(78.8%); Erythema marginatum in 7 (21.2%); Sydenham chorea, 4 (12.1%); Subcutaneous nodules, 2 (6.1%), respectively. And minor criteria were; fever, 27 (81.8%); first degree heart block, 7 (21.2%); elevated inflammatory markers (ESR, CRP), 30 (90.9%). ARF patients were treated with antimicrobial agent therapy 33 (100%); NSAIDs 24 (72.7%); and glucocorticoids therapy 15 (45.4%). All ARF patients were prescribed with antimicrobial agent prophylaxis.

On the other hand, the mean age of PSRA was 8.0 years (3-14 years), and female/male ratio was 17/14. Three patients (9.4%) had monoarthritis, 15 patients (46.9%) had oligoarthritis, and 14 patients (43.6%) had polyarthritis. Two patients had arthritis and enthesitis. And fever in 24 (75.0%) and Elevated inflammatory markers in 29 (90.6%). PSRA patients were treated with antimicrobial agent therapy 25 (78.1%); NSAIDs 27 (84.4%); and glucocorticoids therapy 3 (9.4%). During the follow up, there was no patient with carditis. 18 (56.3%) patients with PSRA were prescribed with antimicrobial agent prophylaxis.

Conclusion: In this study, ARF is rare in the Japanese pediatric population, but ARF has not yet disappeared. We observed high incidence of arthritis, carditis and erythema marginatum. No PSRA case was complicated with carditis. General pediatrician need to have updated information about ARF and PSRA even in industrialized countries.

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SAT0517 MANAGEMENT OF RISK OF VARICELLA INFECTION IN IMMUNOCOMPROMISED CHILDREN: WHAT IS THE EVIDENCE?

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Background: Varicella-naive children on methotrexate are at risk of severe infection if they encounter varicella. Current guidance advises that children on low-dose methotrexate can safely receive live vaccines[1], but management of children on higher doses or combination immunosuppression is challenging as there is no consensus about their management. Acyclovir and varicella-zoster immune globulin (VZIG) are commonly used as post-exposure prophylaxis (PEP).

Objectives: Assess the evidence for use of acyclovir and/or VZIG as PEP in the management of varicella exposure in susceptible children taking methotrexate.

Methods: A literature search using PubMed, the Cochrane Library and EMBASE was conducted from November to December 2017, using the terms methotrexate, immunocompromised, varicella, child, prophylaxis, acyclovir, VZIG and their variations. Only full papers, in English language, studying children were analysed (63 abstracts read for relevance, 28 papers obtained, 11 papers included).

Results: There have been no randomised controlled trials (RCTs) analysing the effectiveness of acyclovir and/or VZIG as PEP in immunocompromised children. The studies that do exist (see table for key publications) are small and uncontrolled and have largely been carried out in oncology rather than rheumatology patients. While they suggest that acyclovir and VZIG are effective at reducing historical infection rates of >70%[1], a significant proportion of recipients still get varicella.

Conclusion: There is only level 3 evidence for the use of acyclovir and/or VZIG as PEP in susceptible children on methotrexate. The literature indicates acyclovir is more effective, but this is a grade 3 recommendation only. A RCT to compare the effectiveness and acceptability of VZIG and acyclovir is needed, however the difficulty of randomising a cohort of similarly immunocompromised patients should not be underestimated.

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SAT0518 READINESS FOR TRANSITION – CROATIAN VERSION AND PILOT EVALUATION OF THE TRANSITION READINESS ASSESSMENT QUESTIONNAIRE (TRAQ) IN RHEUMATOLOGIC PATIENTS

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Background: Rheumatic diseases of childhood extend to adulthood as an active disease in 30-70% of patients, which is the reason for requiring transition of rheumatologic care into adulthood. Transition should be individualized based on patient's readiness, and good transition readiness analysis tools should be available at the time of transition.

Objectives: As systematic review of the available assessment tools for transitional readiness in adolescents with chronic diseases has shown that only the Transition Readiness Assessment Questionnaire (TRAQ) has proven reliability in its key measurement components[1], we have decided to use the TRAQ[2] in pediatric rhematology patients.

