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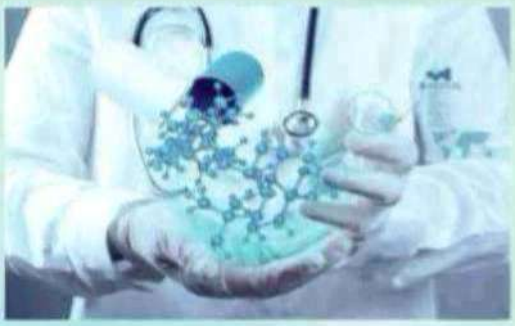

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EXPERT CRITERIA FOR THE FORENSIC DIAGNOSIS OF SUDDEN DEATH IN CHILDHOOD *(Literature review)*

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✓ ***Resume***

The current state of the problem is characterized by ambiguity of approaches to defining criteria for the forensic assessment of cases of sudden infant death. Existing methodological recommendations often do not take into account the full variety of morphological changes and laboratory parameters that are significant for expert assessment. There is no systematic approach to the assessment of risk factors and clinical and anamnestic data.

Keywords: forensic medical examination, sudden infant death, thanatology of childhood, pathomorphological diagnosis, infant mortality, postmortem diagnosis

ЭКСПЕРТНЫЕ КРИТЕРИИ СУДЕБНО-МЕДИЦИНСКОЙ ДИАГНОСТИКИ СЛУЧАЕВ ВНЕЗАПНОЙ СМЕРТИ В ДЕТСКОМ ВОЗРАСТЕ *(Литературный обзор)*

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✓ ***Резюме***

Современное состояние проблемы характеризуется неоднозначностью подходов к определению критериев судебно-медицинской оценки случаев внезапной детской смерти. Существующие методические рекомендации часто не учитывают всего многообразия морфологических изменений и лабораторных показателей, значимых для экспертной оценки. Отсутствует систематизированный подход к оценке факторов риска и клинико-анамнестических данных.

Ключевые слова: судебно-медицинская экспертиза, внезапная детская смерть, танатология детского возраста, патоморфологическая диагностика, детская смертность, посмертная диагностика

БОЛАЛАР ЁШИДА ТЎСАТДАН ЮЗАГА КЕЛГАН ЎЛИМ ҲОЛАТЛАРИНИНГ СУД-ТИББИЙ ТАШХИСЛАШ ЭКСПЕРТИЗА МЕЗОНЛАРИ (АДАБИЁТЛАР ШАРҲИ)

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✓ ***Резюме***

Муаммонинг ҳозирги ҳолати тўсатдан юз берган болалар ўлими ҳолатларини суд-тиббий баҳолаш мезонларини аниқлашда ёндашувларнинг турлича эканлиги билан тавсифланади. Мавжуд услубий тавсиялар кўпинча экспертиза баҳоси учун муҳим бўлган морфологик ўзгаришлар ва лаборатория кўрсаткичларининг барча хилма-хиллигини ҳисобга олмайди. Хавф омиллари ва клиник-анамнестик маълумотларни баҳолашда тизимли ёндашув мавжуд эмас.

Калит сўзлар: суд-тиббий экспертиза, болаларнинг тўсатдан ўлими, болалар танатологияси, патоморфологик ташхислаш, болалар ўлими, ўлимдан кейинги ташхислаш.

Relevance

The relevance of the study is due to the need to improve expert practice and the lack of uniform standardized approaches to the forensic medical assessment of cases of sudden infant death. The problem of sudden death of children remains one of the most urgent in modern forensic medicine and pediatrics. Despite significant advances in the development of medical science and practice, the incidence of sudden infant death remains consistently high, ranging from 0.5 to 1.0 per 1,000 live births in developed countries, according to WHO. The relevance of studying the criteria for the forensic assessment of cases of sudden death of children is due to a number of factors. Firstly, the complexity of the differential diagnosis of causes of death requires an integrated approach using modern research methods. Secondly, there is a lack of uniform standardized criteria for the expert assessment of such cases, which complicates the work of forensic medical experts and can lead to expert errors. Thirdly, the problem has a high social significance, since every case of sudden infant death requires a thorough investigation and determination of the true causes of the incident.

