which 88% had onset of JIA before the age of 6 (all girls). Complete remission of uveitis was achieved in two patients treated with topical corticosteroids, with non-steroid anti-inflammatory medication and methotrexate used in one of the patients. Inactive uveitis was most frequently achieved with a combined therapy which included biologics. A patient suffering from uveitis and JIA was found to be significantly more likely to suffer from oligoarthritic subtype (64.5%, CI 45.4-80.8%, $p<0.001$). We determined that a positive ANA titer occurred statistically significantly more frequent in females (OR inf., CI 3.18-inf., $p=0.001$). No significant statistical difference in ANA titer was found between groups of patients who were diagnosed with rheumatologic or ocular disease before and after the age of 6 (OR 1.85, CI 0.12-29.57, $p=0.6111$) nor between different subtypes of JIA. There was no difference in time passed between the occurrence of rheumatologic and ocular manifestations (median difference =1.75 years, $W=131$, $p=0.2597$), nor between ANA titer (OR 0.57, CI 0.09-3.12, $p=0.4775$) between groups with and without ocular complications. Patients who first presented with ocular manifestations did not have a greater likelihood to develop ocular complications (OR 5.59, CI 0.51-299.98, $p=0.175$).

Conclusion: A group of female patients suffering from RF negative subtypes of JIA and uveitis, with onset occurring before the age of 6 and positive ANA was prominent. These patients were traditionally classified as "early onset ANA positive JIA", concurring the need for reclassification of JIA. Limited by a small number of patients, this was partly confirmed with significant statistical findings, a recurrent positive ANA titer in females with JIA and uveitis and a more frequent occurrence of uveitis in oligoarthritis. The occurrence of ocular complications was not dependent upon ANA titer, nor upon the time difference between rheumatic and ocular symptoms or their order of appearance.

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AB0996

APPLICATION OF YAMAGUCHI CRITERIA FOR CLASSIFICATION OF SYSTEMIC JUVENILE IDIOPATHIC ARTHRITIS

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Background: The ILAR criteria require the presence of arthritis to classify a patient as having sJIA. The sJIA patients prior to the onset of arthritis (suspected sJIA) appeared clinically similar to patients fulfilling the ILAR criteria for sJIA.

Objectives: Children with sJIA may have a delayed onset of arthritis and it fail to fulfill the ILAR criteria for sJIA. This study was undertaken to determine whether the Yamaguchi criteria is useful in classification of child with sJIA particularly in pre-arthritis phase of illness.

Methods: Retrospective chart review all patients with a diagnosis of sJIA in Hallym University Medical Center from Feb. 2002 through April 2016.

Results: Total 73 patients were diagnosed by ILAR classification criteria. 38 boys and 35 girls formed the study cohort. There are only 20 patients who are fulfilled of ILAR criteria in sJIA.

Characteristics sJIA (n=20) suspected (n=53) Total (n=73)

Evanescent Rash 19 24

Sore throat 1 5

Lymphadenopathy 2 11

Hepatomegaly 1 11

Splenomegaly 4 10

37 patients were diagnosed with suspected sJIA due to have not an arthritis. 36 patients were presented the arthritis at the onset of sJIA. The Yamaguchi criteria was fulfilled in a higher number of patients in the study (n=44) as compared to the ILAR criteria (n=20). Among the

patients with the absence of arthritis (n=37), 25 patients fulfilled the Yamaguchi criteria. (67.5%).

ILAR criteria Yamaguchi criteria

Yamaguchi criteria fulfilled 16 28

Yamaguchi criteria not fulfilled 4 25

Total cohort 20 53

Conclusion: Overall the Yamaguchi criteria was fulfilled in a higher number of patients in the study (60.2%) as compared to the ILAR criteria (27.3%). The failure of ILAR criteria was mainly due to the absence of arthritis. The Yamaguchi criteria is more helpful to diagnosis of sJIA. The Yamaguchi criteria may be useful on early diagnosis and treatment for patient with delayed onset arthritis at onset of disease.

