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

Disclosure of Interests: Ariane Klein: None declared, Dennis Conzelmann: None declared, Toni Hospach Speakers bureau: Chugai, Roche, Novartis, Frank Weller-Heinemann: None declared, Sandra Hansmann: None declared, Jasmin Kuemmerle-Deschner Grant/research support from: Jasmin Kuemmerle-Deschner is an employee of University of Tuebingen, Germany, and received consultants/speakers fees from Novartis and SOBI pharmaceuticals and grant support from SOBI and Novartis., Consultant for: Jasmin Kuemmerle-Deschner is an employee of University of Tuebingen, Germany, and received consultants/speakers fees from Novartis and SOBI pharmaceuticals and grant support from SOBI and Novartis., Speakers bureau: Jasmin Kuemmerle-Deschner is an employee of University of Tuebingen, Germany, and received consultants/speakers fees from Novartis and SOBI pharmaceuticals and grant support from SOBI and Novartis., Michael Borte Grant/research support from: Pfizer and Shire, Kirsten Minden Consultant for: AbbVie, Ivan Foeldvari Consultant for: Chugai, Novartis, Christoph Rietschel: None declared, Gerd Ganser: None declared, Ralf Trauzeddel: None declared, Markus Hufnagel: None declared, Dirk Foell Grant/research support from: not specified, Consultant for: not specified, Speakers bureau: not specified, Rainer Berendes: None declared, Gundula Boeschow: None declared, Prasad Oommen: None declared, Angelika Thon: None declared, Gerd Horneff: None declared
DOI: 10.1136/annrheumdis-2019-eular.2750

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.

REFERENCES

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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.6731

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