results included 38 JIA-U patients aged 2 to 15 years and 69 eyes (7 patients treated with IMT was conducted at University Hospital Centre Zagreb, Department of Ophthalmology, University of Zagreb School of Medicine; University Hospital Centre Zagreb, Department of Pediatrics, University of Zagreb School of Medicine, Zagreb, Croatia; Anna Meyer Children’s Hospital, Florence, Italy; Royal Hospital for Children, Glasgow, UK; University Hospital Centre Split, University of Split School of Medicine, Split; University Hospital Dubrava, University of Zagreb School of Medicine, Zagreb, Croatia; Faculty of Pharmacy and Biochemistry, Zagreb, Croatia; Dana Dawe Children’s Hospital, Tel Aviv, Israel; Division of Rheumatology, Giannina Gaslini Institute, Genoa; Division of Clinical Rheumatology, Research Center for Adult and Pediatric Rheumatic Diseases, Università degli Studi di Milano, Milan, Italy; Department of Pediatric Rheumatology, Hacettepe University, Ankara, Turkey; Children’s Hospital Zagreb, Josip Juraj Strossmayer University of Osijek, Medical Faculty Osijek, Zagreb, Croatia).

**Methods** The longitudinal observational study with JIA-U patients treated with IMT was conducted at University Hospital Centre Zagreb in the period from 2011 to 2017. We included 38 JIA-U patients aged 2 to 15 years and 69 eyes (7 patients had unilateral JIA-U).

**Results** At baseline 46 (66.7%) eyes had grade ≤1+ of inflammation in anterior chamber (AC) according to Standardization of Uveitis Nomenclature (SUN) Working group criteria, 11 (15.9%) had grade 2+, 3 (4.4%) had grade ≥3+, 23 children (60.5%) had already received methotrexate (MTX), 8 (21.0%) had grade 2+, 3 (4.4%) had grade 3+. 23 children of Uveitis Nomenclature (SUN) Working Group criteria, 11 patients treated with both biologics and MTX, achieved level of inflammation in AC. In the first 12 months, 3 (8.8%) eyes had a median of 4 daily doses, GC ointment in glucocorticoids (TGC) in the form of drops were used in 61 (88.4%) eyes with a median of 2-7 daily doses of TGC, and in the 48th month in 61.1%. In the 12th month in 75% of eyes the grade 0 of inflammation was achieved with ≤2 doses of TGC, and in the 48th month in 61.1%. According to milder criteria, in the 12th month 90% of the eyes have a degree of inflammation ≤0,5+ with ≤2 doses of TGC, and in the 48th month all patients achieved this goal.

**Conclusion** It was shown that the results of treatment outcomes during follow-up largely depend on the selected outcome measures. This will be important for future research because setting different limits can lead to a more favorable outcome.

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**Abstracts**

**453 DETERMINING THE EFFECTIVENESS OF SYSTEMIC IMMUNOMODULATORY THERAPY IN THE TREATMENT OF PATIENTS WITH JUVENILE IDIOPATHIC ARTHRITIS ASSOCIATED UVEITIS DEPENDING ON THE CHOSEN OUTCOME MEASURES**

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**Introduction** The number of patients with juvenile idiopathic arthritis associated uveitis (JIA-U) who require systemic immunomodulatory treatment (IMT) for disease control is small. Variabilities in the patient selection and the results on the effectiveness of IMT make it difficult to compare studies. We aimed to show the difference between obtained levels of therapy efficacy on the same sample of JIA-U patients depending on the selected outcomes.

**Methods** The longitudinal observational study with JIA-U patients treated with IMT was conducted at University Hospital Centre Zagreb in the period from 2011 to 2017. We included 38 JIA-U patients aged 2 to 15 years and 69 eyes (7 patients had unilateral JIA-U).

**Results** At baseline 46 (66.7%) eyes had grade ≤1+ of inflammation in anterior chamber (AC) according to Standardization of Uveitis Nomenclature (SUN) Working group criteria, 11 (15.9%) had grade 2+, 3 (4.4%) had grade ≥3+, 23 children (60.5%) had already received methotrexate (MTX), 8 (21.0%) had grade 2+, 3 (4.4%) had grade 3+. 23 children of Uveitis Nomenclature (SUN) Working Group criteria, 11 patients treated with both biologics and MTX, achieved level of inflammation in AC. In the first 12 months, 3 (8.8%) eyes had a median of 4 daily doses, GC ointment in glucocorticoids (TGC) in the form of drops were used in 61 (88.4%) eyes with a median of 2-7 daily doses of TGC, and in the 48th month in 61.1%. According to milder criteria, in the 12th month 90% of the eyes have a degree of inflammation ≤0,5+ with ≤2 doses of TGC, and in the 48th month all patients achieved this goal.