Sudden death (SC) is an urgent problem in forensic medicine. In some cases, physical activity is a provoking risk factor for sudden cardiac death [1]: with latent or existing diseases of the cardiovascular system, fatal complications may develop [2-4]. A significant proportion of the total number of young people, including school-age children. According to statistics, VS accounts for about 5% of cases [3]. In the USA, 5,000 to 7,000 such cases are registered annually, in Russia — from 1 to 13 per 100,000 people per year [5, 6]. The forensic diagnosis of the causes of sun in a group of young people causes difficulties due to the paucity of the morphological picture, the absence of pathology of the heart and blood vessels detected by routine laboratory and instrumental studies in medical institutions, as well as the speed and suddenness of its onset. The scientific literature actively discusses the pathogenesis of sudden infant death, the role of various factors in its development, and the possibilities of prevention. However, to date, morphological markers that allow an objective assessment of the causes of sudden death in children remain insufficiently studied. The methodology of conducting a forensic medical examination in such cases needs to be improved. Of particular importance is the development of a system of objective criteria for the forensic assessment of cases of sudden infant death. The need for such a system is conditioned by the need to standardize approaches to expert assessment, improve the quality and evidence of expert opinions, and improve preventive work**.**

Modern advances in morphological diagnostics, molecular genetics, and immunohistochemistry open up new opportunities for developing objective criteria for assessing cases of sudden infant death. The comprehensive use of these methods can significantly improve the accuracy of diagnosis and the validity of expert conclusions. Solving the problem of developing criteria for the forensic medical assessment of cases of sudden death of children is of great practical importance for improving expert activities, improving the quality of forensic medical examinations, and improving preventive work. This determines the relevance and practical significance of this research.

The sudden death of young people attracts the attention of doctors of various specialties - cardiologists, neurologists, pediatricians, sports medicine doctors. The establishment of the cause of death by the legislator in such cases is determined only through a forensic medical examination. The category of deceased persons, as a rule, reflects a young, able-bodied, reproductive and economically promising age - schoolchildren, students, cadets of higher military institutions, athletes or people engaged in sports activities. The circumstances of death in young people are most often active physical activity of various kinds. Death occurs against the background of complete "imaginary" health, without any pathology diagnosed during life. However, a forensic medical examination always reveals a pathology on the part of the cardiovascular system that is latent or not diagnosed in a timely manner. It was found that among all young people who died suddenly, signs of connective tissue dysplasia were revealed, which are detected both during external examination of the corpse and during internal examination. The article describes the main clinical and morphological manifestations of connective tissue dysplasia, pathology of various organs and systems, as well as the causes of sudden death of young people with this pathology.

The study of the causes of sudden death of young people is an urgent task of various fields of medical specialties — cardiologists, sports doctors, neurologists, pediatricians, as well as forensic medical experts and pathologists [1, 2]. The establishment of the causes of sudden death and signs of diseases that led to death is the responsibility of the forensic medical examination [1]. Forensic medical diagnostics of the causes of sudden death of young people of working age is aimed at identifying

pathological conditions or diseases that are not diagnosed during life, which were realized in conditions of provoking factors for the development of terminal conditions. The absence of any information about lifetime clinical observation, the absence of signs of chronic pathology of organs and systems in young subjects causes difficulties in determining the cause of sudden death [3]. Among the causes of death, the arrhythmic symptom complex, individual syndromes associated with valvular insufficiency, vascular syndrome, PE, and acute coronary insufficiency are most often described [4]. However, this list of pathological changes is much broader, most often they are in a compensated state and are not detected by standard clinical studies [5].