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AB0997

MATCHED CONTROLLED SURVEILLANCE OF TOCILIZUMAB TREATMENT FOR POLYARTICULAR JUVENILE IDIOPATHIC ARTHRITIS—AN INTERIM ANALYSIS

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Background: Tocilizumab (TOC) is approved for treatment of polyarticular juvenile idiopathic arthritis (poly-JIA). Real world data are limited.

Objectives: Long-term surveillance of patients newly initiating tocilizumab treatment for at least five years compared to a cohort of patients newly initiating a comparator biologic using the BIKER-registry.

Methods: Baseline demographics, clinical characteristics and disease activity parameters as well as efficacy and safety parameters were compared between patients using TOC and a control cohort using alternative biologics. The cohorts were matched for region and date of therapy start. Efficacy outcome variables were JADAS10 and joint counts. Functional status was determined with the Childhood Health Assessment Questionnaire disability-index (CHAQ-DI). Safety assessments were based on adverse events (AE) reports.

Results: Patients starting on TOC had a longer disease duration (5.2y vs. 2.9 y; $p=0.0001$) and had TOC significantly more often as second line biologic than patients in the comparator group (80% vs 14%, $p<0.0001$). No differences could be observed regarding disease activity parameters (JADAS 10 14.7 +/- 6.4 vs 15.8 +/- 6.0) or disability (CHAQ DI 0.65 +/- 0.63 vs 0.66 +/- 0.66) at baseline. Patients treated with TOC showed a substantial response to treatment with a significant reduction in JADAS 10 from 14.7 to 5.4 ($p<0.0001$). After 6 months of TOC treatment, 40% of patients had reached JADAS minimal disease activity (MDA), 22% of patients had reached JADAS remission. Tolerability was comparable between the two cohorts, with 31 in the TOC versus 34 patients in the control cohort experiencing AE; serious AE (SAE) were documented in 3 patients in the TOC and one patient in the control cohort. The SAE in the TOC cohort were medication intoxication with suicidal intent, JIA flare, gastrointestinal infection and abdominal pain in 1 patient each. In the control cohort there were JIA flare in 2 patients and gastrointestinal infection in 1 patient. No significant differences regarding infectious AEs, cytopenias or elevated transaminases were observed. No other AE were reported more than once; no deaths occurred.

Conclusion: Comparability of efficacy was limited due to the fact that patients in the TOC cohort had longer disease duration and were more often second line biologic users. Tolerability was comparable in both cohorts. Long-term observation is ongoing.

Abstract AB0997 Table 1. Comparison of TOC patients and matched controls. Baseline parameters and adverse events are listed. *Number and rate of patients are given. # t-test or χ^2 -test as appropriate

	TOC cohort N=88	Matched controls N=88	P-value#
Baseline Parameters			
Gender female, n (%)	73 (82.9)	67 (76)	0.35
Age onset, mean (SD), years	7.5 (4.6)	7.5 (4.7)	0.89
Disease duration, mean (SD), years	5.2 (4.4)	2.9 (2.8)	0.0001
RF neg. poly-JIA, n (%)	57 (64.8)	62 (70.5)	0.52
RF pos. poly-JIA, n (%)	10 (11.4)	10 (11.4)	1.0
Extended Oligo-JIA, n (%)	21 (23.9)	16 (18.2)	0.46
Pre-treatment with biologics, n (%)	70 (79.5)	12 (13.6)	<0.0001
Active joint count, mean (SD)	7.3 (7.4)	7.4 (5.9)	0.92
CHAQ DI, mean (SD)	0.65 (0.63)	0.66 (0.66)	0.92
CRP, mg/l, mean (SD)	8.9 (18.9)	9.4 (18.6)	0.86
JADAS10, mean (SD)	14.7 (6.4)	15.8 (6.0)	0.24
Adverse Events			
Patients with AE, n (%)*	31 (35.2)	34 (38.6)	0.75
Patients with SAE, n (%)	3 (3.4)	1 (1.1)	0.62
Patients with infectious AE, n (%)	19 (21.6)	20 (22.7)	1.0
Patients with medically important infections, n (%)	1 (1.1)	3 (3.4)	0.62
Patients with increased transaminases, n (%)	4 (4.5)	3 (3.4)	1.0
Patients with hyperbilirubinemia, n (%)	3 (3.4)	-	0.25
Patients with neutro-/leukopenia, n (%)	3 (3.4)	1 (1.1)	0.62