**Conclusion** It was shown that the results of treatment outcomes during follow-up largely depend on the selected outcome measures. This will be important for future research because setting different limits can lead to a more favorable outcome.

**454 DIFFERENT HISTOLOGICAL CLASSIFICATIONS FOR IGA VASCULITIS NEPHRITIS – WHICH ONE HAS THE BEST ASSOCIATION WITH THE DISEASE OUTCOME?**

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**Introduction** IgA vasculitis nephritis (IgAVN) is almost the only cause of morbidity and mortality among children suffering from this most common childhood-vasculitis. Several histological classifications are used in the analysis of renal biopsy findings in IgAVN, but it remains unknown which one is the best predictor of severity and disease outcome.

**Objectives** The aim was to compare the four most commonly used histologic classifications for IgAVN and to establish which variables of each histological classification have the strongest association with unfavorable outcome.

**Methods** The cross-sectional study included 69 patients with IgAVN (diagnosed by EULAR/PRES/PRINTO criteria) and available renal biopsy specimens for analysis using the four histological classifications for IgAVN (the International Study of Kidney Disease in Children (ISKDC) classification, the Oxford classification, the Haas histologic classification of IgA nephropathy and the modified semi-quantitative classification (SQC), developed by Koskela et al.). The clinical outcome was defined through four categories, graded according to the modified classification of Cournahan (physical examination, hematuria, proteinuria, urine albumin-to-creatinine ratio, hypertension and eGFR). The linear relationships between outcome and histological classifications were analysed using ordinal regressions using the first-order of polynomial orthogonal contrasts.

**Results** The SQC classification proved to be the best, reducing the deviation of the model-predicted outcome value from the observed value by 9.5% (χ2=13,89, p<0,001), followed by the Oxford classification with a deviation reduction of 8.0% (χ2=11,76, p=0,001), then the ISKDC classification with a decrease in deviation of 3.3% (χ2=4,89, p=0,027). The worst was the Haas classification with a decrease in deviation of 2.1% (χ2=3,06, p=0,080). Analysis of individual variables of Oxford and SQC classifications showed that with increasing values in the variables of interstitial fibrosis (t66=3,23, p=0,002), tubular atrophy (t66=2,94, p=0,005) and tubular dilatation (t66=2,40, p=0,019) in the SQC classification, and endocapillary hypercellularity (t66=3,14, p=0,003) and crescents (t66=2,07, p=0,043) in the Oxford classification the outcome worsens.

**Conclusion** This study showed that the SQC classification has the strongest association with the IgAVN severity and outcome. Although crescents on renal biopsy were considered the most important outcome indicators, our study suggests that tubulointerstitial changes could be more important as predictors of poor outcome. Intestinal and renal tubules changes...
should be further explored in order to have better predictive values of IgAVN outcome and to be incorporated into existing or new classifications, on the basis of which guidelines for the treatment of patients would be developed.

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455 SINGLE NUCLEOTIDE POLYMORPHISMS OF GENES HMGB1 AND AGER AND ITS ASSOCIATION WITH CLINICAL FEATURES OF IGA VASCULITIS

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Introduction IgA vasculitis (IgAV) is a disease in which genetic background also plays an important role. Some small studies have indicated the importance of variants in various non-HLA genes in the manifestation of different disease phenotypes.

Objectives The aim of this research was to investigate single nucleotide polymorphisms (SNPs) of genes HMGB1 and AGER encoding for high mobility group box-1 (HMGB1) and receptor for advanced glycation end products (RAGE), both acting as mediators of inflammation, in the susceptibility and clinical features of patients with IgAV.

Methods HMGB1 and RAGE gene polymorphisms were genotyped using a real-time polymerase chain reaction. The presence and frequency of polymorphisms in HMGB1 (rs2249825, rs1045411, rs1060348, rs1412125 and rs41369348) and RAGE (rs1800625, rs1800624, rs2070600 and rs3134940) were determined. Clinical data were collected from database of IgAV patients from two Croatian University Centers for pediatric rheumatology.

Results 81 pediatric IgAV patients were included, of whom 45 were boys and 36 girls, as well as 150 age- and sex-matched healthy controls without any history of autoimmune disease. The median (range) age of IgAV patients was 6.25 (4.60-8.20) years, and among them 71.6% had joint involvement, 28.6% gastrointestinal manifestations, 27.16% developed nephritis. The purpuric rash which extended from lower extremities to the trunk, upper extremities and face (generalized rash) was present in 43.20% of patients and 27.16% had at least one relapse. Among the analyzed polymorphisms, only in the rs1412125 there was a statistically significant association with gastrointestinal involvement. The IgAV patients carrying the T allele (rs2070600) of the AGER had significantly increased risk of nephritis development compared with the IgAV patients with homozygous CC genotype in dominant (OR 2.78, CI 1.04-7.43, p = 0.04).