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Pathomorphology of Vascular Bed in Different Variants of Arrhythmogenic Heart Development

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A total of 200 hearts from patients with various forms of cardiosclerosis and pronounced disorders of the heart rhythm were examined postmortem by contrast polypositional cardioventriculography, coronarography, volume and weight cardiometry, and morphometry. Left-ventricular, right-ventricular, and septal variants of arrhythmogenic heart development were distinguished. Left-ventricular variant is characterized by compensatory restructuring of the vascular bed with appreciably increased volume of vascular density mainly in the left ventricle and with the median left type of blood supply. The right-ventricular variant is characterized by signs of compensation and decompensation of circulation and intensification of the right ventricular vascularization. The septal variant is characterized by signs of vascular bed decompensation with more intense vascularization of the septum. The detected diagnostic criteria indicate appreciable structural rearrangement of the coronary bed of arrhythmogenic heart, which explains the essential shifts in stimulation, contraction, and mechanical characteristics of the heart ventricles.

Key Words: *arrhythmogenic heart; vascular bed; coronarography; cardioventriculography; volume and weight cardiometry*

Arrhythmogenic heart (AH) is now regarded as an organ characterized by intricate and deep structural rearrangement of the myocardium and nervous system in response to lasting and variegated disorders in the rhythmic activity [5]. Changes in the vascular bed play an important role in the multifactorial process of AH formation. Vascular disorders should be taken into consideration in the diagnosis of the patho- and thanatogenesis. Such investigations are very important because relatively individual variants of AH development were recently detected and each of these variants is characterized by specific pathognomonic pathomorphological features.

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We studied changes in the vascular bed in different variants of AH development and distinguished the criteria for pathomorphological diagnosis and some thanatogenesis factors.

MATERIALS AND METHODS

A total of 200 hearts from dead patients (mean age 56.4±0.4 years) with various forms of cardiosclerosis, in whom pronounced arrhythmias were diagnosed during lifetime (during 10.2±0.2 years) were examined. Ectopic arrhythmias (extrasystole and paroxysmal (extrasystolic) tachicardia) were recorded in 112 cases, atrial and ventricular flutter and fibrillation in 28 cases. Various conduction disorders were detected in 60 cases (sinoatrial blockade in 28 cases, atrioventricular block in 20, and intraventricular blockade in 12 cases). For control, 200 hearts of subjects of the same age and

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sex dead from accidental causes without history of cardiac rhythm disorders were examined.

The vascular bed of AH was studied by the method of postmortem contrast polypositional coronarography in our modification and by this method in combination with the standard anatomical method recommended by WHO [2]. Volume density of the vascular bed in the main compartments of AH was estimated and types of its blood supply were studied. The main morphofunctional parameters of AH were evaluated by morphometrical methods [4]: indexes of blood supply to the heart and myocardium (ratio of total and net weight of the heart to summary area of coronary arteries lumen) [2]. The status and volume of the cardiac cavities were evaluated by our method of postmortem contrast polypositional cardioventriculography in combination with volume-weight and planimetric cardiometry [3]. Topographoanatomical peculiarities of the vascular map of AH necessitated visualization of the conduction system by micropreparation under a stereomicroscope with magnification of up to ×36 [1]. Special section of the hearts was made in order to detect additional conduction routes and nodal tracts [6,7]. The detected changes in the vascular bed and the morphofunctional parameters of AH were analyzed for each of the variants of AH development (left-ventricular, right-ventricular, and septal). Quantitative data were statistically processed using Student's *t* test and alternative variation.

RESULTS

Detailed cardioventriculographic analysis of the hearts in the main group in comparison with the control showed 3 variants of AH development: left-ventricular, right-ventricular, and septal. Left-ventricular variant was most incident (92 cases, 46%), right-ventricular (62 cases, 31%) and septal (46 cases, 23%) variants of AH development were less common.