According to the definition recommended by the WHO Committee of Experts, "sudden death" (VS) refers to all cases of nonviolent death that occurred unexpectedly within 6 hours of the onset of symptoms of the disease that caused it, in people who had been practically healthy up to that point, or in patients whose condition was considered stable or improving. Worldwide, diseases of the cardiovascular system are the cause of sudden death in 60-80% [6]. And if the diagnosis of the causes of death of the elderly has been studied well enough, then establishing the true cause of SC in people of young working age is often problematic. In elderly people, 95 % of the cause of VS is acute myocardial ischemia, as an outcome of stenosing atherosclerosis or myocardial hypertrophy. The causes of sudden cardiac death in young people are various types of cardiomyopathies, pathology of the cardiac conduction system, latent myocarditis, aortic stenosis, aortic pathology with non-traumatic rupture in systemic connective tissue diseases [7, 8]. At the age of 15 to 18 years, the causes of SC are channelopathy (syndrome of elongated OT, Brugada syndrome), acute coronary insufficiency without atherosclerotic vascular lesion, anomalies of the coronary arteries, etc. [9].

In recent years, studies have been conducted and data have been obtained on pathological conditions caused by connective tissue dysplasia (DST). There are comprehensive studies and forensic diagnostics of the causes of sudden death of young people with this pathology. It has been established that in 83-87% of cases of sudden death, young people who have died show signs of connective tissue dysplasia [11].

Clinical studies have shown that the pathology of the cardiovascular system in people with DST is the most common, and cardiovascular disorders can lead to sudden cardiac death [10]. However, the widespread use of DST among people who consider themselves practically healthy makes the identification of this pathology, the early detection of its signs, and, consequently, the development of measures to prevent risk factors for sudden death relevant. The lack of long-term prospective clinical observations of patients with signs of DST makes it difficult to epidemiologically assess the prevalence of this pathology and assess the risk factors for sudden death in this pathology [10, 11].

Over the past 20 years, the problem of sudden death of people involved in sports activities, both amateur and high—performance sports, has been widely publicized. There are descriptions in the literature of cases of athletes' sun directly during training or immediately after-after that [12]. At the same time, the proportion of cardiac death in athletes is 10% higher than in people who do not engage in sports [12, 13]. The urgency of the problem has increased even more at the present time, as the number of young people aged 14-18 who are engaged in heavy sports is increasing everywhere, aiming at the formation of muscle relief (bodybuilding, weightlifting, athletic gymnastics, etc.) [13]. Among such cases, individuals with signs of DST are identified, who have the goal of increasing the small volume of muscle tissue in the presence of an asthenic type of constitution. However, high physical exertion, combined with increased psychophysical effort, is a side-causal risk factor for SC in this category of people [14].

In the study of the SUN in young people who died suddenly, changes in the heart during routine macro- and microscopic examination are practically absent or, as a rule, insignificant. The problem of sectional diagnosis in SC of young people is the need to interpret scant morphological changes in order to identify pathogenetic mechanisms that lead to death.

From the point of view of pathophysiological mechanisms, sudden cardiac death (SCD) can have two variants: 1) rapid cardiac arrest associated with ventricular fibrillation and secondary brain death; 2) slow cardiac arrest, resulting in gradual suppression of the activity of the cardiovascular system. The main mechanism of SCD in young people is arrhythmogenic - ventricular fibrillation (80%), less often — bradycardia or asystole (20%). The main causes of arrhythmic death are ventricular arrhythmias and intraventricular or atrioventricular blockages, leading to clinical death [5]. At the same time, coronary artery spasm and a sharp increase in myocardial oxygen demand (especially during physical exertion),

changes in nervous regulation and an increase in myocardial sensitivity to ischemia contribute to the development of arrhythmic conditions. Acute coronary insufficiency, characteristic of people under the age of 40, is often complicated by ventricular fibrillation or electromechanical dissociation, which are also leading to death [8].

The study of connective tissue dysplasia has been comprehensively developed over the past 15 years [6]. A systematic clinical approach to the study of this pathology and separate epidemiological studies have shown that individual phenotypic manifestations of DST, starting from puberty, occur in young people in 0.6—80.0% of cases. Works dedicated to-

With regard to sudden death and the forensic diagnosis of its causes, it is indicated that people with signs of DST die prematurely at the age of 40, against the background of physical exertion [11]. These circumstances dictate the need for a comprehensive comprehensive study of this pathology at both the clinical and morphological levels.

