Goal Henoch-Schönlein purpura (HSP) is the most common vasculitis of the childhood. Among all possible symptoms/complications, nephritis (HSPN) is the main and almost only cause of morbidity and mortality in HSP. The aim of the study was to investigate the value of erythrocyte glutathione S-transferase (e-GST) activity as an early predictor of HSPN.

Methods Ninety-seven children with HSP were enrolled into the study. The control group consisted of 52 children without clinical and laboratory signs of inflammation. In all participants e-GST activity was determined spectrometrically from the whole blood samples, after incubation with a commercial GST assay.

Results At the beginning of the disease the e-GST activity was significantly higher in HSPN patients who developed the nephritis during the six month follow up period, compared to the group of HSP patients without signs of nephritis: median (interquartile range) 5,70 U/mgHb (4,38-7,50 U/mgHb) compared to 3,10 U/mgHb (2,20-4,20 U/mgHb); P<0,001. Similar results were obtained after the comparison of the HSPN patients and control group: 5,70 U/mgHb (4,38-7,50 U/mgHb) vs.3,13 U/mgHb (1,91-4,20 U/mgHb); P<0,001. There were no statistically significant differences between the group of HSP patients without nephritis and control group (P=0,837). During the follow up period, a significant decrease of e-GST activity was observed in the HSPN patients, but it was still significantly higher compared to the group of HSP patients without nephritis (P<0,001/P<0,001).

Conclusion e-GST activity is a reliable, independent marker of early nephritis risk assessment in children with HSP. As a sensitive, specific and feasible laboratory test, it has potential practical utility in the diagnostic algorithm and monitoring of the children with HSP.

440 SAPHO SYNDROME IN CHILDHOOD

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SAPHO syndrome is an acronym for 'Synovitis, Acne, Pustulosis, Hyperostosis and Osteitis'. It is combination of cutaneous and musculoskeletal manifestations, such as osteoarthritis and hyperostosis of bones of the anterior chest wall associated with acne fulminans and hidradenitis suppurativa. We present the case of a 15-year old boy with osteoarthritis of the left hip, osteitis of the left carpal bones and right clavicula with acne conglobata. Bone biopsy of the left hip has been carried out and Propionibacterium acnes was found in the bone lesion.

Identification of the Propionibacterium acnes from bone biopsy in SAPHO syndrome has occasionally been reported, which could suggest that microorganisms could be a trigger for osteitis and hyperostosis and may play the role in the pathogenesis of the disease. Our patient was treated with nonsteroidal anti-inflammatory drugs, antibiotics and a corticosteroids, which resulted in clinical improvement, but not complete remission, so we introduced methotrexate in therapy.

The purpose of this case study is to raise awareness to a set of clinical features of SAPHO syndrome and its early recognition and prompt therapy.

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

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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.

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

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.

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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