The detected variants of AH development were characterized by different volume density of the vascular network in the main compartments of the heart (Table 1). Left-ventricular variant was associated with an appreciable increase in the volume density of vessels in all walls and in the left ventricle in general (Table 1). Right-ventricular variant of AH was characterized by selective increase in the volume density of the vascular network in the right ventricle. The septal variant was characterized by an appreciable increase in the volume density of vessels in the ventricular septum of AH.

The relationship between the detected variants of AH and types of blood supply to the heart was analyzed (Table 2). Predominance (56.5% cases) of median left blood supply was pathognomonic for the left-ventricular variant. Right-ventricular variant of AH was most often (67.7% cases) associated with the median right blood supply, while the median blood supply predominated (56.5% cases) in cases with the septal variant.

Volume and weight cardiometry with individual weighing of individual parts of the heart in combination with the standard WHO anatomical method for studies of coronary arteries abnormalities showed extremely important morphofunctional values allowing evaluation of the adequacy of the coronary bed to AH weight (Table 1). Index of cardiac blood supply (ratio of total heart weight to summary area of coronary arteries lumen) in left-ventricular variant of AH corresponded to the criteria transitional from health to disease, in right-ventricular variant this index corresponded to criteria of disease, and in septal AH the index showed severe disease.

The index of myocardial blood supply (weight of AH myocardium (g) per square millimeter of coronary

TABLE 1. Volume Density of Vessels (%) and Indexes of Blood Supply to the Heart and Myocardium in Different Variants of AH Development (*M*±*m*)

Heart compartment and blood supply index	Control	AH variants		
		left-ventricular	right-ventricular	septal
Left ventricle	68.8±2.4	80.4±1.6*	60.4±2.4*	64.4±2.2
Anterior wall	54.2±1.6	58.6±1.6	52.2±1.2	52.4±1.4
Posterior wall	50.2±1.4	52.2±1.4	48.4±1.6	46.6±1.2
Lateral wall	34.2±1.2	36.6±1.2	32.2±1.8	30.2±1.4
Ventricular septum	44.2±1.4	40.4±1.6	38.6±1.4*	54.2±1.2*
Right ventricle	50.2±1.2	36.6±1.4*	66.4±1.8*	46.6±1.8
Index of cardiac blood supply, g/mm^2	16.8	22.4	25.4	26.8
Index of myocardial blood supply, g/mm^2	16.6	18.8	21.2	22.6

Note. *$p<0.01$ compared to the control.

TABLE 2. Correlation of Types of Myocardial Blood Supply and Variants of AH Development

Blood supply index	Control	AH variants		
		left-ventricular	right-ventricular	septal
Left	32 (16)	20 (21.7)	—	—
Right	20 (10)	—	12 (19.4)	—
Median	46 (23)	16 (17.4)	6 (9.7)	26 (56.5)
Median left	62 (31)	52 (56.5)	2 (3.5)	12 (26.1)
Median right	40 (20)	4 (4.4)	42 (67.7)	8 (17.4)
Total	200 (100)	92 (100)	62 (100)	46 (100)

Note. The percentage of cases is shown in parentheses.

artery lumen) indicated satisfactory blood supply to the myocardium in left-ventricular AH variant and differed negligibly from the upper threshold normal value (18.1 g/mm^2). In the right-ventricular AH variant this parameter corresponded to the lower boundary of the pathology, while in the septal variant the index reflected deep pathological shifts in blood supply to the myocardium (Table 1).

Hence, indexes of blood supply to the heart and myocardium indicated that the coronary bed more or less corresponded to AH weight in left-ventricular variant. Right-ventricular variant was characterized by sufficient throughput capacity of coronary vessels. In septal variant of AH the hemodynamics was sharply modulated because of decreased volume density of the vascular network [8]. This decrease was responsible for disorders in myocardial contractility and mechanical characteristics manifesting in prolongation of isovolumic contraction and shortening of the ejection time. Pronounced disorders in coronary blood flow in the septal zone usually lead to asynchronic stimulation and contraction of AH ventricles [10,15].

