

CONTENT OF TYPE I COLLAGEN ANTIBODIES AND THEIR ASSOCIATION WITH CLINICAL MANIFESTATIONS OF UNDIFFERENTIATED CONNECTIVE TISSUE DYSPLASIA

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Abstract: undifferentiated connective tissue dysplasia (UCTD) is considered one of the diseases of high medical and social significance, and practitioners are not sufficiently aware of the pathological values of UCTD as the background state of many pathologies. One of the features of development connective tissue dysplasia is the restructuring of connective tissue elements, the extracellular matrix, collagen fibers and elastin. The aim of study to identify the content of type I collagen antibodies and their association with clinical manifestations of undifferentiated connective tissue dysplasia. 48 patients with signs of UCTD (24 men and 23 women) were examined. All patients were subjected to general clinical, instrumental and laboratory studies. In the study of the level of auto antibodies to collagen type I, the average was $4.88 \pm 0.095 \mu\text{g/ml}$. A high concentration of titers of auto antibodies to type I collagen ($5.08 \pm 0.56 \mu\text{g/ml}$) was found in those patients ($n = 17$) who had complex clinical manifestations of musculoskeletal system lesions, joint hypermobility syndrome and small heart abnormalities. Thus, the increase in antibody level suggests the value of the autoimmune component in the pathogenesis of this disease, which will help us to carry out early diagnosis, and prevent possible complications. **Keywords:** undifferentiated connective tissue dysplasia, type I collagen antibodies, musculoskeletal system lesions, joint hypermobility syndrome, small heart abnormalities.

СОДЕРЖАНИЕ АНТИТЕЛ К КОЛЛАГЕНУ I ТИПА И ИХ СВЯЗЬ С КЛИНИЧЕСКИМИ ПРОЯВЛЕНИЯМИ НЕДИФФЕРЕНЦИРОВАННОЙ ДИСПЛАЗИИ СОЕДИНИТЕЛЬНОЙ ТКАНИ

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Аннотация: недифференцированная дисплазия соединительной ткани (НДСТ) считается одним из заболеваний, имеющих высокую медицинскую и социальную значимость, и практикующие врачи недостаточно осведомлены о патологических значениях НДСТ как о фоновом состоянии многих патологий. Одной из особенностей развития дисплазии соединительной ткани является перестройка элементов соединительной ткани, внеклеточного матрикса, коллагеновых волокон и эластина. Цель исследования - выявить содержание антител к коллагену I типа и их связь с клиническими проявлениями недифференцированной дисплазии соединительной ткани. Обследовано 48 пациентов с признаками НДСТ (24 мужчины и 23 женщины). Все пациенты были подвергнуты общеклиническим, инструментальным и лабораторным исследованиям. При исследовании уровня аутоантител к коллагену типа I среднее значение составило $4,88 \pm 0,095 \text{ мкг / мл}$. Высокая концентрация титров ауто антител к типу коллагена ($5,08 \pm 0,56 \text{ мкг / мл}$) было обнаружено у тех пациентов ($n = 17$), которые имели сложные клинические проявления опорно-двигательных аппаратов поражений, синдром гипермобильности суставов и малые аномалии сердца. Таким образом, повышение уровня антител свидетельствует о значении аутоиммунного компонента в патогенезе этого заболевания, что поможет нам провести раннюю диагностику и предотвратить возможные осложнения. **Ключевые слова:** недифференцированная дисплазия соединительной ткани, типа коллагена антитела, костно-мышечной системы поражения, синдром гипермобильности суставов, небольшие аномалии сердца.

Introduction. Undifferentiated connective tissue dysplasia (UCTD) is considered one of the diseases of high medical and social significance, and practitioners are not sufficiently aware of the pathological values of UCTD as the background state of many pathologies. In order to improve the quality of medical care for the population

during the years of independence, a program of transformation of the health care system has been implemented in our country and large-scale use of the latest diagnostic methods has been introduced. Thus, effective medical care for children and adolescents ensured timely detection, early diagnosis, treatment and prevention of possible complications from the cardiovascular system of patients with undifferentiated connective tissue dysplasia. Another important problem of organ dysfunctions in UCTD is the disruption of collagen synthesis found in most patients. The close relationship of UCTD with immune system function disorder has been proved. One of the features of development connective tissue dysplasia is the restructuring of connective tissue elements, the extracellular matrix, collagen fibers and elastin. In collagen metabolism, an immune response in the form of circulating auto antibodies arises, that is, a physiological process in which the life products of the connective tissue structures are eliminated - characterized by the level of activity of the immune system, its reduction or formation of autoimmune phenomena and even persisting immune reactions, indirectly indicate certain types of collagen involved in the pathological process against the background of UCTD.

