

## Chronic heart failure: syndrome or disease?

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### ABSTRACT

The authors of the study analyze various definitions of chronic heart failure (CHF). CHF, having many faces, despite the consensus concerning the paradigm of its pathogenesis, is given different definitions, using both the syndromic and nosological approaches. Most authors share a view of CHF as the final stage (outcome or complication) of many diseases in which there is impairment of ventricular filling or ejection of blood, i.e. as a syndrome, and not an independent nosological form. Nevertheless, at the beginning of the XXI century leading Russian specialists in heart failure presented a reasoned point of view on CHF not only as the final stage of the cardiovascular continuum, complicating the course of a disease of the cardiovascular system, but also as an independent nosological form. This approach, which contradicts the standard rules for the formulation of the final clinical and pathological diagnoses, as well as the agreed positions of the International Statistical Classification of Diseases and Related Health Problems, has been the subject of reasonable criticism. Since the identification of the underlying cause of heart failure is crucial for therapeutic reasons, the only correct view is that of CHF as a syndrome, the detailed description of which in clinical diagnosis is an important intranosological characteristic that allows building the most effective differentiated therapy and accurately determining the prognosis of the disease.

**Key words:** chronic heart failure, definition, International Statistical Classification of Diseases and Related Health Problems, syndrome, disease, rules of diagnosis.

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## Хроническая сердечная недостаточность: синдром или заболевание?

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## РЕЗЮМЕ

В лекции проанализированы различные определения хронической сердечной недостаточности (ХСН). Многоликой ХСН, несмотря на консенсус, касающийся парадигмы ее патогенеза, дают различные определения, используя как синдромальный, так и нозологический подход. Большинство авторов объединяет взгляд на ХСН как на финальную стадию (исход, осложнение) многих болезней, при которых нарушается способность желудочка наполняться кровью или изгонять ее, т.е. как на синдром, а не на самостоятельную нозологическую форму. Тем не менее в начале XXI в. ведущие российские специалисты по сердечной недостаточности представили аргументированную точку зрения на ХСН не только как на конечный этап сердечно-сосудистого континуума, осложняющий течение того или иного заболевания кардиоваскулярной системы, но и как на самостоятельную нозологическую форму. Такой подход, противоречащий стандарту правил формулировки заключительного клинического и патологоанатомического диагнозов, а также согласованным позициям Международной статистической классификации болезней и проблем, связанных со здоровьем, стал предметом обоснованной критики. Так как идентификация причины, лежащей в основе сердечной недостаточности, принципиальна для выбора терапии, единственно правильным является взгляд на ХСН как на синдром, развернутая характеристика которого в клиническом диагнозе представляет собой важную интранозологическую характеристику, позволяющую построить наиболее эффективную дифференцированную терапию и точно определить прогноз заболевания.

**Ключевые слова:** хроническая сердечная недостаточность, дефиниция, Международная статистическая классификация болезней и проблем, связанных со здоровьем, синдром, болезнь, правила формулировки диагноза.

Конфликт интересов. Авторы декларируют отсутствие явных и потенциальных конфликтов интересов, связанных с публикацией настоящей статьи.

Источник финансирования. Авторы заявляют об отсутствии финансирования при проведении исследования.

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## INTRODUCTION

CHF, having many faces, despite the consensus concerning the paradigm of its pathogenesis, is given different definitions using both the syndromic and nosological approaches [1–6]. Most authors share a view of heart failure as the final stage (outcome, complication) of many diseases that affect the heart, i.e. as a syndrome, and not an independent nosological form. In particular, experts from the American College of Cardiology and the American Heart Association, in a lapidary and yet comprehensive definition that has not been revised since 2001, interpret CHF as a complex clinical syndrome that can be caused by

any structural or functional heart disease that interferes with the ability of the ventricle to be filled with blood or expel it [7]. Similar definitions are given in the European, Canadian, British, Korean, Australian-New Zealand, Indian and other guidelines [8–13].

Nevertheless, at the beginning of the XXI century leading Russian experts on heart failure presented a reasoned point of view on heart failure not only as the final stage of the cardiovascular continuum, complicating the course of a particular cardiovascular system disease, but also as an independent nosological form: "... a disease with a complex of characteristic symp-

toms (shortness of breath, decreased physical activity, swelling, etc.), which are associated with inadequate perfusion of organs and tissues at rest or during exercise, and often with fluid retention in the body. The root cause is deterioration in the ability of the heart to fill or empty due to damage to the myocardium, as well as an imbalance of vasoconstrictor and vasodilating neurohumoral systems" [14, 15].

The formal basis for designating CHF as an independent nosological form is the existence of a corresponding heading ("Heart failure" – I50) in the International Statistical Classification of Diseases and Health Problems (10th revision, ICD-10); and informal basis is the idea shared by many cardiologists that CHF develops according to uniform laws, regardless of etiology. It is well known that at the late stage of any disease of the cardiovascular system, "remodeled heart syndrome" pushes into the background the importance of the etiological factor and can independently determine the quality of life and prognosis of the patient [16–18].