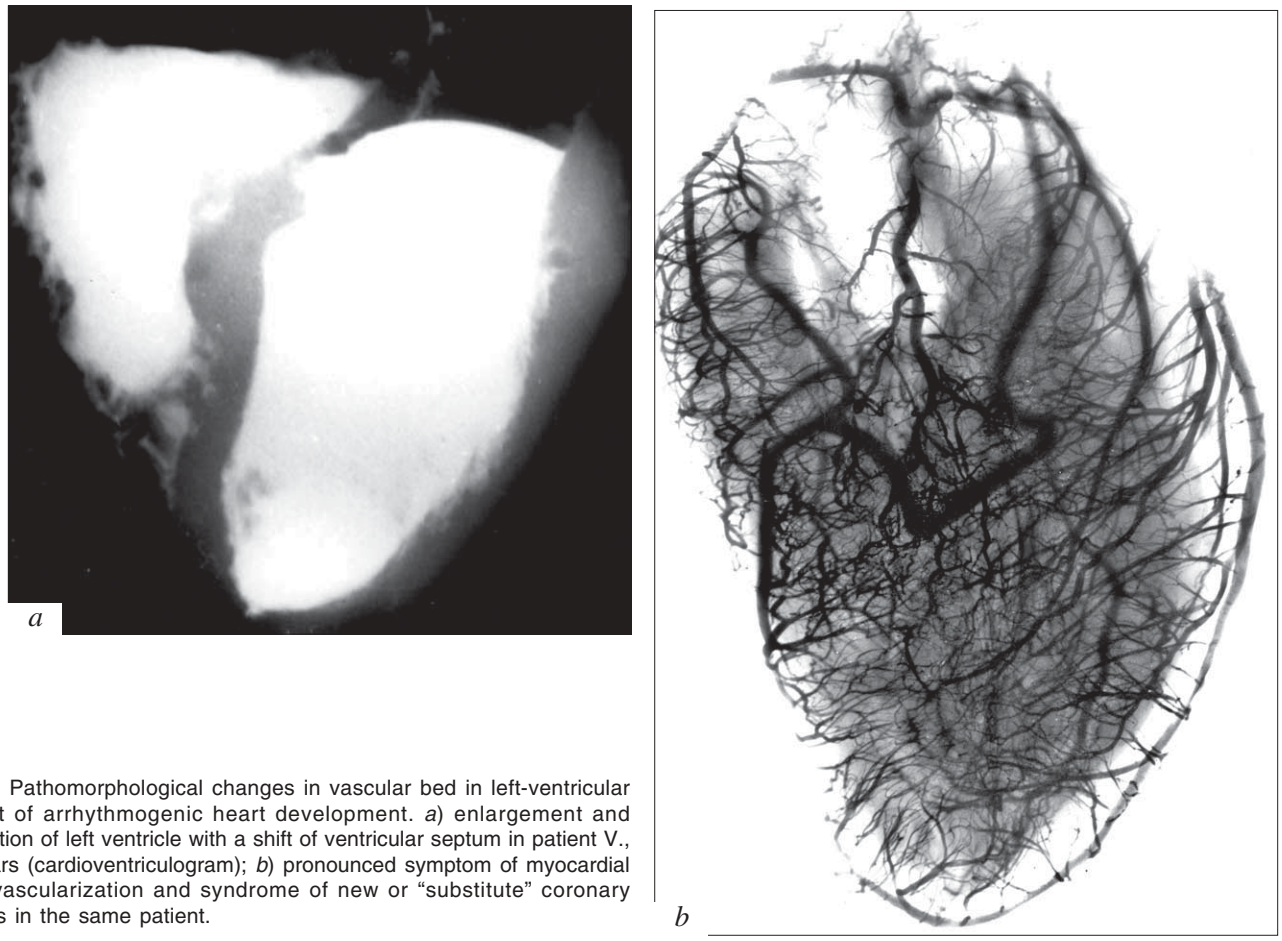

Fig. 1. Pathomorphological changes in vascular bed in left-ventricular variant of arrhythmogenic heart development. *a*) enlargement and elongation of left ventricle with a shift of ventricular septum in patient V., 56 years (cardioventriculogram); *b*) pronounced symptom of myocardial hypervascularization and syndrome of new or "substitute" coronary vessels in the same patient.