Objective: Identify the content of type I collagen antibodies and their association with clinical manifestations of undifferentiated connective tissue dysplasia.

Methods: 48 patients (24 men and 23 women) were examined, on the basis of central medical association of Samarkand City and 1-clinic of Samarkand State medical institute with signs of UCTD. All patients were subjected to general clinical, instrumental and laboratory studies. Type I collagen is a fibrillar protein that forms the basis of the connective tissue. It is most found in skin, bones, tendons, cornea, etc. Determination of auto antibodies titers to type I collagen in blood plasma was determined by immune-enzyme assay using “Imtek” kits (Russia) according to the accompanying instructions (ELISA IEA).

Results: Among those surveyed, we identified the following indicators: the average age was 21.7; The BMI was about 22. The anthropometric and phenotypic characteristics of the patients studied showed that on average the circumference of the chest - $87, 69 \pm 8.15$ sm, the epigastral angle (in degrees 0) - $88, 29 \pm 9.74$, the length of the foot depending on the growth - 0.151 ± 0.08 , the height of the arch of the foot - 7.75 ± 1.15 sm respectively. Hyper mobility of joints (HS) according to the Beighton criteria the average degree was found about 26% of the studied. The podometric index on average consists of 28.87%, respectively. This suggests that most of the patients has foot flat. According to the ECG, a heart rhythm disorder was found by sinus tachycardia and bradycardia, and metabolic changes. We identified MVP, abnormally located chord, small heart abnormalities. In the study of the level of auto antibodies to collagen type I, the average was 4.88 ± 0.095 µg/ml.

In order to study the titers of auto antibodies to type I collagen depending on the phenotypic signs of UCTD, we distributed clinical signs into several groups, depending on the change in the level of auto antibodies. This has important clinical significance and makes it possible to carry out early diagnostics, to detect the progression of the musculoskeletal system - scoliosis, funnel-shaped chest deformation (FSCD), flatness and hyper mobility syndrome of joints, as well as small heart abnormalities.

Values of auto antibodies titers level in case of disorders of musculoskeletal system, hyper mobility syndrome, small heart abnormalities are given in Table 1.

Table 1. Values of auto antibodies with titers level in case of disorders of the musculoskeletal system

Groups of phenotypical signs (n=48)	Level of titers (mkg/ml)
Level of titers of auto antibodies to collagen type I with disorders of musculoskeletal system (n=12)	4,35±0,73
Level of titers of auto antibodies to collagen type I with disorders of musculoskeletal system + hyper mobility syndrome (n=11)	4,84±0,81
Level of titers of auto antibodies to collagen type I with small heart abnormalities signs (n=8)	4,60±0,59
Level of titers of auto antibodies to collagen type I with musculoskeletal system + hyper mobility syndrome + small heart abnormalities signs (n=17)	5,08±0,56

Based on this table, it is possible to compare the values of auto antibodies titers in different combinations of musculoskeletal system lesions, joint hyper mobility syndrome and small heart abnormalities. A high concentration of titers of auto antibodies to type I collagen ($5, 08 \pm 0.56$ µg/ml) was found in those patients (n = 17) who had complex clinical manifestations of musculoskeletal system lesions, joint hypermobility syndrome and small heart abnormalities.

These data indicate that there is a relationship between the clinical manifestations of HPCT and the values of the titers of autoantibodies to type I collagen in the plasma examined.

Conclusions. Thus, the increase in antibody level suggests the value of the autoimmune component in the pathogenesis of this disease, which will help us to carry out early diagnosis, and prevent possible complications. Early diagnosis of the UCTD in adolescents and young person’s makes it possible to take on a dispensary register and carry out secondary prevention in a family advisory polyclinic.

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