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ASSOCIATION BETWEEN IgA DEFICIENCY AND CELIAC DISEASE

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Relevance. Selective IgA deficiency (SIgAD) is the most frequent primary immune defect. It is defined as a serum IgA level below 0.07 g/L alongside normal IgM and IgG levels in an individual aged over 4 years old. Since SIgAD is not characterized by relevant infectious issues in most cases, it is often diagnosed during the diagnostic work up of several and different autoimmune disorders, which are associated with this primary immune defect.

Serum IgA levels are age-related: IgA is basically absent at birth and its concentration gradually increases during the pediatric age until reaching the adult levels during the adolescence (with normal levels ranging between 61 and 365 mg/dl). Total IgA deficiency is defined by serum IgA levels < 7 mg/dl. IgA deficiency is defined as partial when serum IgA levels are >7 mg/dl, but below the lower limit of the normal range according to the age.

Celiac disease (CD) is a gluten-related systemic immune-mediated disorder characterized by a very variable clinical expression, including both gastrointestinal and extra-gastrointestinal manifestations. It is diagnosed by the demonstration of specific autoantibodies, such as anti-tissue transglutaminase antibody and anti-endomysium antibody (which mainly belongs to the IgA isotype), along with the presence of atrophic (small bowel) enteropathy at the histopathological level.

Notably, SIgAD is significantly associated with CD, which can make the diagnosis of the latter disease be more difficult, since the main serological markers are IgA autoantibodies. Indeed, the assessment of total serum IgA concomitantly to the serological screening for CD is a mandatory test in the suspicion of CD.

Aim: to form the alertness of doctors regarding celiac disease in children with SIgAD.

Materials and methods. The object of the study was patient Z., 15 years old, undergoing treatment at the 3rd City Children's Clinical Hospital in Minsk. An analysis of the literature on the relationship between SIgAD and celiac disease was carried out, including the Pubmed and Uptodate databases.

Results and their discussion. Patient Z. was admitted to the hospital with the complaints on fatigue and stomach pain. CD was diagnosed based on the results of serological test that included a high level of tissue transglutaminase IgG and gliadin IgG, with normal levels of antibodies to these antigens of the IgA class. At the same time IgA deficiency was detected with total serum IgA – 0,04 g/l. Gastroscopy with obtaining biopsies from the duodenum and the bulb was perfomed. Crypt hypertrophy and villus atrophy were observed in the duodenal biopsies, and the diagnosis of celiac disease was substantiated. A gluten-free diet was initiated immediately, and the symptoms disappeared shortly after.

Conclusion: SIgAD is considered to be responsible for false negative IgA- tissue transglutaminase in CD patient in our study. Because of such false negatives, CD patients with SIgAD may elude diagnosis thereby lowering the apparent frequency of IgA deficiency in the CD population. Our finding that IgG based testing (Deamidated Gliadin Peptide (DGP)-IgA/IgG screen in this case) is useful for detection of CD in patients with SIgAD. DGP-IgA/IgG screen is also performed well in ruling out CD in patients with either SIgAD or partial IgA deficiency (PIgAD)**.**