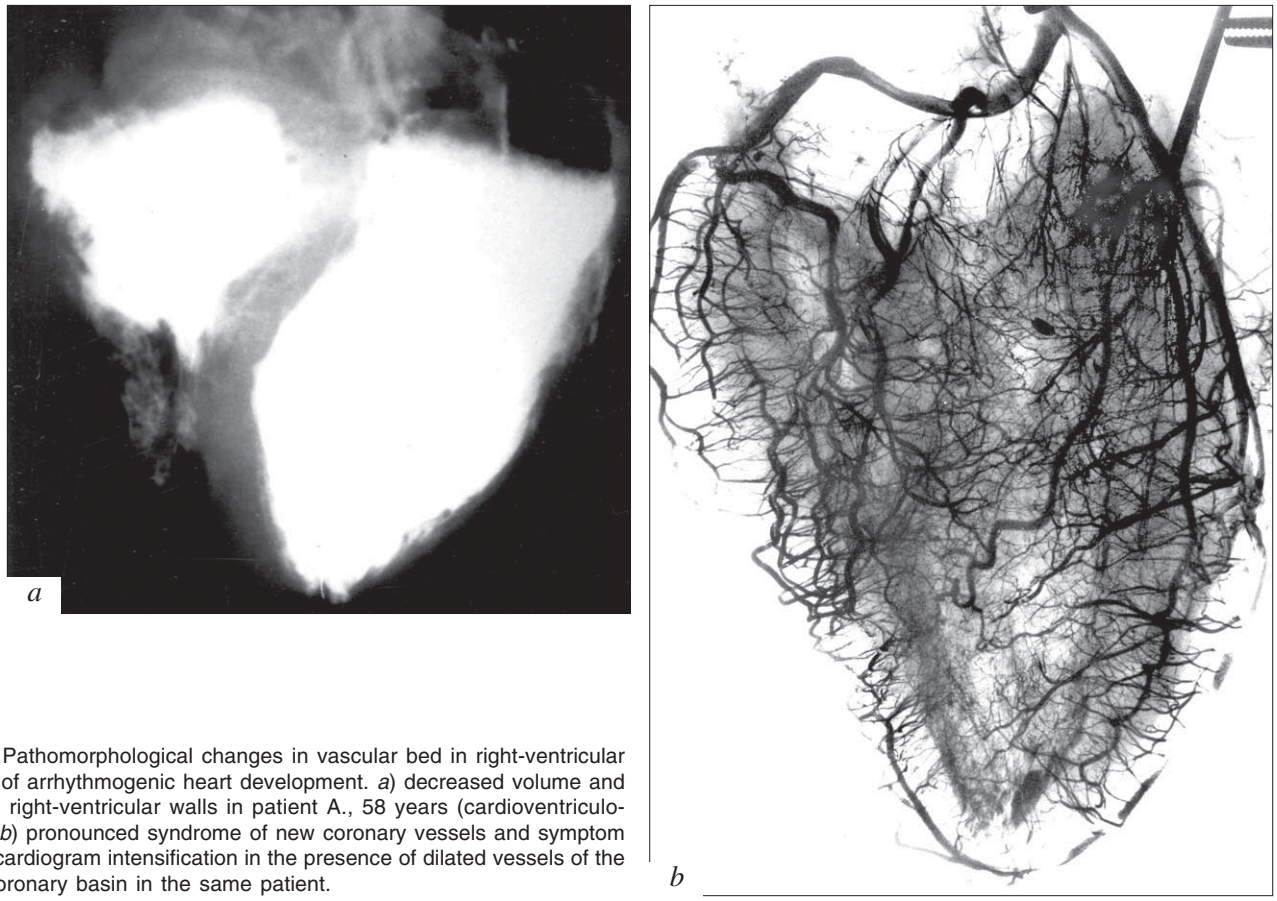

Fig. 2. Pathomorphological changes in vascular bed in right-ventricular variant of arrhythmogenic heart development. *a*) decreased volume and thinned right-ventricular walls in patient A., 58 years (cardioventriculogram); *b*) pronounced syndrome of new coronary vessels and symptom of myocardiogram intensification in the presence of dilated vessels of the main coronary basin in the same patient.

Multipositional analysis of cardioventriculograms and coronarograms showed characteristic features of the coronarographic picture of AH variants. In left-ventricular variant the cavity and volume of the left ventricle were always enlarged, with moderately hypertrophic walls and a slight shift of the interventricular septum towards the right ventricle. The left ventricle acquired an elongated shape (Fig. 1, *a*). Pathognomonic compensatory restructuring of the vascular network of AH with development of myocardial hypervascularization symptom and the syndrome of new or "substitute" coronary bypass corresponded to these cardioventricular changes (Fig. 1, *c*).

Hypervascularization in this variant was achieved at the expense of pronounced collateral blood flow in the intra- and intersystemic fusion of the coronary arteries and their branches. This fusion of collateral vessels was mainly zonal with regard to the left coronary basin. The formation of substitute coronary vessels, in turn, was due to connection of the anterior interventricular branch of the left coronary artery to the right coronary artery, circumflex and anterior interventricular branches of the left coronary artery. Transatrial, epicardial, and subendocardial anastomoses of AH were often involved in the formation of these collaterals.

The parameters of volume density of the left-ventricular vascular network and indexes of blood supply to the heart and myocardium objectively confirm the compensatory shifts in AH angioarchitectonics (Table 1). These indexes corresponded to the transitional criteria and little differed from the normal, which attested to adequate compensatory restructuring of the coronary bed and blood flow level in AH [11].

Decreased cavity and thinning of the right-ventricular walls were pathognomonic for the right-ventricular variant of AH development. The volume of the right ventricle was one-third of the volume of the left ventricle of AH (Fig. 2, *a*). These characteristic manifestations of the so-called arrhythmogenic dysplasia of the right ventricle [14] were paralleled by a combination of signs of compensation and decompensation of AH vascular bed. The symptom of more intensive myocardiogram indicating transition of compensatory processes into decompensation of coronary circulation developed against the background of the new coronary bypass syndrome and compensatory changes characteristic of this syndrome [12]. This symptom manifested in capillary phase of coronarogram combined with dilatation of the vessels in the main coronary basins, including their distal portions (Fig. 2, *b*).