According to literature data, connective tissue dysplasia (DST) (dis — disorder, plasio — development, education) is a violation of the development of connective tissue in the embryonic and postnatal periods due to genetically altered fibrillogenesis of the extracellular matrix, leading to a disorder of homeostasis at the tissue, organ and organizational levels in the form of various morphofunctional disorders of visceral and locomotor organs with progressive course. DST is a genetically determined systemic progressive process. The group concept of DST includes genetic diseases that differ in clinical characteristics— such as Marfan syndrome, Ehlers—Danlos syndrome, and osteogenesis imperfecta. Signs of DST are also found in some chromosomal diseases, such as Down syndrome, Shereshevsky—Turner syndrome, and Klinefelter syndrome. Recently, not only primary hereditary connective tissue diseases have been identified and described, but also undifferentiated forms of DST — MASS factor. This group usually includes congenital deformities of the chest, spine, varicose veins, pathology of the skin, joints, and other external signs [8]. However, it has been established that with severe clinical syndromes of DST and with undifferentiated variants, type I collagen damage is observed. Type I collagen is the main component of collagen in the skin, arteries, and bones, which reflects the high phenotypic and organ manifestations of DST. The wide variety of clinical and morphological manifestations of DST is due to the fact that type I collagen damage can capture the extracellular matrix in the body as a whole. However, DST also disrupts another fibrous component of connective tissue, elastin, whose abnormalities were detected in Marfan syndrome with a normal collagen content. Damage to elastin fibrils has been described in cutis laxa and Ehlers—Danlos syndrome [9]. In addition, the cause of the clinical and morphological manifestations of DST are abnormalities of structural proteins and protein-carbohydrate complexes of connective tissue. For example, osteogenesis imperfecta revealed a decrease in osteonectin, proteoglycans, and sialoproteins. The consequence of a defect in the fibronectin molecule that binds collagen fibers to glycoproteins is the clinical manifestations of Ehlers—Danlos syndrome type X, abnormal fibronectin is also found in type II of this syndrome [12].

The results of biochemical studies have shown that the basis of "non-syndromic" forms of DST, such as mitral valve prolapse, hyperelasticity of the skin, deformities of the chest and spine, hypermobility of joints, are genetically determined disorders of collagen, elastin, fibronectin, proteoglycans. At the same time, the inferiority of type V collagen and additional type III collagen, which is produced by fibroblasts of the myxomatously altered mitral valve, was revealed.

In addition, a violation of the processes of fibrillogenesis may be the result of a defect in various enzymes that determine the regenerative potencies of connective tissue [10].

Thus, diseases associated with congenital pathology of connective tissue are based on molecular disorders that lead to changes in the structure and function of connective tissue. DST is morphologically characterized by structural changes in collagen and elastic fibrils, disorders of glycoproteins, proteoglycans, and fibroblasts, which leads to disorganization of the matrix at the fiber levels, fiber bundle, and tissue as a whole. The specific phenotype of an individual, the severity of clinical and morphological manifestations of DST depend on the level of genetic damage to the elements of connective tissue. The systemic nature of the lesion in DST is due to the widespread distribution of connective tissue in the body, which makes up the stroma of internal organs, forms the musculoskeletal system and the outline of the body. The nature of damage to internal organs and systems is caused by violations of their own parenchymal-stromal relationships [3].

The absence of any complaints or clinical manifestations in patients with signs of DST at a certain age period reflects only the effectiveness of compensatory mechanisms. However, this does not mean that such a patient has no pathology of internal organs. Sectional observations show that in individuals with external signs of DST, autopsy reveals serious disorders of the heart, blood vessels, and other organs and systems [11]. Hence, there is a need for a comprehensive clinical approach in the search for stigmas of connective tissue dysplasia in young people. Connective tissue dysplasia has a number of syndromes, which can be diagnosed both at the clinical level and by pathomorphological examination at the onset of sudden death. Therefore, the integration of knowledge on the main manifestations of DST is important primarily for the prevention of the development of terminal processes and risk factors for sudden death among young people.