Methods: English version of TRAQ was translated to Croatian, afterwards a back-translation to English was done. Due to difference in the insurance policy in Croatia (health care is free for children under 18 years of age), question 9 was modified into: "Are you aware of the fact that after 18 years of age you have to start paying for the additional health insurance?"

A pilot study was performed in order to validate translated TRAQ by applying questionnaire to pediatric rheumatology patients at the time of transition.

Results: A total of 41 patients at the median age of 18.25 years (range 17.8-21.1 years) were enrolled, 28 female and 13 male patients. Most of

Authors	No. of Patients & VZV Status	Cause of Immunosuppression	Treatment	Outcomes
Evans et al (1980)[2]	102 immunosuppressed children, 80 of whom were known to be seronegative for VZ IgG antibodies	Variable	Zoster immune globulin (ZIG) as a single dose - up to 1 year, 100 mg; 1-5 years, 250 mg; 6-10 years, 500 mg; 11-14 years, 750 mg; 15-16 years, 1000 mg	24/80 became infected; 17 with symptoms
Zaia et al (1983)[3]	164 immunocompromised children	Variable 131: malignancy 30: other immunosuppressive treatment 3: primary immunodeficiency	81 given VZIG 83 given ZIG Dose: 1.25 ml/10 kg (max dose, 6.25 ml), given within 96 hours of exposure	VZIG: 49/81 became infected ZIG: 57/83 became infected Varicella pneumonia occurred in 3 cases in each group
Shinjoh et al (2009)[4]	65 immunocompromised children (some with a history of varicella); 76 immunocompetent children; 11 immunocompetent controls	Variable	Oral acyclovir (10 mg/kg/dose; 400 mg max dose; 4 times daily). Minimum of 7 days treatment, starting from 7 days after exposure	2/65 immunocompromised children developed varicella. 1/76 immunocompetent children developed varicella. 2/11 controls developed varicella

the patients had juvenile idiopathic arthritis (28 patients, 68.3%), 7 (17.1%) had mixed connective tissue disease, 3 (7.4%) had Raynaud's syndrome and there was one patient (2.4%) with each of the following diagnosis: SLE, fibromyalgia, and polyarteritis nodosa. The mean follow up time before the transition was 5.3 years (3 months - 14 years).

In general, the TRAQ was well understood and was completed in a short time by the study participants. No major difficulties were observed and all the patients were able to read and answer the questionnaire.

Average score for each of the subparts of TRAQ was: managing medication 4.29 (range 2.25-5.0), appointment keeping 3.82 (1.43-5.0), tracking health issues 3.63 (1.67-5.0), talking with providers 4.68 (2.0-5.0), managing daily activities 4.39 (3.0-5.0) with overall score 4.05 (2.50-5.0), showing good overall readiness of our patients for transition. Readiness for transition in our group of patients is likely due to the age of transitioning over 18 years and good preparing for transition done by the team in the months preceding transition.

Conclusion: The Croatian version of the Transition Readiness Assessment Questionnaire was validated in the population of transitional patients with chronic rheumatic diseases. It has proven to be easily applied and well understood, and its results showed adequate readiness for transition among our patients. According to our 7-year experience and 81% of follow up visits in adult rheumatology among transitioned patients in previous years, we believe that these results are correct.

We shall continue following these patients through adult rheumatology visits and in few years' time we shall verify whether they continue regular follow ups and prove to have been ready for transition as TRAQ has suggested.

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SAT0519 CLINICAL CHARACTERISTICS, TREATMENT AND OUTCOMES OF ENTHESITIS-RELATED ARTHRITIS IN TAIWAN

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Background: Juvenile idiopathic arthritis (JIA) has been categorized into seven different subtypes according to International League of Associations for Rheumatology system (ILAR) criteria [1]. Enthesitis-related arthritis (ERA) has represented the largest subtype in Taiwanese cohort study [2].

Objectives: The aim was to compare the clinical characteristics, treatments and outcomes of ERA in one of tertiary medical center in Taiwan to other subtypes of JIA. Further, to determine patients' characteristics and risk factors help to predict the development of active and non-active treatment outcomes in ERA.

