http://dx.doi.org/10.35630/2199-885X/2020/10/3//ed

EDITORIAL

Dear clinicians, research fellows, colleagues and friends!

Development and introduction of novel diagnostic techniques aimed at detecting heritable disorders in children are priority tasks of modern pediatrics. Identifying susceptibility to latent pathological conditions is crucial for prevention and interrupting their chronicity. In this connection the issues of studying heritable disorders such as connective tissue dysplasia (DCT) in children population are relevant and timely. DCT symptoms, which are frequently combined with a number of somatic diseases, dramatically affect their severity and course.

Differentiated hereditable diseases of connective tissue (Marfan syndrome, Ehlers-Danlos syndrome, Stickler syndrome, osteogenesis imperfecta) are well-studied. They have clinical phenotypes formed due to heritable disruptions in biosynthesis and collagen degradation on structural and metabolic levels. On the contrary, many facets of undifferentiated dysplasia remain understudied. These subjects, however, have attracted great interest in different medical areas including pulmonologists, cardiologists, ophthalmologists, pediatricians, orthopedic trauma physicians, nephrologists, gastroenterologists, sonographers.

Patients with undifferentiated forms of DCT represent a large heterogenic group encompassing simple as well as complicated conditions. Whereas phenotypic and clinical manifestations confirm the defect of connective tissue, on the other side, they do not match any known syndrome among genetically induced syndromes of mesenchimal cell deficiency. DCT is not a nosological unit, it presents an onotgenetically systemic progredient process resulting in structural and functional changes of organs and tissues. The variety of clinical symptoms in patients with connective tissues diseases indicates on systemic damage, as the connective tissue accounts for 50–80% of the body mass. Its functions (biomechanical, trophic, barrier, plastic, morphogenetic) are the major ones for a human body. Clinical manifestations of undifferentiated connective tissue are extremely varied and have a polysystemic nature. DCT is not a separate disease; it is a set of symptoms determined by an inherited qualitative or/and qualitative defect in protein synthesis defects.

There is dependence between external phenotypic manifestation of DCT and detected pathology of internal organs. Clinical systems of DCT may be displayed during years, whereas functional decline is progressing with the age of the patient.

Specialists observe DCT dependence with cardio-respirative syndromes, urological and gastroenterological diseases, bronchial asthma, and vegetative disorders.

It is scientifically proved that most of the craniofacial tissues are composed of connective tissues. Their structural and functional components play an active part in inflammatory, destructive and protective processes which develop at acute and chronic pathological conditions. There is evidence that CDT facilitates incidence of teeth and jaw abnormalities, diseases of periodontium, occlusion and temporomandibular joint.

In the current issue of our journal you may read about new data on pathogenetic mechanisms causing the development of craniofacial malformations. The authors have systemized external phenotypes and morphological features in children with different severity of CTD. It will allow forming groups of patients with a high risk of development of polyorgan pathology. Therefore, *pre-clinical* diagnosis in children aimed at early detection of latent hereditary diseases can reduce risk and frequency of disability; improve quality of life and social adaptation in children with CTD, as well as save the costs of medical treatment.

Executive Editor
Prof. Dmitry Domenyuk