Pathology of the musculoskeletal system. Musculoskeletal pathology and joint changes in DST are the defining and leading links in the formation of anatomical and structural prerequisites for the pathology of internal organs. The characteristic outline of the body, the constitutional type, characterized by tall stature, asthenic type of constitution, and the characteristic shape of the chest (flat, long, narrow, with a rib angle of less than 90°), creates the classic phenotype of a subject with DST syndrome. Developmental disorders of the upper and lower extremities are represented by elongation of the arms, dolichostenomy, arachnodactyly, curvature of the legs, clubfoot, and the presence of various types of flat feet. The detection of pathology from the sternum — funnel-shaped, keeled, as well as spinal pathology — scoliosis, lordosis, combined forms, straight back complement the clinical picture of a patient with signs of a differentiated or undifferentiated form of DST. Skeletal pathology is the basis for the formation of thoracophrenic syndrome, which is clinically manifested by hyperfunction of the respiratory muscles, disorders of anatomical and topographic relationships with a decrease in the volume of pleural cavities and, as a result, a violation of the structural and functional parameters of the bronchopulmonary system. Dysplastic-dependent changes in the musculoskeletal system are extra-cardiac factors that negatively affect the functioning of the circulatory system as a whole. Deformities of the chest and spine lead to mechanical compression of the heart and displacement of blood vessels. With minor sternal disorders, the heart undergoes compression, and in cases of severe bone deformities, direct mechanical action occurs on the heart, with its rotation and displacement to the left, with a decrease in the lumen and torsion of large vessels. Prolonged functioning of the heart in unfavorable conditions leads to the development of metabolic disorders in the myocardium, as well as energy depletion of cardiomyocytes, subsequently creating a substrate for the development of myocardiodystrophy, heart failure, rhythm disturbances and cardiac conduction.

External signs of bone skeleton pathology, detected both during clinical examination of patients and during autopsy examination, are confirmed by characteristic morphological changes detected during examination of the cartilaginous part of the ribs and sternum: internal tortuosity-

There was a violation of the three-dimensional vessels, a violation of their orientation, and a bone of a desmal structure was determined at the junction with the cartilage. Thinning of the cortical layer of the sternum was revealed. Thinning of bone beams was observed in the spongy structures, and accumulations of fat cells were observed in the inter—girder cells. Attention was drawn to a slightly smaller volume of red bone marrow compared to the average age norm. When examining the hyaline cartilage of the ribs, there was a violation of the order of alternating layers of hyaline cartilage, and alcyanophilia of osteocytes and osteoblasts was also noted in bone structures. Vacuoles with a positive reaction to mucus were detected in chondrocytes and chondroblasts. The disordered nature of the isogenic groups also attracted attention. An active CHIC+ reaction was observed around them, paling when treated with testicular hyaluronidase. A positive reaction with alcyan blue was observed, reflecting an increase in depolymerized sulfated and unsulfated glycosaminoglycans in cartilage structures.

Cardiovascular pathology. Pathology of the sternum and spine with a violation of the shaping of the chest as a whole and functional changes detected during life on the part of the respiratory system and the cardiovascular system form a cardiovascular syndrome, the severity of which is proportional to the degree of violation of musculoskeletal structures. Pathology of the sternum and spine in DST leads to structural changes in the heart and large vessels — the aorta, pulmonary trunk. Violation of the formation of the heart in people with DST is manifested by a violation of its position and shape: autopsy examination of sudden death of young people reveals hypoplasia of the heart (weighing less than 240 g), its vertical position, the predominance of vertical dimensions, torsion or partial torsion of the heart is diagnosed, which during life contributes to pronounced hemodynamic disorders. Valve pathology in

individuals with DST is represented by mitral valve prolapse, aortic valves, and pulmonary artery, with characteristic histological signs in the form of mitral valve myxomatosis and decreased collagen structures, which is the morphological substrate of valve prolapse. Thus, the functioning of the cardiovascular system in DST occurs in unfavorable intra- and extracardial conditions, which is due to anatomical, structural and morphological features.