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AB0998 SEVERE IGA VASCULITIS: CYCLOPHOSPHAMIDE IN QUESTION

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Background: Cyclophosphamide (CYC) has been a mainstay in the treatment of severe childhood primary systemic vasculitis. Although major

organ involvement is rare in IgA vasculitis (IgAV), CYC is among therapeutic options in moderate and severe IgAV nephritis.(1) No specific recommendations are available for cerebral vasculitis of IgAV.

Objectives: To describe disease course and treatment response in 2 patients with severe IgAV with CNS and renal involvement.

Methods: Case reports with disease assessments using Paediatric Vasculitis Activity Score (PVAS).(2)

Results: Both children were Caucasian boys with unremarkable previous history aged 4 (Patient 1) and 8 (Patient 2) years. In both cases the initial typical presentation included purpura and abdominal pain in both, arthritis in Patient 1 and haematuria in Patient 2. After 2 weeks of prednisone therapy for severe gastrointestinal (GIT) symptoms Patient 1 was admitted for right-sided weakness and facial palsy. His brain MRI revealed an ischaemic lesion. His PVAS reached 21/63 for skin, abdominal, renal (haematuria) and CNS systems. When Patient 2 was admitted for abdominal symptoms he developed a focal epileptic paroxysm. His brain MRI was compatible with cerebral vasculitis. His nephritic-nephrotic syndrome prompted renal biopsy showing mesangioproliferative nephritis with IgA deposits. His PVAS was 27/63 for skin, abdominal, renal (hypertension, haematuria, proteinuria) and CNS systems. Both patients received 3 doses of intravenous methylprednisolone 30 mg/kg followed by oral prednisone 1 mg/kg with subsequent tapering and 3 doses of CYC 500mg/m²(monthly i.v.). At follow-up 3 months after the 1st CYC dose both patients fully recovered neurologically. Patient 2 was on prednisone (0.2 mg/kg) and ACE inhibitor and his urine was negative for the first time, PVAS=0. Patient 1 had been developing significant proteinuria during prednisone withdrawal and his PVAS counted 10/63 for renal disease. Prednisone was re-instituted and ACE inhibitor added while renal biopsy has been pending.

Conclusion: In both cases therapy with corticosteroids and CYC led to the full recovery of CNS disease but did not prevent progression of nephritis in Patient 1. Response of Patient 2 nephritis to CYC appeared satisfactory at 3 months visit but would require confirmation after full prednisone withdrawal. Although generally benign, when IgAV presents with major organ involvement its treatment is challenging. Further research is needed in order to gather better evidence to support treatment recommendations.

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AB0999 TREATMENT FOLLOW UP AFTER TRANSITION FROM PAEDIATRIC TO ADULT RHEUMATOLOGY CARE

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Background: Many juvenile onset rheumatic diseases continue to have activity in adulthood. Therefore, a continuous uninterrupted healthcare of these patients is needed.

Objectives: To determine whether and how therapy has changed in patients after transition from paediatric to adult rheumatology service in our medical centre.

Methods: A retrospective single centre study was performed. Patients seen at least once at both paediatric and adult rheumatology outpatient service were enrolled. Data was collected from patients medical records.

Results: A total of 50 patients were included. Most of them were female (82%). Average age at transition was 19.6 years (range 18- 27 years). Mean time of follow up was 44 months (range 2- 119 months). Juvenile idiopathic arthritis (JIA) was the most common juvenile onset chronic rheumatic disease. The medications used either as monotherapy or in combination before transition at last visit at paediatric rheumatology