KD is currently unknown, according to the available literature and our experiences, it is important to point out that therapy for KD also leads to the resolution of lung problems and findings.

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ASSOCIATION BETWEEN GASTROINTESTINAL MANIFESTATIONS AND THE RISK OF RENAL DISEASE IN CHILDREN WITH IGA VASCULITIS

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Introduction IgA vasculitis (IgAV) is the most common childhood-vasculitis in which more than 50% of children develop gastrointestinal (GI) symptoms. In 10-20% of patients serious complications such as intussusception, bowel perforation, and massive bleeding may occur. The most important complication is the development of nephritis with progression to chronic renal failure in about 3% of children. The aim of the research was to analyze clinical and biochemical parameters in patients with IgAV and GI manifestations.

Methods This retrospective study included children with IgAV reviewed in five Croatian University Centers for pediatric rheumatology in the period 2009 to 2019.

Results Out of 611 children with IgAV, 320 were males and 291 were females. The overall GI symptoms prevalence was 45.9% and the median (range) age at diagnosis was 6.42 (4.5-8.83) years. Among patients with GI symptoms there were 1.44 times more males (N=166) than females (N=115), which was statistically significant ($p=0.003$). Patients with GI symptoms had less infections before the appearance of purpura (59.8% vs. 70.9%, $p=0.005$) and were found to be significantly more likely to have rash distributed on the trunk (61.9% vs. 48.5%, $p=0.001$), and upper extremities (35.2% vs. 24.7%, $p=0.006$), as well as generalized rash (38.8% vs. 28.3%, $p=0.008$). These patients also had significantly higher values of C-reactive protein, leukocyte count, erythrocytes and platelets, hemoglobin, hematocrit and D-dimer concentrations and lower levels of IgG and IgM. In our cohort 42 out of 281 children (14.9%) had the most severe GI manifestations (intussusception and/or massive GI bleeding) with significantly higher values of 24-hour urine protein levels and D-dimer concentrations and lower total serum protein, albumin, IgG, IgM and C3 levels in comparison with children whose GI manifestations were not severe. Predictors of severe GI involvement were: relapse of the disease, generalized rash, rash extended on upper extremities, rash extended to the face, recurrent rash and renal involvement, as well as lower values of prothrombin time, fibrinogen and IgM among the laboratory parameters. Patients with GI symptoms were 1.68 times more likely to develop nephritis, and this probability was 2.58 times higher if GI symptoms occurred before other symptoms. Other predictors of nephritis were: severe and

moderate GI manifestations, recurrent rash, one or more relapses of IgAV, and older age.

Conclusion Older children with IgAV and severe GI manifestations in whom IgAV begun with GI symptoms had a higher risk of acute and chronic complications of the disease.

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THE IMPACT OF SYSTEMIC IMMUNOMODULATORY THERAPY ON THE INTRAOCULAR INFLAMMATION AND THE NEED FOR TOPICAL GLUCOCORTICOID THERAPY IN PATIENTS WITH JUVENILE IDIOPATHIC ARTHRITIS- ASSOCIATED UVEITIS

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Introduction Juvenile idiopathic arthritis associated uveitis (JIA-U) is the most common and potentially most destructive extraarticular manifestation of JIA. The aim of this research was to determine the need for topical glucocorticoid therapy (TGC) in patients with JIA-U on systemic biological therapy in comparison to patients treated with methotrexate (MTX) only.

Methods We have conducted a longitudinal observational study with JIA-U patients in whom systemic immunomodulatory treatment (IMT: biologics and/or MTX) was introduced and who were followed at least 3 months in the period between 2011 and 2017. The data about the number of cells in the anterior chamber (AC) according to Standardization of Uveitis Nomenclature (SUN) Working Group criteria, related to TGC, systemic therapy and JIA complications were collected during each examination. Generalized linear mixed models were used to analyze the relationships between treatment with biologics, MTX, TGC and the grade of inflammation in AC according to SUN criteria.

Results 38 JIA-U patients (69 eyes) with median (range) age of 4.9 (2-15) years and follow up period of 209 (19-381) weeks were included. There were a total of 1205 examinations. At the first examination JIA-U was detected in 16 (42.1%) of patients, 59 (79.7%) of the eyes had $\leq 1+$ cells in the AC, and in 19 (50%) of JIA-U patients complications were already present. MTX was introduced in 23 (60.5%) JIA-U patients before the initiation of the study. 8 (21%) had already received biologics, while in 4 (10.5%) prior systemic glucocorticoids were also used. Before the completion of the study, all patients received MTX and 40% JIA-U were treated with biologics. The average number of TGC doses decreased from 3.74 at baseline to 0.72 in the 48th month. After Friedman and the post hoc test a significant difference in the daily doses of TGC could be seen from the 12th month after application of systemic IMT. The number of daily doses of TGC as well as the degree of inflammation in AC per eye decreased over time. Using generalized linear mixed models it was shown that the treatment with biologics, but not with MTX and systemic glucocorticoids, was associated with lower

intensity of TGC therapy. Treatments with biologics and systemic glucocorticoids, but not with MTX, were associated with lower degree of inflammation in AC.

Conclusion The results showed that the application of systemic biological therapy may result in less intensive TGC therapy, resulting in glucocorticoid-sparing potential, and reducing intraocular inflammation.

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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 $\leq 1+$ 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%) biologics, 4 (10.5%) systemic glucocorticoids (GC). Topical glucocorticoids (TGC) in the form of drops were used in 61 (88.4%) eyes with a median of 4 daily doses, GC ointment in 75.4% of the eyes with a median of 1 dose. Until the end of the follow-up, all children received MTX, and 40% of them biologics. The results of the effectiveness of IMT are presented according to the reduction of the need for TGC and achieved level of inflammation in AC. In the first 12 months, among JIA-U patients treated with both biologics and MTX, in 65% of eyes there was no need for TGC therapy, in the 48th month in 50% of eyes, while the rest required 1-2 daily doses of TGC. At the end of the first year, with MTX and biological therapy 75% of eyes had grade 0 of inflammation and in 48th month 61.1%. In the 12th month in 75% of eyes a 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 $\leq 0,5+$ 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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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 Counahan (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% ($\chi^2_{21}=13,89$, $p<0,001$), followed by the Oxford classification with a deviation reduction of 8.0% ($\chi^2_{21}=11,76$, $p=0,001$), then the ISKDC classification with a decrease in deviation of 3.3% ($\chi^2_{21}=4,89$, $p=0,027$). The worst was the Haas classification with a decrease in deviation of 2.1% ($\chi^2_{21}=3,06$, $p=0,080$). Analysis of individual variables of Oxford and SQC classifications showed that with increasing values in the variables of interstitial fibrosis ($t_{66}=3,23$, $p=0,002$), tubular atrophy ($t_{66}=2,94$, $p=0,005$) and tubular dilatation ($t_{66}=2,40$, $p=0,019$) in the SQC classification, and endocapillary hypercellularity ($t_{66}=3,14$, $p=0,003$) and crescents ($t_{66}=2,07$, $p=0,043$) 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. Interstitial and renal tubules changes