The pathology of the aorta in people with DST is the most striking and is always revealed in the study of cases of sudden death in the form of general hypoplasia, decreased diastasis, decreased aortic wall thickness and the formation of ascending aneurysms or arches. Ruptures of the pathologically altered aorta at the height of physical activity with the development of hemorrhagic shock are the cause of sudden death in people with DST. Morphological examination of the aorta in Marfan syndrome revealed uneven thinning of its wall, loosening of the endothelial layer, its rarefaction and fragmentation, focal absence of elastic fibers, their thinning, a decrease in the number of smooth muscle fibers surrounded by connective tissue with pseudocysts containing a metachromous mucoid substance, the formation of cavities in the media area, its necrosis periarterial fibrosis. Similar changes were found in the aorta and the main arteries of the large circulatory system. In the area of the aortic aneurysm, changes in the feeding vessels are detected — the intrahepatic vessels, defined in the middle shell, are worn out and dilated, often with perivascular hemorrhages. Sometimes perivascular, mainly lymphohistiocytic infiltrates are detected in adventitia and the media, which gives a resemblance to nonspecific aortitis. With pathological tortuosity and looping of the carotid arteries in the looping zone, inflammatory processes in the walls of the vasa vasorum are also noted, obviously of a traumatic (mechanical) nature due to constant pulsation. In this case, trophic disorders of the vessel lead to early dystrophic and necrobiotic changes in its wall, followed by replacement with coarse fibrous connective tissue and the development of an aneurysm [5].

In individuals with DST, pathology of extra- and intracerebral vessels is revealed. Such factors as arterial hypertension, stressful or traumatic situation, high physical activity, etc. can contribute to the rupture of a pathologically altered wall. Pathology of cerebral vessels is combined with external and internal (organ) signs of DST, represented by a violation of the shape, structure of the vessel and a change in the morphology of the vascular wall. The pathogenesis of arterial aneurysm formation consists of two components: the effects of hemodynamic factors and the characteristics of the resistive properties of the vascular wall. The combination of impaired resistive properties of the vascular wall and the effects of hemodynamic factors (for example, a sudden sharp increase in blood pressure with acute excessive skeletal muscle tension, high blood flow velocity) largely determine the progressive aneurysm of the arteries in DST, potentially dangerous rupture of their walls with fatal outcome [4].

Modern descriptions of muscular and mixed types of arteries (carotid, renal, cerebral) with connective tissue dysplasia indicate the existence of several types of their damage: bifurcation-hemodynamic aneurysms, tubular aneurysms, arterial dolichoectasia (fusiform shape), pathological tortuosity (up to looping). Morphological examination of vascular aneurysms in DST shows thinning of its wall with the phenomena of uneven intimal thickening with fibroelastic phenomena, with thinning of the middle shell and its focal atrophy, signs of hyperelastosis and fragmentation. As a result, the folding of the inner elastic membrane increases. In the study, the aneurysm wall is usually devoid of blood vessels and consists of tightly packed bundles of collagen fibers with scant cellular elements. In about 2/3 of cases, rupture of an aneurysm of the arteries of the base of the brain is preceded by an intrahepatic exfoliating hemorrhage. The formation of cerebral vascular aneurysms in people with DST occurs over the age of 15, when risk factors such as hypertension, hemodynamic disorders and lifestyle changes (increased physical activity) are added. The main morphological criterion for the diagnosis of congenital vascular wall pathology is a violation of the structure and order of elastic membranes, a congenital pathology of the muscular layer of the vessel, which is one of the leading mechanisms of aneurysm formation in hemodynamic and regulatory disorders. These circumstances increase the capacitive function of the vessel, lead either to the formation of aneurysms, or to uneven narrowing and expansion of the vessel, with a high risk of rupture of the pathologically altered vascular wall in conditions of provoking factors, with the development of hemorrhagic infarction, subarachnoid hemorrhage and the onset of sudden death. Thus, lesions of the cardiovascular system in DST are dominant in the study of sudden death cases. Pronounced and widespread changes in the heart and blood vessels are noted in severe systemic forms of DST — Marfan syndrome, Ehler-sa—Danlos syndrome, etc. However, isolated lesions of the heart (mitral valve prolapse) and blood vessels are also recorded,