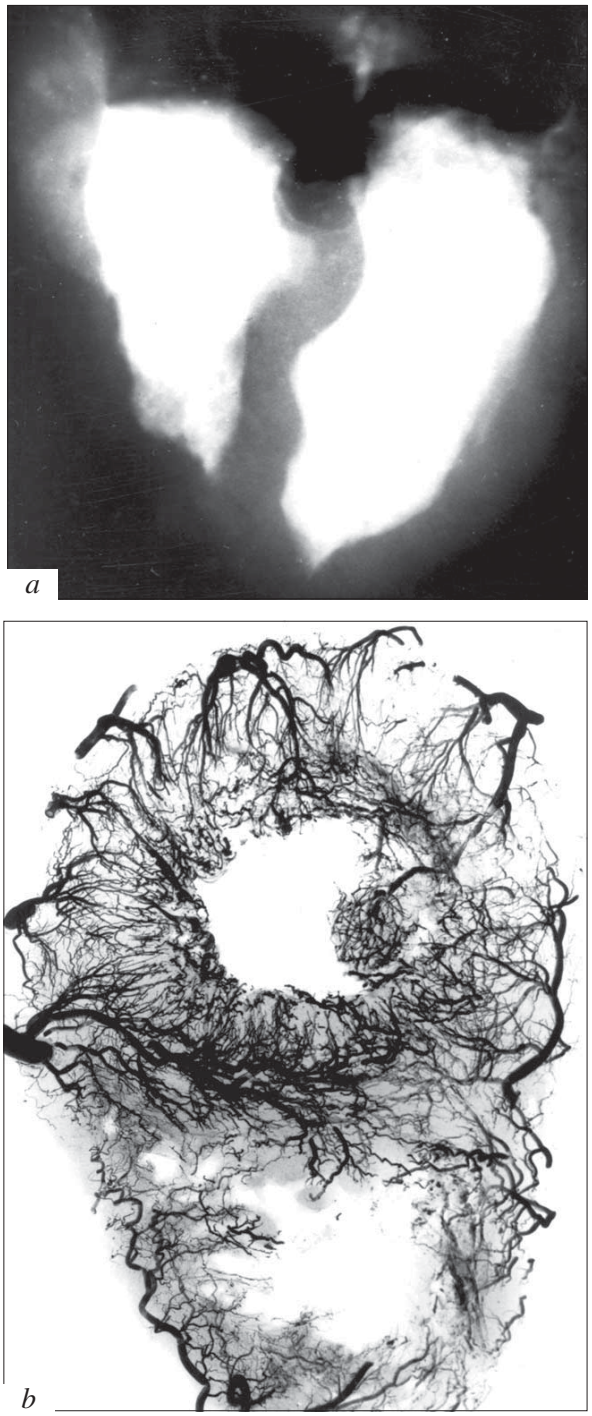

Fig. 3. Pathomorphological changes in vascular bed in septal variant of arrhythmogenic heart development. *a*) sharp wave-like deformation and signs of pathological mobility of the ventricular septum in patient G., 54 years (cardioventriculogram); *b*) numerous small-vessel bundle plexuses and loss of the principal functions by the main coronary arteries in the ventricular septum in the same patient (coronarogram of the transverse median section of the heart).

Enlarged volume density of the vessels confirmed the persisting compensatory trend of changes in AH angioarchitectonics in the right-ventricular variant of AH development. However, this trend did not eliminate the deficient blood supply to AH, which was seen from indexes of blood supply to the heart and myocardium (Table 1).

Pathognomonic sign of the septal variant of AH was moderately hypertrophic ventricular septum with pronounced wave-like deformation and signs of pathological mobility (Fig. 3, *a*). This was paralleled by signs of decompensation of coronary circulation (coronary decompensation triad): syndrome of complete bypass blood flow, decreased coronary accommodation, and symptom of myocardiogram intensification [2]. Coronarographic manifestations of this triad presented as intricate restructuring of AH angioarchitectonics. Zones where the main coronary arteries lost their principal functions with the formation of numerous twisted capillary bundles were seen on coronarograms of transverse sections of the hearts of dead subjects (Fig. 3, *b*). These angioarchitectonic changes in the ventricular septum were responsible for high volume density of vessels in this topographic zone in patients with the septal variant of AH. On the other hand, they had no positive impact on the degree of blood supply to AH, which was clearly seen from blood supply indexes (Table 1) reflecting pronounced disease status.

Deep pathomorphological shifts in the structure of ventricles and ventricular septum in different variants of AH create favorable conditions for the development of extremely intricate vascular changes with separate manifestations of compensation and decompensation of coronary circulation. The totality of these changes leads to clear-cut discrepancy between the weight of AH and its regions and the summary area of coronary arteries lumen. This discrepancy leads to the formation of sufficiently stable ischemic foci in the zones of conduction system, nodal tracts, and in the adjacent areas of the myocardium. These foci become sort of recitative centers of electrical instability and formation of arrhythmogenic substances [9], thus underlying the basis of patho- and thanatogenesis in AH. The type of electrical instability, in turn, determines the specific features in the clinical functional and pathomorphological differential diagnosis of variants of AH development [4,13].

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