Recognizing the formal logic of the arguments presented, we allow ourselves to criticize the nosological interpretation of CHF. Firstly, the presence of the appropriate code in the ICD-10 does not necessarily justify the possibility of its use for encoding the underlying disease. For example, pulmonary embolism (I26) or acute renal failure (N17) is indicated in the diagnosis under the heading "complications of the underlying disease", since replacing the underlying disease with its complication or one of the manifestations is not allowed by the standard rules for formulating the final clinical and pathological diagnoses [19]. However, doctors who have been making diagnostic mistakes for years, ignoring key postulates of the rules for formulating final clinical and pathological diagnoses and calling this clinical experience, are not very rare [2]. Even if we displace from our consciousness the provisions of the rules for the formulation of the diagnosis, the clinician must understand that the diagnosis of "heart failure" (by the way, according to ICD-10, it excludes: conditions caused by arterial hypertension – I11.0, including with kidney damage – I13. -, the consequences of heart surgery or in the presence of a heart prosthesis – I97.1, as well as heart failure in a newborn – P29.0) may be at least some alternative to a detailed diagnosis, say, "ischemic cardiomyopathy" or "dilated cardiomyopathy", only when it is first formulated at the bedside "without past medical history" with

symptoms and signs of heart failure, the corresponding stage III classification of N.D. Strazhesko and V.Kh. Vasilenko, i.e. in a situation where it is extremely difficult to make a reasonable conclusion about the nature of the disease and (or) this, unfortunately, is almost meaningless.

It is another thing when it comes to a patient with initial, yet reversible manifestations of heart failure. In this case, the development of an effective program of rehabilitation treatment is not conceived without an accurate and timely recognition of the etiological essence of the disease underlying heart decompensation. No one will doubt that the management of patients with common occlusive coronary atherosclerosis, mitral stenosis, hemochromatosis or amyloidosis in a cardiology clinic will differ significantly, even if the stage and functional class of heart failure are completely the same, since the best treatment is always etiotropic [2, 20].

Indeed, one of the main target issues of medical diagnosis is the rationale for treatment. Let us turn to the section "Diagnosis, diagnostics" of the Big Medical Encyclopedia [21], in which V.Kh. Vasilenko writes: "For the possibility of a real impact on the patient's condition, knowledge of not only the general nature of the process and its initial and final links, but also the entire chain of phenomena in their sequence and interdependence (pathogenesis) is necessary; only then does it become possible to break the chain in the most accessible and decisive link" [19]. Therefore, the clinical diagnosis should be not only pathogenetic (contain additional intranosological characteristics of the pathological process), but also nosological (meet the requirements of international classifications and the nomenclature of diseases, taking into account the peculiarities of domestic classifications).

Another objective of the diagnosis is a unified statistical study of morbidity and mortality. Let us return to the currently relevant revision of the International Statistical Classification of Diseases and Health Problems. For obvious reasons, the possibility of using the code I50.0 ("Congestive heart failure" – right ventricular failure secondary to left ventricular heart failure), which some experts usually insist on, is limited only to clinical cases with a far advanced stage (IIB–III) of CHF. Therefore, using this code, only part (and not the largest one) of patients with heart failure will be taken into account in statistical reports.

Be that as it may, the experts of the Society of Heart Failure Specialists, the Russian Cardi-

ology Society and the Russian Scientific Medical Society of Therapists in the section “Determining Heart Failure” of the 4th revision of the National Guidelines for the Diagnosis and Treatment of CHF no longer consider the latter as a nosological unit, rightly indicating that “from a practical point of view, heart failure is a syndrome characterized by certain symptoms (shortness of breath, swelling of the ankles, fatigue) and clinical signs (swelling of the cervical veins, small bubbling rales in the lungs, displacement of the apical impulse to the left) resulting from a violation of structure or function of the heart” [22]. However, apologists for the nosological approach to heart failure, while maintaining their beliefs in clinical practice, continue to formulate a diagnosis of heart failure without reference to its etiology.

Such rigidity of thinking requires educational effort. First of all, the fundamental difference between the nosological form and the syndrome regarding their nature should be recalled. In accordance with the industry standard “Terms and definitions of a standardization system in healthcare” (All-Union Standard No. 91500.01.0005-2001, enforced by order of the Ministry of Health of the Russian Federation No. 12 dated 01.22.2001), in the first case, albeit with certain reservations, about monocausal pathology: “... a combination of clinical, laboratory and instrumental diagnostic signs that make it possible to identify a disease... and attribute it to a group of conditions with a common etiology and pathogenesis, clinical manifestations, unified approaches to the treatment and correction of the condition”. Whereas the syndrome is “a condition that develops as a consequence of a disease and is determined by a combination of clinical, laboratory, instrumental diagnostic features that allow it to be identified and assigned to a group of conditions with different etiologies, but common pathogenesis, clinical manifestations, general treatment approaches that also depend on the diseases underlying the syndrome.”

Persistent in the nosological independence of CHF, they are under the illusion that the cause of CHF does not matter or the treatment is the same in all cases: whether it is coronary heart disease, heart disease or tachycardiomyopathy. At the same time, they forget that any structural or functional heart disease underlying CHF has its own specificity, which leaves an imprint on the course of heart failure and prognosis, and identification of the cause underlying CHF is

crucial for the choice of therapy (for example, valvuloplasty or valve prosthetics for malformations, surgical or endovascular revascularization of the ischemic myocardium or rhythmic therapy for tachycardiomyopathy) [8, 23].

Those persistent in the nosological independence of CHF are under the illusion that the cause of CHF does not matter or the treatment is the same in all cases: whether it is coronary heart disease, heart defect or tachycardiomyopathy. At the same time, they forget that any structural or functional heart disease underlying CHF has its own specificity, which leaves an imprint on the course of heart failure and prognosis, and identification of the cause underlying CHF is crucial for the choice of treatment (for example, valvuloplasty or valve prosthetics for defects, surgical or endovascular revascularization of the ischemic myocardium or rhythm-reducing therapy for tachycardiomyopathy) [8, 23].

## CONCLUSION

Thus, the only correct view on CHF is as a syndrome, a detailed description of which in the clinical diagnosis is an important intranosological characteristic that allows for the construction of the most effective differentiated therapy and accurately determining the prognosis of the disease [24].

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