especially in young people. As DST progresses, there is a progressive qualitative and quantitative increase in pathological changes in the heart and blood vessels, leading to the development of cardiovascular disorders. The pathology of the heart and blood vessels of persons with DST during autopsy is detected in 83% of cases, however, this pathology has not been diagnosed in adolescents and young people. persons during their lifetime. When provoking factors were added, the existing structural disorders led to the development of terminal symptom complexes and fatal arrhythmias. Changes in the heart contribute to the development of arrhythmic syndrome, impaired innervation of the heart and blood circulation, which cause the onset of instant cardiac death. Dilated changes in the heart cavities during physical exertion (sudden sports death) are the cause of a decrease in the contractility of the myocardium, its electrical instability and the development of various types of arrhythmias. Pathology of arterial vessels — from aneurysms caused by a violation of the elasticity of the vascular wall, to changes in shape and position, and structural disorders of the vascular wall — can cause the main cause of death when the pathologically altered wall ruptures. Venous vascular pathology, which also occurs within the framework of DST, is fatal only at the level of the central nervous system and is realized by pulmonary embolism as the cause of sudden death. In other cases, it only increases the deposition of blood in certain localizations.

Pathology of respiratory organs. In people with DST, this pathology is clinically represented by bronchopulmonary syndrome in the form of tracheobronchial dyskinesia, bullous emphysema, recurrent chronic bronchitis with an asthmatic component and the presence of signs of hyperventilation syndrome. The causes of death in such individuals are spontaneous pneumothorax and hemothorax, the development of focal bronchopneumonia on the background of immunological insufficiency syndrome and pulmonary heart failure in the presence of thoracodiaphragmal syndrome in combination with heart pathology. Functional changes in the bronchopulmonary system in DST are caused by a violation of the structure of the musculocartilage framework, both the tracheobronchial tree and lung tissue, which increases their elasticity, with the formation of signs of tracheobronchomegaly, tracheobronchomalacia and bronchiectasia. "Changes in the tracheobronchial tree are accompanied by dyskinesia of the airway wall (inflating on inspiration and collapse on exhalation), which contributes to a delay in the evacuation of secretions. It is these mechanisms that form the bronchopulmonary syndrome. Violations of the functions of the bronchial tree caused by these factors contribute to the infection and chronization of the inflammatory process. Fibrosclerotic changes that develop in this case worsen the valvular mechanism of bronchial obstruction" [7].

Pathology of the gastrointestinal tract (GIT). In people with DST, pathology of the gastrointestinal tract is usually not the main leading pathology and the cause of sudden death, but in all cases there is a definite relationship between DST and the severity of the pathology of the digestive system. The pathomorphological picture of the lesion of the digestive tract is of a syndromic nature and does not have a specific nosological outline. Cases of manifestation of lesions of the gastrointestinal tract, which directly caused death, are described, such as bleeding from rupture of an aneurysm of the esophageal artery, perforation of a small curvature ulcer of the stomach and bleeding from the vessels of the wall of the diverticulum of the colon. Disorders of parenchymal-stromal relationships in individuals with DST are manifested by the presence of gastroptosis, dolichosigma, diverticulum of the colon, atypical position of the gallbladder with incomplete torsion of the neck and cystic duct and its horizontal position with significant elongation of the neck of the bladder. When examining the bile ducts, their elongation is observed with thinning and some tortuosity of the bile ducts. All changes in the biliary tract are accompanied by signs of atony of the walls, which indicated the presence of intravital dyskinesia of the gallbladder and biliary tract. Bile stasis and calculous cholecystitis are also constant signs identified during the study of cases of sudden death of people with signs of DST. In a sectional study of the sudden death of young people with signs of DST, it is necessary to determine the relationship of gastrointestinal tract damage with connective tissue dysplasia in order to assess the role of this pathology in the onset of death [1].