Methods: Retrospective review of patients diagnosed with JIA between March 1993 and December 2018 at a pediatric rheumatology clinic in National Taiwan University Hospital (NTUH), Taipei, Taiwan were enrolled. The outcome assessments were based on Wallace criteria to categorize patient into active and non-active (inactive, remission on medication and remission off medication) group.

Results: One hundred and eighty-three patients were included for 8 years mean follow up duration. Distribution of JIA subtypes in Figure 1 shown ERA was the single largest category of JIA (39.89%) in Taiwan. The demographic details of ERA patients in Table 1 revealed: male predominant (86%), late onset age (11.1±3.2 yrs), majority with HLA-B27 positive (92%), sacroiliac join or lumbosacral involvement (16%) and anterior uveitis (10%). Category specific outcomes in Table 2 showed ERA and extensive oligoarthritis were less likely to achieve non-active treatment response compared to persistent oligoarthritis. Among risk factors contributed to poorer treatment response in ERA were any clinical signs of sacroiliitis with P value of significant (0.0057).

Table 1. ERA patients and disease characteristic, n=73

Characteristics	N (%)
Demographic characteristics	
Male sex	63 (86)
Age at onset, mean ±SD years	11.1±3.2
Age at follow-up visit, mean ±SD years	17.6±5.4
Disease duration at follow-up visit, mean ±SD years	6.5±5.0
Disease characteristic	
Enthesitis and arthritis	71 (97)
Presence of HLA-B27 antigen	67 (92)
Presence of or a history pf sacroiliac joint tenderness and/or inflammatory lumbosacral pain	12 (16)
Onset of arthritis in a male> 6y/o	68 (93)
Acute (symptomatic) anterior uveitis	7 (10)
History of AS/ERA/sacroilitis with IBD, Reiter's syndrome, or acute anterior uveitis in 1st degree relative	8 (11)

Table 2. JIA category-specific outcomes

	ERA n=73	Persistent Oligo n=33	Extensive Oligo n=22	Polyarticular RF(+) n=9	Polyarticular RF(-) n=19	Systemic n= 25
Active	46 (63)	14 (42)	15 (68)	7 (78)	10 (53)	14 (56)
Non-active	24 (33)	18 (55)	5 (23)	2 (22)	8 (42)	8 (32)
Inactive	13 (18)	2 (6)	3 (14)	2 (22)	3 (16)	0 (0)
Clinical remission on medication	5 (7)	1 (3)	0 (0)	0 (0)	0 (0)	0 (0)
Clinical remission off medication	6 (8)	15 (46)	2 (9)	0 (0)	5 (26)	8 (32)
Loss follow up	3 (4)	1 (3)	2 (9)	0 (0)	0 (0)	2 (8)
Expire	0 (0)	0 (0)	0 (0)	0 (0)	1 (5)	1 (4)
Active/Non-active (P)	46/24 (**0.036)	14/18 (————)	15/5 (** 0.027)	7/2 (0.071)	10/8 (0.422)	14/8 (0.151)

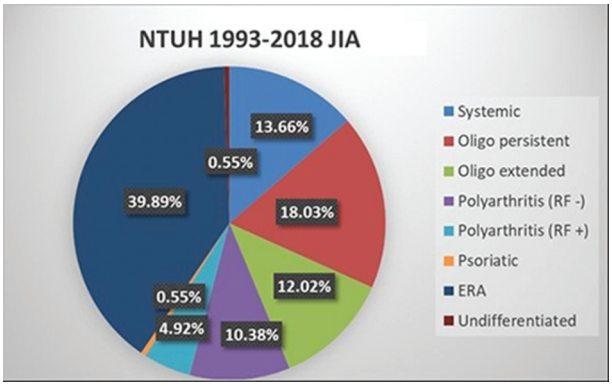

Figure 1. Distribution (%) of JIA subtypes in 183 Taiwanese children from NTUH between year 1993~2018

*Comparison to Persistent Oligoarthritis. **P<0.05 with significant

Conclusion: ERA has represented the most common subtype of JIA in Taiwanese cohort study and has poorer treatment responses when compared to other JIA subtypes. To identify risk factors that contributing to poorer ERA treatment response might help more aggressive therapeutic strategy and improve outcome of ERA.

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