OBLITERATING CARDIOMYOPATHY. PARIETAL FIBROBLASTIC LEFFLER'S ENDOMYOCARDITIS AND DEVS' ENDOMYOCARDIAL FIBROSIS. MODERN METHODS OF EXAMINATION AND TREATMENT

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Abstract. Obliterative Cardiomyopathy in this article. Detailed information about parietal fibroblastic Leffler's endomyocarditis and endomyocardial fibrosis of Devs is given, and modern methods of its examination and treatment are described.

Key words: cardiogram, hereditary diseases, parietal fibroblastic, endomyocardial fibrosis, etc.

Cardiomyopathy is called primary myocardial damage that is not caused by inflammation, tumor or ischemic process. Often, the pathology has an unclear etiology. In order for the patient to be diagnosed with cardiomyopathy, it is necessary to exclude other diseases: congenital defects, heart valve defects, systemic vascular diseases, arterial hypertension, pericarditis, etc. Types of cardiomyopathy There are three main types of cardiomyopathy:

1. dilated cardiomyopathy,
2. hypertrophic cardiomyopathy,
3. restrictive cardiomyopathy.

Dilated cardiomyopathy In this type of cardiomyopathy, the left and right ventricles are stretched and, therefore, the volume of their cavities increases. If the

healthy heart of an adult is about the size of a fist and weighs 240-310 grams, with dilated cardiomyopathy it becomes larger. All this leads to a decrease in myocardial contraction: the force and speed of contraction decrease. An enlarged heart becomes more difficult to pump blood, which leads to the development of progressive heart failure. Dilated cardiomyopathy is 2 times more common in men than in women. Hypertrophic cardiomyopathy In hypertrophic cardiomyopathy, there is an increase in the size of the heart muscle. In addition, hypertrophy is often detected in the area of the interventricular septum. This causes the septum to protrude into the ventricular region and prevents the normal outflow of blood from the ventricle to the aorta when the heart contracts. This phenomenon is called obstruction, so this type of cardiomyopathy is also called obstructive hypertrophic cardiomyopathy. Physical activity is contraindicated in hypertrophic cardiomyopathy: it increases obstruction and causes heart failure. The sensational cases of sudden death of young athletes during the competition were mainly related to this violation. Since it is almost asymptomatic, it looks like a sudden death against the background of iron health. However, in fact, it is a lack of timely diagnosis. In almost 100% of cases, hypertrophic cardiomyopathy is caused by a genetic defect - hereditary or sporadic (random).

The importance of the epicardium for myocardial and valvuloseptal development has been well established; perturbation of epicardial development results in cardiac abnormalities, including thinning of the ventricular myocardial wall and malformations of the atrioventricular valvuloseptal complex. To determine the spatiotemporal contribution of epicardially derived cells to the developing fibroblast population in the heart, we have used a mWt1/IRES/GFP-Cre mouse to trace the fate of EPDCs from embryonic day (ED)until birth. EPDCs begin to populate the compact ventricular myocardium around ED12. The migration of epicardially derived fibroblasts toward the interface between compact and trabecular myocardium is completed around ED14. Remarkably, epicardially derived fibroblasts do not migrate into the trabecular myocardium

until after ED. Migration of EPDCs into the atrioventricular cushion mesenchyme commences around ED. As development progresses, the number of EPDCs increases significantly, specifically in the leaflets which derive from the lateral atrioventricular cushions. In these developing leaflets the epicardially derived fibroblasts eventually largely replace the endocardially derived cells. Importantly, the contribution of EPDCs to the leaflets derived from the major AV cushions is very limited. The differential contribution of EPDCs to the various leaflets of the atrioventricular valves provides a new paradigm in valve development and could lead to new insights into the pathogenesis of abnormalities that preferentially affect individual components of this region of the heart. The notion that there is a significant difference in the contribution of epicardially and endocardially derived cells to the individual leaflets of the atrioventricular valves has also important pragmatic consequences for the use of endocardial and epicardial cre-mouse models in studies of heart development.

Endomyocardial fibrosis (EMF)—also called tropical endomyocardial fibrosis—is a restrictive cardiomyopathy of unknown cause. It is characterized by the deposition of fibrous tissue in the endomyocardium, which leads to a restrictive physiology accompanied by a very poor prognosis without a specific management. The etiopathogenesis of EMF remains in the field of hypotheses and far from exact knowledge; therefore, it demands systematic research with the aid of current technologies. The seemingly implicated factors, besides ethnicity, poverty, eosinophilia, autoimmunity, and serotonin, are related to: (i) the excessive immune response against certain parasitic infections; (ii) dietary scarcity (malnutrition); (iii) herbal preparations; and (iv) the use of improperly processed or cooked cassava as the primary source of carbohydrate (because of the ingestion of toxic levels of cyanogenic glycoside). The occurrence of familial cases supports the participation of genetic predisposition. Although not yet a consensus, the high prevalence of anti-heart antibodies detected in patients with EMF somehow suggests the involvement of autoimmunity in the pathogenesis.

Similarly, the usual association of hypereosinophilia with EMF has led some authors to consider this entity as the tropical variant of hypereosinophilic syndrome, which is found in temperate climates with the overproduction of interleukin and fibrotic lesions identical to those seen in EMF.

Typically, EMF presents an insidious onset, usually associated with fever, pancarditis, and eosinophilia, which are morphologically abnormal. This initial active form also presents dyspnea, itching, and periorbital edema. The clinical features of EMF will depend on the predominantly affected cardiac chamber and the duration of disease. Lower limbs edema, ascites, and non-specific gastrointestinal complaints (e.g. nausea, vomiting, and anorexia) are characteristic of the right ventricular and tricuspid valve involvement. However, when the left ventricle is affected, dyspnea, exertional dyspnea, orthopnea, nocturnal paroxysmal dyspnea, and fatigue will predominate. Thromboembolic events, angina-like chest pain, arrhythmias, and syncope, may also take part of the clinical features of EMF involving the left chamber. Growth retardation, testicular atrophy, clinical feminization, finger and toe clubbing, and cachexia are the consequence of low cardiac output in a chronic disease. Left ventricular involvement, isolated or in combination with biventricular disease, is most often encountered in the chronic form of the disease, followed by isolated right-side involvement. In the latter, the physical examination shows signs of systemic venous hypertension with multi-visceral congestion, accompanied (or not) by pulmonary hypertension due to pulmonary thromboembolism. Some characteristic signs include central cyanosis, exophthalmos, giant ascites without pedal edema (sometimes accompanied by peritoneal fibrosis), hyperpigmentation of the lips and gums, proptosis, and parotid swelling. Although chest x-ray and electrocardiogram may show several abnormalities, none of them are specific. However, the echocardiogram is the gold standard technique for the diagnosis of the chronic disease. Dense endocardial echograms along the mural and valvular endocardium, valvular dysfunction, a restrictive filling pattern with shrinkage of

the cavity, the presence of thrombus, and the detection of pericardial effusion are the most typical echocardiographic findings. MRI adds precision to the diagnosis, showing hypoperfused fibrotic areas, and confirms the presence of thrombus and calcifications.

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