Pathomorphology of urinary organs. An analysis of all sectional cases showed that the spectrum of kidney damage in DST is quite wide. Pathological changes affect almost all structural and functional units of the kidneys. Kidney pathology in people with DST in sudden death was characterized by a violation of the anatomical shape, topography, structure and, consequently, organ function. In addition, there were abnormalities of the renal vessels, which were the main and triggering link in the occurrence of vasorenal hypertension. The most frequently detected pathology was represented by nephroptosis,

the displacement of the kidneys ranged from 8 to 15-20 cm, and the degree of organ prolapse varied depending on the age of the patient: the older the subject, the more pronounced the degree of nephroptosis. In individuals with pronounced skeletopias, signs of kidney dystopia are revealed — lumbar, iliac, and pelvic dystopia. All types of dystopias were accompanied by structural changes in the ureters, renal vessels, and nephron changes. The phenomena of nephroptosis and dystopia have always been accompanied by changes in the shape and size of the organ, as well as varying degrees of rotation. Morphometric examination of the kidneys in individuals with syndromic forms of DST (Marfan syndrome, Ehlers—Danlos syndrome, osteogenesis imperfecta) revealed doubling and incomplete doubling of the kidneys with signs of doubling and hypoplasia of the calyx-lobular system. This pathology of the calyx-lobular system in all cases was accompanied by complications expressed to varying degrees: pyelonephritis, secondary stone formation, sometimes with the development of hydronephrosis. With osteogenesis imperfecta, obstructive processes along the urinary tract were detected in the form of ureteral abnormalities with torsion. Dysplastic changes also affected the renal vessels, which are represented by general tortuosity, hypoplasia, and sometimes with the formation of aneurysmal protrusions. In all cases, there was an expansion of the diameter of the aorta at the level of the outlet of the renal arteries, as well as an increase in the diameter of the renal arteries and veins. The main pathology of the kidneys in people with DST most often leads to the development of arterial hypertension, which, together with arrhythmic and other cardiac syndromes of DST, forms the basis of the thanatogenesis of sudden death. The presence of chronic inflammatory changes in the kidneys and ureteral segment, such as calculous or stone-free pyelonephritis, is also important in the development of latent hypertension syndrome, but which collectively aggravates the course of cardiac DST syndromes [11]. Thus, the kidney pathology of people with DST in sudden death is significantly higher than in people without connective tissue pathology.

Clinical syndromes in people with DST, diagnosed both in children, adolescents, and over the age of 18, are the syndrome of neurological disorders and mental disorders, manifested in autonomic dysfunction, neurotic and depressive states. When examining the neurological status of people with DST, signs of increased anxiety, unmotivated fear, the presence of various phobias, hypochondriac mood and a general decrease in the emotional background are revealed. Arterial hypertension syndrome usually reflects pathology from both the musculoskeletal system, the cardiovascular system, and the kidneys.

The study of the obstetric and gynecological status in people with DST revealed a whole range of pathological disorders, both in adolescent girls in the form of delayed formation of secondary sexual characteristics, the development of juvenile uterine bleeding and menstrual cycle disorders, and the pathology of women of reproductive age — genitourinary prolapse.-

waistline, pathology of pregnancy and childbirth (rapid and rapid delivery, premature birth, high frequency of spontaneous abortions) [2].

Conclusions

Thus, connective tissue dysplasia is a systemic pathology that covers almost all organs and systems, with progressive development starting from puberty. This pathology, depending on the severity and organ specificity, is a risk factor for sudden death, primarily of young people of working age who lead an active lifestyle and consider themselves practically healthy. Timely detection of signs of DST at the clinical level will prevent the development of life-threatening complications and adjust the lifestyle of such a subject. An integrated approach to the study of DST will make it possible to develop forensic criteria for the diagnosis of sudden death in this pathology. The need to develop a scientifically based system of criteria for the forensic medical assessment of cases of sudden death of children, improve methodological approaches to conducting examinations and standardize expert activities determines the relevance of the chosen research topic and its practical significance for forensic medical examination.

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