INTRODUCTION AND AIMS: Metalloprotease ADAMTS13 is a natural regulator of coagulation and complement hyperactivation. The low activity of the metalloprotease indicates the need for RRT, which is shown in patients with aHUS (71.4% vs 42.4%). The activity of ADAMTS13 was determined by the method of FRET (fluorescence resonance energy transfer).

METHODS: A total of 667 children were enrolled into the study, 220 of them were diagnosed with STEC-HUS, 441 with aHUS.

RESULTS: Above the cut-off value, ADAMTS13 activity was significantly higher in STEC-HUS (3613.5 ± 800.7 vs 2524 ± 420.3) than in aHUS (2524 ± 420.3 vs 206 ± 312.3). This was a result of different presentation of renal involvement: STEC-HUS group showed more often proteinuria, hematuria, hypertension, and anuria. In the aHUS group, the need for RRT was higher (87.3% vs 53.7%). Among patients with aHUS, in comparison to STEC-HUS, anemia was significantly more severe (50.43 ± 29.9 vs 44.3 ± 25), and the CD4/CD8 ratio was lower (0.75 ± 0.37 vs 0.56 ± 0.3). The proportion of patients with the development of various renal complications was higher, i.e. anuria (83.9% vs 35.7%), cortical necrosis (15.2% vs 5.5%).

CONCLUSIONS: Decrease in ADAMTS13 activity makes an additional contribution to the decompensation of the renal function. It is combined with the need for RRT, which is more often, and the development of severe complications such as anuria and cortical necrosis. The CD4/CD8 ratio and the severity of anemia are more severe than in control group (IgAN 4549.3 ± 688.7 vs 1688.7 ± 206 vs 19.5 days). With STEC-HUS, complications were observed after 20.46% (19-21) and 2.1 years (group II) were enrolled into the study. The age of the onset, IgA and time of the appearance of proteinuria, haematuria, creatinine and IgA, C3 levels, glomerular filtration rate (GFR) and recording the presence of hypertension (HT). Kidney biopsy findings are stated as the presence of mesangial proliferation, hypereosinophilia of mesangial cells, or focal and segmental glomerulosclerosis. The result was calculated as a sum of M, E, S, T. At the follow up the serum level of GDIgA1 and IgA/C3 ratio were associoated with a corresponding confidence interval for specific MEST scores. The aim of the study was to assess the significance of serum GDIgA1 and IgA/C3 ratio in children with nephrotic syndrome.

OUTCOMES IN CHILDREN WITH HENOCH-SCHONLEIN NEPHRITIS AND NEPHROTIC-RANGE PROTEINURIA

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INTRODUCTION AND AIMS: Henoch-Schönlein purpura (HSP) is the most common vasculitis in children. Its long-term prognosis depends on renal involvement. The management of Henoch-Schönlein purpura nephritis (HSPN) remains controversial. This study tries to characterize children with HSPN and nephrotic-range proteinuria treated initially by Pozzi protocol (intravenous bolus injections of 1 g Methylprednisolone for 3 days each at months 1, 3, and 5, followed by oral steroid 0.5 mg/kg prednisone on alternate days for 6 months).

METHODS: Our study included 18 children with HSPN and nephrotic-range proteinuria (> 40 mg/kg/m²) diagnosed between 2005 and 2016 in our Pediatric Nephrology Institute. All patients were treated initially by Pozzi protocol. Demographic, clinical,
laboratory and histological data were collected from the medical files. The primary endpoint was proteinuria remission defined as a proteinuria≤ 200 mg/24h.

**RESULTS:** The mean age of these 18 patients (9 girls, 9 boys) at the time of diagnosis was 9.5 years (range 3.2–17.3 years). Nine patients (50%) responded to Pozzi protocol treatment and maintain complete remission over time. The other 50% remained with persistent proteinuria and all were treated by Cyclophosphamide, in this group, 5 patients (55.6%) maintained complete remission and 4 patients (44.4%) patients remained with persistent proteinuria. Univariate analysis found a higher likelihood of response to Pozzi protocol in: older children 11.8±4.1 years (p = 0.016), in patients with longer period passed between skin manifestation (purpura) and nephrotic range proteinuria (median 30 [14.5–45] days), (p = 0.05), in patients with lower 24-hour urine protein to creatinine ratio (p = 0.036) and higher glomerular filtration rate (GFR) at presentation (p = 0.034).

**CONCLUSIONS:** Our study shows 50% response to treatment by Pozzi protocol in HSPN patients who presented with nephrotic range proteinuria. Time between skin and renal manifestations, age, GFR and 24-hour urine to creatinine ratio at presentation have been found to be predictive for treatment outcomes.