±3	14	44
A. G. Islabão, 2020	Retrospective cohort	Multi center	APS in pSLE	67	12 (4-18)	0	67
E. Sloan, 2021	Retrospective cohort	Single center	APS and aPL in pCTD	15	12.7 ±3.3	0	15
J. A. Madison, 2022	Retrospective cohort	Single center	pAPS	21	16 (8-18)	10	11

Acknowledgements: NIL.
Disclosure of Interests: None Declared.
DOI: 10.1136/annrheumdis-2023-eular.2272

AB1421

DOES THE SACROILIAC JOINT MAGNETIC RESONANCE IMAGING ENHANCE THE ENTHESITIS-RELATED ARTHRITIS DIAGNOSIS?

Keywords: Imaging

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Background: Magnetic Resonance Imaging (MRI) is the gold standard in the diagnosis of adult spondyloarthritis [1]. MRI sensitivity and specificity seem to be less studied in juvenile spondyloarthritis, especially in Enthsitis-related Arthritis (ERA).
Objectives: We aimed to determine the sensitivity and specificity of MRI in the diagnosis of ERA.
Methods: We conducted a retrospective study including 44 patients with Juvenile Idiopathic Arthritis (JIA) meeting the International League of Associations for Rheumatology (ILAR) 2001 criteria. For each patient, we collected the following data: age, age at the onset of JIA, JIA subtype, disease duration, C-reactive protein (CRP), HLA-B27 typing, and Erythrocyte sedimentation rate (ERS) levels. Each patient had a sacroiliac joint MRI. Sacroiliitis was defined using the OMERACT JAMARIS scoring system [1]. Disease activity was assessed using the JSpADA score for ERA and JADAS-CRP-10 for the rest of the JIA subtypes. We divided patients into two groups: Group 1 (G1) patients with ERA and Group 2 (G2) patients with non-ERA subtype. Statistical analysis was performed using SPSS software.
Results: We included 28 boys and 16 girls. The mean age was 13.65 ± 4.62 years. The mean age at the onset of the disease 9.57 was ± 3.97 years. The mean disease duration was 4.34 ± 3.09 years. There was enthesitis-related arthritis in 61% of the cases (n=27), oligoarticular JIA in 14% of the cases (n=6), polyarticular JIA in 11% of the cases (n=5), and psoriatic arthritis in 7% of the cases (n=3). JIA was undifferentiated in 7% of the cases (n=3). Sacroiliac joints were painful in 15 patients (34%). The mean CRP and ESR were 14.42 ± 19.67mg/L and 26.56 ± 20.87mm, respectively. The mean JADAS and JSpADA were 6.6 ± 4.7 and 3.09 ± 1.55, respectively. HLA-B27 was performed in 22 patients and was positive only in five patients. IRM was normal in 25 patients (57%) (G1: 11 versus G2:14) and showed bone marrow in the sacroiliac joints in 19 patients (34%) (G1=16 versus G2=3, p=0.005). The sensitivity and specificity of the sacroiliac joints MRI in the diagnosis of ERA were 61.54% and 82.35%, respectively. Positive predictive value and negative predictive value were 84.21% and 58.33%, respectively. Furthermore, sacroiliac joint pain in the clinical examination was able to predict sacroiliac bone edema in MRI with an odds ratio of 6.8 (95% confidence interval 1.68 to 28.09; p=0.006).
Conclusion: According to other studies, ours showed that MRI inflammatory lesions could be observed in JIA regardless of JIA sub-types [2]. Interestingly, we found that MRI abnormalities were not sensitive but had a good specificity in the diagnosis of ERA. Thus, MRI could guide the diagnosis of ERA but could not be the gold standard in this disease.

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Acknowledgements: NIL.
Disclosure of Interests: None Declared.
DOI: 10.1136/annrheumdis-2023-eular.4072

AB1422

CLINICAL MANIFESTATIONS AT THE ONSET OF PEDIATRIC MIXED CONNECTIVE TISSUE DISEASE (PMCTD): A SYSTEMATIC REVIEW.

Keywords: Systematic review, Mixed connective tissue disease, Rare/orphan diseases

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Background: pMCTD is a rare disorder that includes features of systemic lupus erythematosus, polymyositis/dermatomyositis, juvenile idiopathic arthritis, and systemic sclerosis. Fifty years have passed since Sharp identified MCTD in 1972, and diagnosis of this disorder remains challenging.
Objectives: The aim of this review is to identify any clinical features at the diagnosis of pMCTD and manifestations that are not currently part of the available diagnostic criteria.
Methods: A systematic literature review was performed in accordance with PRISMA guidelines using electronic bibliographic databases: MEDLINE via PubMed and EMBASE. Data obtained were extracted using a dedicated database containing clinical data that best categorize patient characteristics. Criteria for inclusion: studies including patients with a pMCTD diagnosis with onset before 18 years of age and reporting a description of initial clinical features.
Results: The search returned a total of 1372 results: 409 articles were excluded as duplicates and 790 based on title/abstract, 133 because of publication type (n = 4) or because the full text was not available (n = 65) or not in English (n = 64). One (n=1) eligible article resulted from manual screening of references cited in the selected publications and in the reviews. Finally, 41 articles were included: 23 case reports, 9 case series, 5 prospective, and 4 retrospective studies from 1973 to 2019, with a total number of 218 patients. They were predominantly female (81.56%, n=167), and the mean age at onset was 147 months (median 126 months, 10.5 years). When indicated, the most commonly used criteria for diagnosis were Kasukawa criteria (50%, 11 studies), then Alarcon-Segovia criteria (31%, 7 studies), then Sharp criteria (23%, 5 studies); no Khan criteria were used. Clinical features are listed in Figure 1.

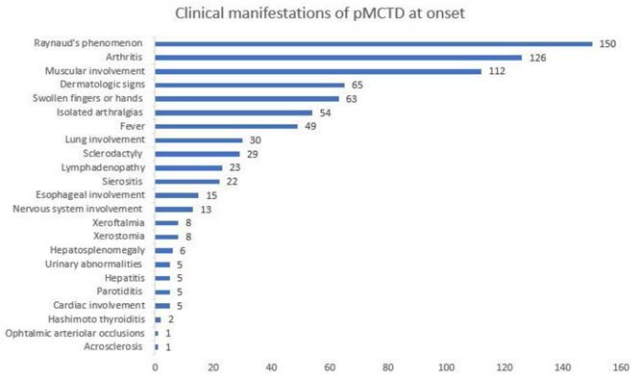

Figure 1. Number of patients for clinical feature at onset.

Joint involvement, Raynaud's phenomenon, myositis, and swollen fingers/hands are the most common clinical features at diagnosis according to the data reported in the literature, although with slightly lower percentages than in other reviews. Dermatologic signs are very heterogeneous, but were found to be a very present feature at disease onset, affecting 1/3 of patients. Fever, not covered by any of the diagnostic criteria, was noted in 1/4 of cases. Pulmonary and esophageal involvement are reported in a lower percentage at the onset of the disease, indicating a more developmental nature of these conditions.